

An unusual submandibular tumour

A woman in her 60s presented with a non-tender, non-enlarging swelling in the left submandibular triangle of the neck for 3 months. She had no significant past medical or surgical history. Examination showed a 2cm firm round lump in the left submandibular triangle, not attached to the skin, mandible and not palpable in the floor of the mouth bimanually. The rest of the head and neck examination including flexible nasolaryngoscopy was normal. Computed tomography (CT) of the neck showed a lobulated, well-circumscribed, heterogeneously enhancing mass in the left submandibular triangle. The mass was caudal to and abutting the left mylohyoid muscle; it also abutted the anterior belly of the left digastric muscle (Fig. 1). An ipsilateral level 1B (submandibular) lymph node showed asymmetric focal cortical thickening, suspicious for metastasis. Fine needle aspiration cytology of the mass in the left submandibular triangle showed salivary gland neoplasm of uncertain malignant potential, suggestive of pleomorphic adenoma. However, CT showed no radiological abnormality in the submandibular or parotid glands bilaterally.

What is your diagnosis?

- Metastatic pleomorphic adenoma to a submandibular lymph node
- Metastatic salivary gland carcinoma of unknown primary
- Sublingual gland carcinoma invading through the floor of mouth
- Sublingual gland tumour herniating through a mylohyoid boutonnière
- Chondroid syringoma

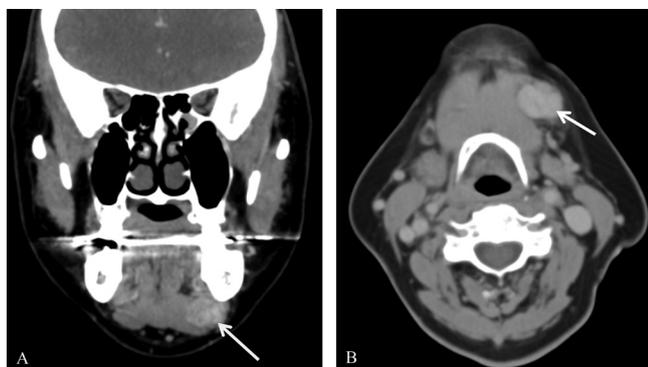


Fig. 1. Contrast-enhanced computed tomