Forty-one Cervicofacial Vascular Anomalies and Their Surgical Treatment – Retrospection and Review
Gavin CW Kang, MBBS, Colin Song, FRCS (Edin), FAMS

Abstract
Introduction: Haemangiomas in children usually involute spontaneously and surgical treatment is exceptional. Vascular malformations do not regress spontaneously and resection may become necessary. We present a series of surgically treated face and neck vascular anomalies during a 9-year period, assessing the epidemiology, presenting signs and symptoms, diagnostic modalities, indications for surgery, treatment methods and clinical outcome post-treatment. Materials and Methods: The medical and pathological records of all patients with cervicofacial vascular anomalies treated surgically at our department from 1997 to 2005 were retrospectively reviewed in relation to current evidence. Results: Forty-one patients were identified. Of these, 9 patients had haemangiomas and the remaining 32 had a variety of vascular malformations. Cervicofacial vascular anomalies were most commonly located at the lip. Atypical looking vascular anomalies like masseteric intramuscular haemangiomas and parotid malformations were diagnostic problems. All 41 had surgical excision of their vascular anomalies for troubling symptoms, cosmesis or diagnostic purpose. For cervicofacial arteriovenous malformations, 28% were classified as Schobinger stage I, 50% stage II, and the remainder stage III. Combined embolisation-resection was used to treat 6 arteriovenous malformations (stage II to III) and of these, 3 required flap reconstruction. Conclusions: Accurate diagnosis distinguishing between cervicofacial haemangiomas and vascular malformations is key to best treatment. The diagnosis can usually be made by history and physical examination aided by early magnetic resonance imaging (MRI). Although cervicofacial haemangiomas can be managed conservatively or with medical therapy, surgery is indicated for preventing psychological distress and in cases of chronic aesthetic alteration resulting from partial regression. Aesthetic concerns and prevention of psychosocial distress point to early excision of venous malformation as the treatment of choice. Lymphatic malformations are best treated by excision. Outcome after excision of localised cervicofacial haemangiomas and low-flow vascular malformations is excellent. Large extensive low-flow malformations as well as those located at the lips may require multiple procedures including reconstruction; patients should be informed that the outcome is generally not as good. Combined embolisation-resection is definitive treatment for arteriovenous malformations and flap reconstruction may prevent their recurrence. Tissue expansion is a useful reconstructive tool after the excision of large vascular anomalies.

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Key words: Face and neck, Haemangioma, Single institution outcome, Vascular malformation

Introduction
Vascular anomalies of the head and neck can be categorised as either haemangiomas or vascular malformations. Haemangiomas are common in young children (hence the term “common infantile haemangioma”) and a conservative wait-and-see treatment is usually favoured because most lesions regress spontaneously.1 Haemangiomas characteristically present shortly after or, in rare cases, at birth with a rapid proliferative phase over the first 12 months. They then stabilise and slowly involute, with regression complete in 50% of cases by age 5 years and in 70% by age 7 years.1 Histologically, there is endothelial hyperplasia and increased number of mast cells.

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during the proliferative phase; the involutive phase is marked by fibrofatty replacement, diminished cellularity and a normal mast cell count.

Surgery is indicated in situations of chronic unaesthetic alteration, psychosocial trauma, bleeding, ulceration, pain and functional compromise. Large periorbital haemangiomas occluding vision and predisposing to ocular complications, head and neck haemangiomas compressing airway or vital structures and labial tumours causing feeding difficulty – all are documented instances of functional compromise with cervicofacial haemangiomas. Vascular malformations, while always present at birth and growing commensurately with body growth, most commonly present in adulthood and do not resolve spontaneously. They are not tumours but collections of abnormal vessels displaying normal flat non-proliferative endothelium and normal mast cell count. Vascular malformations are further separated into low-flow (capillary, venous, lymphatic or combination) and high-flow (arterial component, typically arteriovenous) lesions. Resection is usually necessary, especially for arteriovenous malformations. Preoperative superselective embolisation is increasingly utilised for arteriovenous malformations.

Materials and Methods

All patients with face and neck vascular anomalies treated with surgical resection at our department between 1997 and 2005 were retrospectively reviewed from the medical records of the Singapore General Hospital, the largest tertiary hospital in Singapore.

Data collected included patient age at surgery, sex, race, presenting signs and symptoms, anatomical location of the anomaly, size, diagnostic modality, indication for surgery, surgery performed and other treatment modality, histopathological finding, and outcome after treatment. The vascular anomalies were categorised based on histopathology into haemangiomas and vascular malformations.

Results

Forty-one patients were identified. Of these, 9 had haemangiomas and the remaining 32 had a variety of vascular malformations. All 41 had surgical resection of their face or neck vascular anomalies, with or without the use of other treatment modalities. For the sake of clarity, the patients are discussed separately in the 2 categories.

Haemangioma

The haemangioma series (Table 1) consisted of 3 males and 6 females (male-to-female ratio, 1:2), ranging in age from 3.5 to 48 years (mean, 22.4).

Excluding Case 8 who had an extensive confluent haemangioma affecting most of the face, haemangiomas for the other cases in this series ranged in size from 5 x 5 mm to 30 x 30 mm and were found with equal frequency in each location – lip, cheek, neck and nose.

The presenting signs and symptoms varied according to the tumour site. Every patient had a lump or swelling, with 5 patients reporting multiple symptoms. All tumours were conspicuous where they were located. In all cases, tumour resection was indicated either for diagnostic purpose or for relief of symptoms and signs including incomplete involution, chronic unaesthetic alteration, discomfort, pain, and bleeding. One patient (Case 8) presented with the persistence of a previously treated haemangioma. The patient had undergone surgical excision, laser resection, and even radiotherapy.

Five (55.6%) of the haemangioma patients underwent preoperative computed tomography (CT) or magnetic resonance imaging (MRI) to facilitate diagnosis, determine extent and delineate related anatomy. One (Case 2) had only Doppler ultrasound of the tumour done preoperatively. It was considered necessary to perform diagnostic angiography with preoperative embolisation in Case 8.

With the exception of Case 8, the tumours in the other cases were completely resected surgically with no recurrence at last follow-up (Table 1). For all vascular anomalies, the surgical approach varied depending on the preoperative presumptive diagnosis, location and size of the tumour, as well as surgeon preference. Haemangiomas of the neck were resected via a transcervical approach. The 2 cheek massteric haemangiomas (Cases 5 and 9) were approached as parotid masses and each was excised through a lazy S parotidectomy incision preserving facial nerve branches where necessary. The nasal haemangiomas were both excised via an external rhinoplasty approach. The lip lesions were elliptically excised. In Case 8, the patient had had multiple previous subtotal facial haemangioma resections and a recently skin-grafted palate for a post-resection palatal defect; surgical excision was aimed at palliation in view of her bulky epatulous upper lip and bulky hemipalate with maxillary alveolar protrusion impinging on the dentures.

Apart from minor transient complications encountered with the massteric haemangiomas and Case 8, there were no significant postoperative problems. The follow-up period for this group varied from 5 months to 8 years, with a mean of 2.1 years from the very latest operation.

The results were evaluated for each patient based on the general aspect of the operated area, reduction of tumour volume, correction of functional impairment, improvement of skin texture and cosmetic appearance. The results were classified as very good, good or fair. This method of result
Surgery for Cervicofacial Vascular Anomalies—Gavin CW Kang

assessments were applied uniformly to all vascular anomalies in our entire series. At the latest follow-up for the haemangiomas, the results were very good in 33%, good in 45% and fair in 22% of cases.

Vascular Malformation

The vascular malformation series comprised 18 low-flow lesions (2 lymphangiomas and 16 largely venous malformations) and 14 high-flow lesions (arteriovenous malformations).

Low-Flow Vascular Malformations

Eighteen patients (male-to-female ratio, 1:1.6) aged between 6 and 53 years (mean age, 28.6) were treated for low-flow vascular malformations. All had a lump or swelling of some sort, with 7 (38.9%) having multiple symptoms like recent progressive enlargement (6), pain (3) and itch (1). Eleven (64.7%) noted the lesion at birth or shortly after at a young age. Three had multiple venous malformations distributed over varying areas of the face (Cases 12, 14 and 19). Excluding the patient (Case 16) with extensive venous malformation, the 2 lymphangiomas were primarily sited at the cheek, while the remainder were found at the lip (5) predominantly at the upper lip, frontal (3), cheek (4), neck (3), temporal (1), medial canthus (1) and nose (1). Excluding Case 16, the size of the lesions ranged from 15 x 11 mm to 130 x 30 mm.

Four patients presented with lesion recurrence (Cases 10, 18, 19 and 27) or persistence (Case 16) in spite of prior treatment. Lesion imaging using MRI and/or CT was done in 7 (38.9%) patients, and angiography was performed in cases 16 and 19. Diagnostic biopsy was done in Cases 23 and 27.

Except Case 16, all (Table 2) low-flow vascular malformations were surgically excised completely with no recurrence detected at the latest follow-up. Neck lesions were excised transcervically. The patient in Case 19 presented with recalcitrant recurrent facial lesions that required preoperative embolisation and even adjunctive liposclerotherapy. In case 16 with such a widespread venous malformation, the patient agreed to staged excision and flap reconstruction that was ongoing at the time of writing. The lesion was excised in Case 22 and reconstruction was done utilising tissue expanders – this was complicated by prosthetic infection. One patient (Case 15) required a superficial parotidectomy in order to excise a parotid malformation. Three patients (Cases 12, 14, 19) with lip involvement and 1 patient with a cheek
lymphangioma (Case 26) underwent further minor revision procedures to improve their appearance after the primary surgery. All other low-flow malformations were excised elliptically with direct closure.

There were 2 recurrences (Cases 10 and 19) along follow-up post-excision; the recurrent lesions were again excised with no further recurrence at last follow-up. Postoperative complications were few and minor, involving mainly the cheek lymphangiomas. Case 27 was excised via a parotidectomy approach and suffered transient buccal branch palsy. All complications had resolved by the latest follow-up. The follow-up period for this group ranged from 4 to 72 months, with a mean of about 2.7 years. The results were very good in 6%, good in 72% and fair in 22% of cases.

High-flow Arteriovenous Malformations

The 9 men and 5 women (Table 3) with arteriovenous malformations had an age range of between 10 and 45 years (mean, 29.2). The most common primary zone was the lip (5), predominantly the upper lip, followed by the forehead (3), cheek (3), nose (1), mandible (1) and neck (1). Dimensions of the lesions ranged from 10 x 10 mm to 70 x 30 mm. Four patients presented with recurrence (Cases 35, 37, 39 and 30) of a previously treated high-flow arteriovenous malformation. Every patient in this group noted a lump or swelling; this was associated with progressive enlargement (6), pulsatility and thrill (3), bleeding (2), pain and discomfort (1), positive Valsalva sign (1) and elevated local temperature (1).

Six patients (Cases 30, 33, 35, 37, 40 and 41) noted the congenital history of the lump; in 2 patients (Cases 28 and 29) the arteriovenous malformation was first noted during childhood/puberty. The malformation appeared soon after trauma in 1 patient (Case 32); in the remaining 5 patients, the lesion became apparent in adulthood. MRI and/or CT of the vascular malformation were done in 7 cases and angiogram with embolisation in 6 cases.

Excision (Table 3) alone was used in 8 patients with discrete arteriovenous malformations. Six arteriovenous malformations (43%) – 4 Schobinger stage II and 2 stage III malformations – were treated with superselective embolisation followed by en bloc resection after 24 hours. One patient (Case 37) required a radial forearm free flap for lip reconstruction and later flap debulking. The radial forearm flap was utilised again, this time for right cheek reconstruction in Case 41.

There was recurrence upon follow-up in 1 patient who initially had been treated with resection alone (Case 32); he subsequently had preoperative embolisation and excision of the recurrent (and larger than before) lesion, eventually requiring scalp tissue expansion as part of reconstruction. All other lesions were excised with direct closure. There were no recurrences for all other cases treated. The follow-up period ranged from 3 to 48 months (mean, 2 years). At latest follow-up, the results were very good in 29%, good in 57% and fair in 14% of cases.

Discussion

The appropriate management of vascular anomalies is first dependent on an accurate diagnosis. It is important to distinguish between haemangiomas and vascular malformations, as well as between fast-flow and low-flow vascular malformations. This leads to selection of the most suitable treatment modality for the lesion and appropriate follow-up and assessment of the functional and aesthetic outcome. This approach for the series of anomalies will be discussed separately. We will provide an algorithmic framework for the management of cervicofacial vascular anomalies based on our experience and current evidence, emphasising distinctly the place of surgical excision in overall management.

Haemangioma

Haemangiomas are the most common tumours during infancy, affecting up to 12%, 1.4%, and 0.8% of Caucasian, African American and Asian paediatric populations respectively. There is a female preponderance as was evident from our series. Among affected children, 80% have a solitary lesion while the rest have multiple lesions. Some 60% of haemangiomas are cervicofacial and the bulk of these, appearing shortly after birth and expected to involute with time notwithstanding initial rapid growth, are commonly managed conservatively at paediatric or dermatologic centres. The majority of cases do not present to our centre for plastic surgical excision and treatment – herein lies the significance of this review.

Up to 93% of haemangiomas are easily diagnosed without additional diagnostic tests. Superficial lesions turn raised, bosselated and vivid crimson (strawberry-like). Deeper lesions arise in the lower dermal or subcutaneous planes, appearing with healthy overlying skin or as pale blue or purple masses easily confused with venous malformations. Lesions generally feel firm, rubbery and incompressible, becoming less turgid and fading with involution.

All cases in our series had histology compatible with a diagnosis of haemangioma. Histologically, haemangiomas show plump endothelial cells with multilaminated basement membranes and numerous mast cells; immunohistochemistry demonstrates increased vitronectin, perlecain. The routine hospital use of the immunohistochemical marker GLUT-1 to accurately distinguish haemangiomas (GLUT-1 positive) from vascular malformations has been described and may be practical in our context.

Cases 1, 3, 4, 6, 7, 8, and 9 (Table 1) presented with a
Table 2. Clinical Details of Patients with Face or Neck Low-flow Vascular Malformations

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (y)</th>
<th>Sex</th>
<th>Location</th>
<th>Presenting symptoms and signs</th>
<th>Size (mm²)</th>
<th>Histology</th>
<th>Initial diagnosis</th>
<th>Diagnostic modality and interpretation</th>
<th>Prior treatment</th>
<th>Number of operations: type of surgery</th>
<th>Other treatment involved</th>
<th>Recurrence or complication</th>
<th>Follow-up (months)</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>6</td>
<td>M</td>
<td>Upper lip</td>
<td>Congenital swelling</td>
<td>15 x 11</td>
<td>Venous</td>
<td>Haemangioma</td>
<td>Nil</td>
<td>Nil</td>
<td>2: Excision</td>
<td>nil</td>
<td>Recurrence 4 years after operation</td>
<td>60</td>
<td>Good</td>
</tr>
<tr>
<td>11</td>
<td>7.5</td>
<td>F</td>
<td>Upper lip</td>
<td>Swelling at birth, partial involution by 6 years</td>
<td>20 x 20</td>
<td>Venous</td>
<td>Haemangioma</td>
<td>Nil</td>
<td>Nil</td>
<td>1: Excision</td>
<td>nil</td>
<td>nil</td>
<td>13</td>
<td>Good</td>
</tr>
<tr>
<td>12</td>
<td>15</td>
<td>F</td>
<td>Right cheek and lower lip</td>
<td>Prominent lower lip, regressing cheek lesion</td>
<td>Lower lip – 30 x 10</td>
<td>Venous</td>
<td>Haemangioma</td>
<td>CT – lip lesion haemangioma or lymphangioma</td>
<td>Nil</td>
<td>2: Excision</td>
<td>nil</td>
<td>nil</td>
<td>30</td>
<td>Good</td>
</tr>
<tr>
<td>13</td>
<td>18</td>
<td>F</td>
<td>Neck</td>
<td>Swelling</td>
<td>25 x 25</td>
<td>Venous</td>
<td>Malformation</td>
<td>Salivary tumour or lymph node or lipoma</td>
<td>Nil</td>
<td>1: Excision</td>
<td>nil</td>
<td>nil</td>
<td>11</td>
<td>Very good</td>
</tr>
<tr>
<td>14</td>
<td>19</td>
<td>F</td>
<td>Right cheek and lower lip</td>
<td>Congenital lesions</td>
<td>40 x 35</td>
<td>Venous</td>
<td>Haemangioma</td>
<td>MRI – vascular malformation</td>
<td>Systemic steroid</td>
<td>3: Excision, cheek and lip reconstruction</td>
<td>nil</td>
<td>nil</td>
<td>72</td>
<td>Fair</td>
</tr>
<tr>
<td>15</td>
<td>19</td>
<td>F</td>
<td>Left cheek</td>
<td>Painful swelling</td>
<td>20 x 10</td>
<td>Venous</td>
<td>Malformation</td>
<td>Parotid tumour</td>
<td>CT – slow flow vascular malformation</td>
<td>Nil</td>
<td>1: Superficial parotidectomy</td>
<td>nil</td>
<td>16</td>
<td>Good</td>
</tr>
<tr>
<td>16</td>
<td>22</td>
<td>M</td>
<td>Extensive right cervicofacial involving right chest and upper limb</td>
<td>Persistent recurrent lesions since young</td>
<td>Extensive</td>
<td>Venous</td>
<td>Malformation</td>
<td>Vascular malformation</td>
<td>Angiogram – vascular malformation</td>
<td>Multiple excisions, laser</td>
<td>10: Excision, flap reconstruction</td>
<td>nil</td>
<td>nil</td>
<td>96</td>
</tr>
<tr>
<td>17</td>
<td>28</td>
<td>F</td>
<td>Neck</td>
<td>Congenital swelling, growing and itchy</td>
<td>35 x 20</td>
<td>Venous</td>
<td>Malformation</td>
<td>Nil</td>
<td>Nil</td>
<td>1: Excision</td>
<td>nil</td>
<td>10</td>
<td>Good</td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>30</td>
<td>F</td>
<td>Right temporal</td>
<td>Congenital swelling, previously excised, recurred and painful</td>
<td>30 x 30</td>
<td>Venous</td>
<td>Malformation</td>
<td>Nil</td>
<td>Nil</td>
<td>1: Excision</td>
<td>nil</td>
<td>10</td>
<td>Good</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>31</td>
<td>M</td>
<td>Right upper eyelid</td>
<td>Upper lip and columnella</td>
<td>Recurrence of lesions despite previous intervention</td>
<td>Upper eyelid – 10 x 20</td>
<td>Venous</td>
<td>Malformation</td>
<td>Haemangiomas</td>
<td>Angiogram – upper lip capillovenous malformation</td>
<td>Multiple excisions, embolisation</td>
<td>7: Excision, scar revision</td>
<td>nil</td>
<td>Eyelid recurrence 5 years after first operation</td>
</tr>
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</table>
Table 2. Contd.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (y)</th>
<th>Sex</th>
<th>Location</th>
<th>Presenting symptoms and signs</th>
<th>Size (mm²)</th>
<th>Histology</th>
<th>Initial diagnosis</th>
<th>Diagnostic modality and interpretation</th>
<th>Prior treatment</th>
<th>Number of operations: type of surgery</th>
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<th>Recurrence or complication</th>
<th>Follow-up (months)</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>20</td>
<td>31</td>
<td>F</td>
<td>Neck</td>
<td>Swelling</td>
<td>40 x 30</td>
<td>Venous malformation</td>
<td>Lymph node or sternomastoid tumour</td>
<td>CT – sternomastoid tumour, or haemangioma</td>
<td>Nil</td>
<td>1: Excision</td>
<td>Nil</td>
<td>Nil</td>
<td>4</td>
<td>Good</td>
</tr>
<tr>
<td>21</td>
<td>35</td>
<td>F</td>
<td>Medial canthus</td>
<td>Swelling</td>
<td>5 x 5</td>
<td>Venous malformation</td>
<td>Vascular malformation</td>
<td>Nil</td>
<td>1: Excision</td>
<td>Nil</td>
<td>Nil</td>
<td>5</td>
<td>Good</td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>43</td>
<td>M</td>
<td>Frontal</td>
<td>Swelling from young</td>
<td>130 x 30</td>
<td>Venous malformation</td>
<td>Haemangioma</td>
<td>Nil</td>
<td>Sclerotherapy</td>
<td>Nil</td>
<td>Infected tissue expander</td>
<td>18</td>
<td>Good</td>
<td></td>
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<tr>
<td>23</td>
<td>43</td>
<td>M</td>
<td>Frontal</td>
<td>Swelling</td>
<td>40 x 40</td>
<td>Venous malformation</td>
<td>Lipoma</td>
<td>Incision biopsy – venous malformation</td>
<td>Nil</td>
<td>1: Excision</td>
<td>Nil</td>
<td>Nil</td>
<td>53</td>
<td>Good</td>
</tr>
<tr>
<td>24</td>
<td>52</td>
<td>M</td>
<td>Frontal</td>
<td>Congenital lesion growing proportionately with age</td>
<td>37 x 18</td>
<td>Venous malformation</td>
<td>Vascular malformation</td>
<td>Nil</td>
<td>1: Excision</td>
<td>Nil</td>
<td>Nil</td>
<td>12</td>
<td>Good</td>
<td></td>
</tr>
<tr>
<td>25</td>
<td>53</td>
<td>F</td>
<td>Nasal tip and right cheek</td>
<td>Pedunculated nasal swelling after childbirth Congenital right cheek capillary malformation</td>
<td>15 x 13</td>
<td>Capillary-venous malformation</td>
<td>Vascular malformation</td>
<td>MRI – haemangioma or low flow vascular malformation</td>
<td>Nil</td>
<td>1: Excision</td>
<td>Laser to capillary malformation</td>
<td>Nil</td>
<td>12</td>
<td>Good</td>
</tr>
<tr>
<td>26</td>
<td>28</td>
<td>F</td>
<td>Right cheek</td>
<td>Swelling</td>
<td>20 x 20</td>
<td>Lymphangioma</td>
<td>Lymphangioma</td>
<td>Nil</td>
<td>Liposuction</td>
<td>Laser to scar revision</td>
<td>Haematoma</td>
<td>21</td>
<td>Fair</td>
<td></td>
</tr>
<tr>
<td>27</td>
<td>35</td>
<td>M</td>
<td>Left cheek</td>
<td>Previous excision of cheek lymphangioma Now recurrent swelling</td>
<td>40 x 40</td>
<td>Lymphangioma</td>
<td>Lymphangioma</td>
<td>MRI – lymphangioma Trucut – lymphoid cells</td>
<td>Excision</td>
<td>2: Excision</td>
<td>Facial (buccal) palsy</td>
<td>12</td>
<td>Good</td>
<td></td>
</tr>
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</table>

Table 3. Clinical Details of the 14 Patients With Face or Neck High-flow Vascular (Arteriovenous) Malformations

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (y)</th>
<th>Sex</th>
<th>Location</th>
<th>Presenting symptoms and signs</th>
<th>Size (mm²)</th>
<th>Histology</th>
<th>Initial diagnosis</th>
<th>Diagnostic modality and interpretation</th>
<th>Prior treatment</th>
<th>Number of operations: type of surgery</th>
<th>Other treatment involved</th>
<th>Recurrence or complication</th>
<th>Follow-up (months)</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>28</td>
<td>10</td>
<td>F</td>
<td>Right cheek</td>
<td>Swelling with thrill</td>
<td>25 x 15</td>
<td>II</td>
<td>AVM</td>
<td>MR angiogram – AVM</td>
<td>1: Excision</td>
<td>Nil</td>
<td>Nil</td>
<td>Nil</td>
<td>7</td>
<td>Good</td>
</tr>
<tr>
<td>29</td>
<td>14</td>
<td>M</td>
<td>Nasal dorsum</td>
<td>Swelling that enlarges upon dependency</td>
<td>10 x 10</td>
<td>II</td>
<td>AVM</td>
<td>MRI – small haemangioma</td>
<td>Nil</td>
<td>Nil</td>
<td>Nil</td>
<td>Nil</td>
<td>5</td>
<td>Good</td>
</tr>
<tr>
<td>30</td>
<td>16</td>
<td>M</td>
<td>Left mandible</td>
<td>Congenital swelling with left eye proptosis and blindness Lower jaw gum bleeding</td>
<td>40 x 30</td>
<td>II</td>
<td>AVM</td>
<td>CT - AVM MRI - AVM</td>
<td>Preoperative embolisation</td>
<td>1: Excision</td>
<td>Nil</td>
<td>Nil</td>
<td>Nil</td>
<td>48</td>
</tr>
<tr>
<td>Case</td>
<td>Age (y)</td>
<td>Sex</td>
<td>Location</td>
<td>Presenting symptoms and signs</td>
<td>Size (mm²)</td>
<td>Histology</td>
<td>Initial diagnosis</td>
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</tr>
<tr>
<td>31</td>
<td>20</td>
<td>M</td>
<td>Forehead</td>
<td>Pulsatile swelling</td>
<td>25 x 16</td>
<td>II AVM</td>
<td>Nil</td>
<td>Nil</td>
<td>1: Excision</td>
<td>Nil</td>
<td>Nil</td>
<td>5</td>
<td>Good</td>
<td></td>
</tr>
<tr>
<td>32</td>
<td>21</td>
<td>M</td>
<td>Forehead</td>
<td>Posttraumatic forehead swelling</td>
<td>30 x 20</td>
<td>II Traumatic aneurysm</td>
<td>Angiogram – AVM</td>
<td>Preoperative embolisation</td>
<td>7: Excision, tissue expansion, scalp reconstruction</td>
<td>Nil</td>
<td>Recurrence of AVM 2 years after first operation Tissue expander infection</td>
<td>48</td>
<td>Fair</td>
<td></td>
</tr>
<tr>
<td>33</td>
<td>21</td>
<td>F</td>
<td>Forehead</td>
<td>Congenital discoloured swelling with positive Valsalva sign</td>
<td>10 x 10</td>
<td>I Cavernous malformation</td>
<td>Nil</td>
<td>Nil</td>
<td>1: Excision</td>
<td>Nil</td>
<td>Nil</td>
<td>3</td>
<td>Very good</td>
<td></td>
</tr>
<tr>
<td>34</td>
<td>22</td>
<td>M</td>
<td>Left cheek</td>
<td>Compressible swelling with discomfort</td>
<td>32 x 25</td>
<td>III Haemangioma</td>
<td>MRI – Haemangioma</td>
<td>Preoperative embolisation</td>
<td>1: Excision</td>
<td>Nil</td>
<td>Facial (mandibular) palsy</td>
<td>56</td>
<td>Good</td>
<td></td>
</tr>
<tr>
<td>35</td>
<td>26</td>
<td>F</td>
<td>Lower lip</td>
<td>Congenital swelling with lip ectropion</td>
<td>30 x 32</td>
<td>II Vascular malformation</td>
<td>Nil</td>
<td>Excision, embolisation</td>
<td>1: Excision of recurrence</td>
<td>Laser, sclerotherapy</td>
<td>Nil</td>
<td>12</td>
<td>Good</td>
<td></td>
</tr>
<tr>
<td>36</td>
<td>32</td>
<td>M</td>
<td>Neck</td>
<td>Swelling</td>
<td>20 x 20</td>
<td>I Lymph node</td>
<td>CT – cold abscess or TB adenitis</td>
<td>Nil</td>
<td>1: Excision</td>
<td>Nil</td>
<td>Nil</td>
<td>13</td>
<td>Very good</td>
<td></td>
</tr>
<tr>
<td>37</td>
<td>35</td>
<td>M</td>
<td>Upper lip</td>
<td>Congenital swelling with elevated local temperature</td>
<td>70 x 30</td>
<td>II AVM</td>
<td>MR angiogram – Haemangioma – AVM</td>
<td>Partial resection, laser, sclerotherapy Preoperative embolisation</td>
<td>2: Excision of recurrence, free flap reconstruction</td>
<td>Nil</td>
<td>Nil</td>
<td>96</td>
<td>Very good</td>
<td></td>
</tr>
<tr>
<td>38</td>
<td>39</td>
<td>M</td>
<td>Upper lip</td>
<td>Pulsatile swelling with thrill and bleeding</td>
<td>20 x 10</td>
<td>III AVM</td>
<td>Nil</td>
<td>Nil</td>
<td>1: Excision</td>
<td>Nil</td>
<td>Nil</td>
<td>4</td>
<td>Good</td>
<td></td>
</tr>
<tr>
<td>39</td>
<td>44</td>
<td>F</td>
<td>Upper lip</td>
<td>Swelling</td>
<td>30 x 32</td>
<td>I AVM</td>
<td>MRI – AVM</td>
<td>Excision</td>
<td>1: Excision of recurrence</td>
<td>Nil</td>
<td>4</td>
<td>Good</td>
<td></td>
<td></td>
</tr>
<tr>
<td>40</td>
<td>45</td>
<td>F</td>
<td>Upper lip</td>
<td>Congenital swelling</td>
<td>30 x 10</td>
<td>I Haemangioma</td>
<td>Nil</td>
<td>Intrallesional steroid, laser</td>
<td>2: Excision, skin grafting</td>
<td>Nil</td>
<td>Nil</td>
<td>12</td>
<td>Good</td>
<td></td>
</tr>
<tr>
<td>41</td>
<td>45</td>
<td>M</td>
<td>Right cheek</td>
<td>Congenital swelling</td>
<td>30 x 10</td>
<td>II AVM</td>
<td>Nil</td>
<td>Preoperative embolisation</td>
<td>1: Excision, free flap reconstruction</td>
<td>Nil</td>
<td>Nil</td>
<td>24</td>
<td>Very good</td>
<td></td>
</tr>
</tbody>
</table>

AVM: Arteriovenous malformation
Annals Academy of Medicine

Surgery for Cervicofacial Vascular Anomalies—Gavin CW Kang

lesion shortly after or at birth. The lesions in Cases 1, 4, 6 and 8 were superficial and brighter. In Case 3 the lesion was more dermal and exhibited a purplish blue hue. The remaining 4 patients had haemangiomas that did not resemble any vascular anomaly and became apparent only later in life, manifesting as aesthetic worry or symptoms for concern (swelling, pain, tenderness, discomfort). The haemangiomas in these 4 patients were the largest in our series and this seems to suggest that large cervicofacial haemangiomas may look atypical and are also more likely to cause pain and discomfort. One (Case 2) complained of a tender right neck lump that turned out to be a partially regressed subcutaneous haemangioma. Two patients (Cases 5 and 9), diagnosed initially with a lipoma and a parotid tumour respectively, were distressed by the presence of a cheek lump in adulthood – in each a masseteric intramuscular haemangioma was excised. Intramuscular haemangiomas, which are uncommon to begin with, are even rarer in the head and neck, constituting 0.8% of all haemangiomas in the region.8 When present the masseter is most frequently involved. They may be painful and are often misdiagnosed preoperatively. The provisionally diagnosed nasal dermoid in Case 7 turned out to be a capillary haemangioma. These behave differently from common infantile haemangiomas in that they tend not to involute.

Deeper lesions mimicking other lesions, questionable superficial lesions and vascular-like lesions presenting in later life (to exclude malignancy and the occasional non-involuting or partially involuted haemangioma) require imaging. CT and MRI are useful in confirming diagnosis and for evaluating the extent of the haemangioma, intracranial involvement or any associated abnormalities (such as in the PHACE syndrome in which extensive facial haemangiomas are linked to the central nervous system, vascular, cardiac and ocular anomalies).9,10 MRI of a haemangioma demonstrates a lobulated soft tissue mass with flow voids; T1 images are isointense or hypointense to muscle and T2 images are hyperintense; gadolinium provides intense homogenous enhancement. CT provides similar findings. Tumours bearing little resemblance to haemangiomas (Cases 5, 7, 9) were diagnostic problems solved with the aid of MRI and CT. The diagnosis of haemangioma was suggested or made in 100% of the CT or MRI scans done. The Doppler ultrasound utilised in Case 2 provided a mis-

Table 4. Details of Embolised Face or Neck High-Flow Vascular (Arteriovenous) Malformations

<table>
<thead>
<tr>
<th>Case</th>
<th>Location</th>
<th>Vessels</th>
<th>Embolisation technique</th>
<th>Embolisation result</th>
</tr>
</thead>
<tbody>
<tr>
<td>28</td>
<td>Right cheek</td>
<td>Feeder – right facial artery (superior labial branch)</td>
<td>PVA of 300 to 600 microns with occlusion of the facial artery supply</td>
<td>90% reduction in the filling of AVM with only residual filling noted at superior facial artery</td>
</tr>
<tr>
<td>30</td>
<td>Left mandible</td>
<td>Feeder – left external carotid artery (left facial branches, left internal maxillary branches, distal left external carotid artery)</td>
<td>PVA of 335 to 500 microns followed by gel foam pledgets</td>
<td>Significant reduction in flow through AVM</td>
</tr>
<tr>
<td>32</td>
<td>Forehead</td>
<td>Feeder – ophthalmic arteries, branches of superficial temporal artery</td>
<td>PVA of 255 to 500 microns was administered</td>
<td>Residual AVM supply from ophthalmic artery sympathetic branch and superficial temporal veins</td>
</tr>
<tr>
<td>34</td>
<td>Left cheek</td>
<td>Feeder – branches of left internal maxillary artery and left temporal artery</td>
<td>PVA of 150 to 255 microns and gel foam pledgets</td>
<td>80% to 90% reduction of flow residual supply from left transverse facial artery and small vessels</td>
</tr>
<tr>
<td>37</td>
<td>Upper lip</td>
<td>Feeder – facial artery and branches of left internal maxillary artery and facial artery</td>
<td>PVA and gel foam</td>
<td>Significant reduction in flow through AVM</td>
</tr>
<tr>
<td>41</td>
<td>Right cheek</td>
<td>Feeder – facial artery</td>
<td>PVA and gel foam</td>
<td>Significant reduction in flow through AVM</td>
</tr>
</tbody>
</table>

AVM: arteriovenous malformation; PVA: polyvinyl alcohol
taken diagnosis. Nevertheless, an ultrasound in experienced hands is a portable and available tool that can easily confirm a suspected haemangioma without additional testing.\footnote{1} Doppler colour flow imaging is notable for its ability to distinguish between high-flow and low-flow lesions.

In children, uncomplicated haemangiomas can be managed conservatively (“masterful neglect”), since 70% of haemangiomas involute spontaneously by age 7 and there is continued improvement in the remaining until age 10 to 12. However, periorbital, intranasal and laryngeal, and parotid haemangiomas may threaten vision, airway and hearing respectively; bleeding, ulceration, infection, or even congestive cardiac failure can complicate haemangiomas. These complicated haemangiomas deserve active treatment and a wide array of both medical and surgical therapeutic options is available—pharmacotherapy (steroids, interferon, chemotherapy), surgical excision, and less commonly, laser, embolisation, radiation, cryotherapy and compression.\footnote{5,6} Pharmacotherapy is frontline treatment for complicated haemangiomas, with surgical excision indicated in those unresponsive to pharmacologic therapy, those with psychological problems, and those with partially involuted or non-involuting unaesthetic lesions.

Surgical excision was carried out in all our cases. Unsurprisingly, in our series of mainly adult patients, chronic unaesthetic alteration (56%) and partially regressed haemangioma (44%) were the most common principal indications, followed by need for diagnosis (44%), and symptoms of pain, tenderness and discomfort (33%). By chronic aesthetic alteration we refer to disruption of the involved aesthetic unit with excessive tissue or skin irregularities that are psychologically debilitating. For the patient in Case 8, her minimally regressed facial haemangioma engulfing her central face and extending into the palate was grotesque and a source of great psychological distress in spite of multiple prior debulking interventions. With such extensive cervicofacial haemangiomas, surgery serves only a palliative role. Children with disfiguring cervicofacial haemangiomas are susceptible to psychological body image problems, especially at the school-going age of 4 to 5, yet for these children judging the optimal timing for surgical intervention is difficult since it is not possible to predict the timing of involution. Earlier haemangiomas that present after childhood would not be surgically treated until age 8 to 12. However authors are now making a paradigm shift in thinking towards surgical intervention in children from 2 or 3 years of age.\footnote{1,12,13} One child (Case 1) was troubled with frequent bleeding from a neck haemangioma that mandated early surgical excision. She incidentally has the Klippel-Trenaunay syndrome characterised in her case by right upper limb capillary malformation and hypertrophy. The presence of a partially regressed haemangioma on the nose (Cases 4 and 7) causes great psychological distress to a young adult. In extreme cases, the haemangioma can create a Cyrano de Bergerac nasal deformity,\footnote{14} and a case of a 4-year-old girl who tried to remove her nasal tip haemangioma with scissors has been reported.\footnote{15} The tumour mass effect can cause nasal cartilage aplasia in young children; also, they are slow to regress and contour deformities may result from the remnant fibrofatty tissue after any involution. Surgical excision with preservation of anatomy seems to be the treatment of choice. Various external rhinoplasty approaches allowing for open access, correction of cartilaginous disruption and cosmesis have been suggested.\footnote{15,16} Both our cases did well after excision by external rhinoplasty. For larger lesions, excision and reconstruction by local tissue transfer or rearrangement may be necessary.

Lip haemangiomas were encountered twice as partially involuted lesions in our series. Zide et al\footnote{12} feel that lip haemangiomas may not spontaneously involute as often as or to the degree observed in haemangiomas involving other facial areas. Moreover, this limited involution may result in an abnormal distorted lip contour as the endpoint. Early surgical treatment is advocated while observing key tenets that include avoidance of aggressive muscle excision, the fact that surgery and secondary intention healing may expedite involution, and avoidance of denervation by making reductions in a central or paracentral fashion.

Morbidity was associated with cheek tumours and extensive haemangiomas. Postoperative dysesthesia with the massteric haemangiomas was probably caused by iatrogenic neuropraxia of the surrounding cutaneous nerves; the right facial pain in Case 9 settled with transcutaneous electrical nerve stimulation. Our results show that outcome and prognosis after surgical excision of localised cervicofacial haemangiomas is excellent.

**Low-flow Vascular Malformations**

Vascular malformations are often present at birth and grow commensurately with the patient, usually only becoming significant later in childhood. There is generally no spontaneous involution. Venous and lymphatic malformations are low-flow lesions, while malformations with an arterial component (most commonly arteriovenous) are considered high-flow lesions. All 18 cases in our series had histology compatible with a diagnosis of vascular malformation,\footnote{2} displaying normal non-proliferative endothelium, normal basement membranes and a normal number of mast cells.

**Venous Malformations**

Like haemangiomas, the diagnosis of a venous malformation is usually straightforward at clinical
examination. Venous malformations are the most common symptomatic vascular malformations, becoming symptomatic in older children or young adults with blush skin discoloration, local swelling and pain. They are characteristically soft and compressible blush masses that engorge with dependency; deep lesions may not be visible at birth.\textsuperscript{4,14} Cervicofacial venous malformations can range from tiny lesions to huge vascular malformations that cause significant facial asymmetry and progressive distortion. No sexual predilection is recognised.

As is usually the case with venous malformations,\textsuperscript{4,14} the majority in our series had swellings noted at birth or soon after that grew with the patient and, upon obtaining full growth remained stable until presentation to medical attention much later either for aesthetic or other concerns. Those with onset of clinical symptoms at a later age and/or recent progressive enlargement in all likelihood had deeper inconspicuous malformations whose acute enlargement was triggered by some unknown or unmemorable traumatic or hormonal event, much like for high-flow lesions. The nasal malformation in Case 25 in particular appeared after the birth of her child, becoming cosmetically undesirable later in life. In our series, more than a quarter of low-flow vascular malformations were located at the lip (notably the upper lip) making it the most common site of occurrence. One left cheek lesion (Case 15) was preoperatively diagnosed as a parotid tumour but found intraoperatively to be an infrequently encountered parotid venous malformation. This patient presented with 3 months of a parotid swelling that fluctuated in size through the day and caused pain whenever it enlarged; her features were typical of previously reported parotid vascular malformations.\textsuperscript{17}

The initial preoperative diagnosis was correct in 6 of 16 patients (37.5\%) while an equal number were mistakenly diagnosed with haemangiomas. This was more likely to occur in younger patients (Cases 10, 11, 12 and 14) in whom the venous malformation would have been easily diagnosed as a partially regressed haemangioma. The remaining were diagnosed as salivary tumours, lymph nodes, lipomas, and other soft tissue tumours.

Imaging is required for indeterminate lesions and to assess the extent of the lesion and associated abnormalities. Some large vascular malformations are associated with sinus pericranii and developmental intracranial venous anomalies.\textsuperscript{18} Those of trigeminal localisation are associated in 15\% of patients with glaucoma, or choroidal and leptomeningeal haemangiomas. On radiographs and CT scans, they typically appear as soft tissue masses containing phleboliths. Venous malformations are high-signal intensity lesions on MRI T2 images and low-signal intensity lesions on T1 images; they have lobulated margins and multiple round signal voids representing phleboliths. Their gadolinium contrast enhancement is more patchy and central, compared to lymphatic malformations, which have no or minimal peripheral enhancement. MRI was done in 2 cases, CT in 3 cases and angiography in 2 cases. All MRI scans, 2 CT scans and both angiograms were able to diagnose or suggest the diagnosis of a vascular malformation. Low-flow vascular malformations on imaging were usually confused with haemangiomas in our series. Angiography was applicable to Case 16 with an extensive right cervicofacial and upper body vascular malformation in helping to differentiate venous malformations from high-flow vascular anomalies; it demonstrated low-flow, low shunt vascular dynamics but does not have diagnostic value in assessing low-flow anomalies. Valuable information obtained included its arterial supply, the absence of fast-flow lesion or aneurysm, and the absence of intracranial aneurysm or concurrent arteriovenous malformation. Biopsy performed in 2 cases provided histopathological confirmation but is usually unnecessary unless history, physical examination or imaging is confounding.

While venous malformations in most areas can be treated conservatively with the use of compressive therapy for local pain and swelling and aspirin for thrombosis, cervicofacial venous malformations are cosmetically significant and best treated actively. This may entail the use of sclerotherapy, embolisation, surgical resection or a combination of these techniques. Sclerosing agents used in our series included sodium tetradecyl sulphate and thrombavar.

Because venous malformations do not involute and grow commensurately with the patient even turning nodular and thickened with age, early surgical intervention is appropriate and psychosocially beneficial.\textsuperscript{14,19} Based on our experience, complete surgical excision is probably the best definitive treatment for discrete small to moderate-sized venous malformations considering the minimum morbidity and low overall recurrence rate. It may be better to do MRI imaging at an early stage (possibly by age 5) for vascular anomalies that are diagnosed as haemangiomas but not regressing as quickly as expected. This allows earlier identification of a misdiagnosed venous malformation that can then be expediently excised, preventing a futile wait for spontaneous involution and associated psychosocial distress. This also avoids ineffective treatment such as steroid (Case 14). In the recurrent cases, we observe that recurrent lesions (Cases 16, 19) were more likely to recur after excision; also, a recurrent lesion may grow to be much larger than the original lesion (Case 10).

Nearly a third of the venous malformations were found on the lips. Of these 5 lip anomalies, 3 were on the upper lip. Lip lesions tended to require multiple procedures, be it...
for excision of recurrence or revision, and the aesthetic result was generally not as good as for malformations in other locations. Surgeons should anticipate the need for multiple procedures and manage patient expectations accordingly. Zide et al\(^\text{19}\) recommended the following tenets for optimal surgical outcome: for upper lip lesions – minimal (about 10%) over-correction of deformities, adjustment of the affected side using the contralateral side as an exact template, correction of vermillion discrepancies as a final stage with expectation of future revisions in this area especially due to gravity; for lower lip lesions – overcorrect (about 20%) deformities expecting the need for revisions, and step the vermilion in large central and vertical reductions to reduce notching.

Complex cervicofacial malformations (Cases 16, 19, 22) may be more suited to a combination of preoperative embolisation, resection and sclerotherapy, and if needed followed by ingenious reconstruction using skin grafts, local tissue flaps or free flaps.\(^\text{14}\) Multiple procedures and operations were often performed for large or multi-focal vascular malformations; they appeared to have a high risk of recurrence and the surgical result was not as good in general. In Case 16, an extensive venous malformation required judicious staged debulking and reconstruction using a pedicled contralateral deltopectoral flap. The deltopectoral flap was delayed and “waltzed” stepwise from the opposite chest wall and strategically inset into the serially created right-sided excision defect. At the time of writing this patient would require further surgery to clear the vascular anomaly satisfactorily. Case 22 with a large frontal malformation needed a tissue expanded local flap for defect coverage and did well despite superficial prosthetic infection. Jackson et al\(^\text{20}\) compartmentalised massive head and neck vascular malformations using non-absorbable sutures followed by large dose sclerosant injection into each compartment; this reduces the lesion vascularity for a safer subsequent resection and for haemostasis in life-threatening haemorrhage. There were no instances of haemorrhage as a complication in our series.

**Lymphatic Malformations**

These lesions, also known as lymphangiomas or lymphovenous malformations, are usually present at birth and their appearance varies ranging from blebs to large or multiloculated cysts to the diffuse involvement of a body part or organ. They are classified as microcystic, macrocystic or mixed. About 75% of lymphatic malformations are cervicofacial. Again, most cases are clinically obvious and require no imaging studies for diagnosis.

However, MRI is especially useful for full evaluation: lymphatic malformations have low signal-intensity on T1 images and marker hyperintensity on T2; most are non-enhancing after intravenous contrast – this is the main MRI feature distinguishing lymphatic and venous malformations. Only Case 27 had an MRI done that demonstrated a recurrent lymphangioma.

These lesions rarely resolve spontaneously and surgical excision is the most commonly accepted form of treatment.\(^\text{14}\) Other therapeutic modalities are less useful and although suction-assisted lipectomy has been used, this has not been effective. The patient in Case 26 had been treated with liposuction twice without satisfactory effect and required surgical resection of the lymphangioma.

**High-flow Arteriovenous Malformations**

Like most vascular malformations, arteriovenous malformations are usually present at birth but may not be clinically evident. The arteriovenous malformation was recognised at initial presentation in most patients (50%), while in others it was suspected to be a venous vascular malformation, haemangioma, aneurysm or lymph node. Expansion involves increased blood flow, collateral formation and recruitment of normal adjacent vessels. Of these 14 patients, 78.6% of the arteriovenous malformations became apparent either at birth or in adulthood, with equal incidence for both. The majority of our arteriovenous malformations were found at the lip (36%), cheek (21%) or forehead (21%), and there were no auricular malformations in our series, in contrast to the largest series to date of arteriovenous malformations (n = 81) in which cheek (31%) and ear (16%) were the most common locations.\(^\text{21}\)

Arteriovenous malformations may progress acutely due to activating stimuli like trauma, pregnancy, puberty, infection, or even iatrogenic trauma (biopsy, proximal feeder ligation, or subtotal excision).\(^\text{5,14,21}\) The patient in Case 32 had run into a tree trunk, traumatising his forehead. The arteriovenous malformation in Case 29 was noted in childhood but suddenly expanded at puberty to become more obvious. Physically, as was seen with our patients, the overlying skin can appear normal and there may be a pulsatile mass, a thrill, increased warmth and redness; other possible features include pain, buzzing sound, overgrowth, haemorrhage and heart failure. Arteriovenous malformations often remain undiagnosed until dramatic bleeding occurs as a result of dental manipulation.\(^\text{22}\) In Case 30, the patient had congenital left facial swelling but became troubled by lower jaw gum bleeding shortly after the onset of puberty; he sought medical attention on the advice of his dental surgeon. Categorised clinically by the Schobinger classification, 28% of patients were in stage I (cutaneous blush or warmth), 50% were stage II (bruin, audible pulsations, expanding lesion), 22% were stage III (pain, ulceration, bleeding, infection) and none were stage IV (cardiac failure).
The high-flow nature of the lesion can be confirmed through Doppler examination. On MRI, the anomaly is characterised by enlarged vascular channels with dilated feeding and draining vessels; there is no discrete soft tissue mass. Abnormal arteriovenous connections are easily recognisable as linear or punctuate signal voids or as hyperintensities. Magnetic resonance angiography used in Cases 28 and 37 confirmed the high-flow nature of the lesion and mapped out the feeding and draining vasculature.

Angiography, performed in 43% of our case series, is usually required to evaluate the abnormality in more detail before surgical excision or at embolisation; it shows a high degree of shunting with high blood flow though the lesion and multiple feeders are usually seen. Angiography (Table 4) revealed involvement of the facial artery (feeding vessel) and facial vein in all these cases. Angiography in Case 30 demonstrated, in addition to the left mandibular arteriovenous malformation, hypothalamic and left optic pathway arteriovenous malformations consistent with features of the Wymburn-Mason syndrome.

The arterial and venous phases are present in the same angiographic image if there is significant shunting. Microshunts not demonstrated on angiography open up after main feeding vessel ligation, incomplete embolisation or subtotal surgical resection leading clinically to recurrence of the arteriovenous malformation often larger than the primary lesion. Therefore, selective embolisation on its own, subtotal resection, or proximal ligation of vessels are rarely successful procedures with high-flow anomalies because of the establishment of new flow pathways.14,21,23

The recurrences in Cases 32, 37, 39 were consequent on incomplete first resection, and the recurrence in Case 35 resulted from both inadequate initial resection and the subsequent suboptimal use of embolisation alone 6 years later. All recurrent lesions were notably larger than the primary counterparts. Authors advise careful assessment and combined treatment consisting of good highly selective embolisation followed by total resection ideally within 48 hours for most symptomatic arteriovenous malformations because of the establishment of new flow pathways.14,21,23

Compared to haemangiomas and venous malformations, arteriovenous malformations are rare in the head and neck, and the experience of most surgeons is limited. Kohout et al11 recommended combined embolisation and resection for early (stage I and II) lesions with the aim of preventing progression to more complicated lesions, while painful or rapidly enlarging lesions warrant early intervention owing to a high likelihood of progression and risk of serious haemorrhage. In our series, combined treatment was practised in only 6 patients; the other 8 were subjected to excision alone with no recurrence at a mean follow-up of 2 years. Typically, a residual incompletely resected arteriovenous malformation re-expands 1 to 2 years postoperatively but may remain quiescent for decades.24

Certainly resection alone has a place in the management of small discrete non-expanding lesions if the surgeon is confident of obtaining clear resection margins.

Of the 14 arteriovenous malformations, 6 (Cases 28, 30, 32, 34, 37, 41) were subjected to preoperative superselective embolisation (Table 4). There are 2 forms of embolisation.24

Primary embolisation aims to deliver an ablative embolic agent to the nidus of the arteriovenous malformation to facilitate endothelial destruction – this may have a role in patients who are not ready for mutilating surgery. Particle embolisation, which was practised in this series, uses particles such as polyvinyl alcohol foam (PVA) or acrylic microsheres to occlude flow into the nidus achieving preoperative devascularisation, thus minimising intraoperative haemorrhage and providing a dry operative field. The particles come in different sizes and it is important to choose the optimum size: oversized particles cause physiological proximal ligation and suboptimal preoperative devascularisation whereas undersized particles do not serve their occlusive purpose and may occlude the subdermal plexus leading to skin necrosis. Preoperative embolisation is not without its risks. It has the potential to cause stroke, cranial nerve ischaemia, skin necrosis, bleeding, blindness, adverse haemodynamic changes and even possible pulmonary embolism.4

Some larger arteriovenous malformations have feeders arising from the internal carotid artery (Cases 30 and 32) and in these situations embolisation may not be possible such that significant and even lethal bleeding may occur at resection.20 In such cases, the patient should be informed of the high risk of resection and if surgery proceeds cardiopulmonary bypass may be considered, or the compartmentalisation technique may be used in the event of massive bleeding. Case 30 required emergency surgical haemostasis and excision via a hemimandibulectomy owing to extensive post-embolisation blowout bleeding from the arteriovenous malformation. This was not totally unexpected looking at its rich feeder vascularity originating from both internal and external carotid systems. The patient declined mandibular reconstruction and was left with an acceptable jawline.

More sophisticated reconstructive solutions were utilised in this category of anomalies. Of note, Case 32 (Fig. 1) had a traumatic forehead arteriovenous malformation that was excised only to recur 2 years later. Preoperative embolisation was performed on the recurrent malformation followed by resection and split thickness skin grafting to cover the defect. Scalp tissue expansion was used to advance his disrupted hairline but this was complicated by infection of the tissue expander that had to be removed. Tissue expansion
Fig.1. (Above, left) A 21-year-old man (Case 32) with a traumatic forehead enlarging arteriovenous malformation that was excised. (Above, centre) Recurrence 2 years after the first operation. Note the old scar. (Above, right) Angiogram at preoperative embolisation showing the embolised feeding vessels. (Below, left) Resected specimen of the recurrent lesion. (Below, centre) Result – 17 months after preoperative embolisation with aesthetic unit resection of the recurrence, showing a disrupted anterior hairline post split skin grafting of the defect. (Below, right) Tissue expander inserted to facilitate hairline advancement.

Fig.2. (Above, left) A 37-year-old man (Case 37) with an upper lip gradually enlarging arteriovenous malformation. (Above, centre) Coronal magnetic resonance imaging (MRI) showing the vessels. (Above, right) Angiogram of the lesion done showing preoperative embolisation of the facial artery. (Below, left) The left radial forearm free flap donor site 4 days postoperatively. (Below, centre and right) Result – 3 months after resection with reconstruction.

Fig.3. (Above, left and centre) A 45-year-old man (Case 41) with a right cheek enlarging arteriovenous malformation. (Above, right) Angiogram at preoperative embolisation showing the embolised feeding vessels. (Below, left) Resected specimen of the lesion. (Below, centre and right) Result – 3 months after resection and immediate radial forearm flap reconstruction.
is particularly suited for scalp reconstruction here where resection of the vascular anomaly disrupts the hairline. In Case 37 (Fig. 2) the patient suffered from a persistent upper lip arteriovenous malformation that had previously been subjected to subtotal resection; he also had laser treatment and sclerotherapy – modalities, along with steroid and irradiation, found to be ineffective for these anomalies. His malformation was finally subdued with radical en bloc resection and reconstruction with a left radial forearm free flap anastomosing the radial artery to the facial artery and the radial venae comitantes to the facial veins, as well as anastomosing lateral cutaneous forearm nerve to the cervical cutaneous branch. At 8-year follow-up there was no recurrence and the cosmetic and functional result was very satisfactory to both the patient and the surgeon. The patient in Case 41 had a post-excision right cheek defect reconstructed using the versatile radial forearm flap once again fulfilling form and function (Fig. 3). As practised in Cases 16, 37 and 41, the coverage of resulting defects with myocutaneous flaps serves as a haemostypt and the transfer of healthy tissue also helps create normal micro-revascularisation, hence preventing recurrence. Adequacy of malformation resection is best judged intraoperatively from the bleeding quality and with Doppler assessment and frozen section, as well as clinically at follow-up and again with Doppler.

This single-institution study explored a dedicated series of surgically treated cervicofacial vascular anomalies in terms of clinical characteristics, diagnosis, treatment and its outcome. Accurate diagnosis to distinguish between cervicofacial haemangiomas and vascular malformations is key to optimum management (Fig. 4). The diagnosis can be made in the majority by history and physical examination. Cervicofacial vascular anomalies were most commonly found at the lip and atypical-looking vascular anomalies were often misdiagnosed. In questionable cases diagnosis should be aided at an early age with MRI, so that best treatment may be chosen before the lesion progresses further. Although haemangiomas are expected to involute spontaneously and effective medical therapy is available in cases threatening function or life, surgical treatment has shed its traditional “last resort” image and is now clearly indicated from a young age for preventing psychological distress, in cases of chronic aesthetic alteration resulting from partial regression of haemangiomas, and where medical therapy has failed. While low-flow malformations usually
cause little morbidity, they do not involute and aesthetic concern and prevention of psychosocial distress points to early excision as the treatment of choice. Lymphatic malformations are best treated by excision. Outcome after surgical excision of localised cervicofacial haemangiomas and low-flow vascular malformations is generally excellent, with low recurrence rate and minimum morbidity. Large extensive low-flow malformations as well as those located at the lips often require multiple procedures including reconstruction; the outcome is generally not as good and the patient must be made aware in these cases. Except for small localised lesions which can be directly excised, combined preoperative embolisation followed by complete excision within 48 hours is the treatment of choice for arteriovenous malformations. Myocutaneous flap reconstruction may prevent the recurrence of arteriovenous malformations. Tissue expansion is a useful technique for reconstruction after the excision of large vascular anomalies, particularly in the scalp and forehead.

REFERENCES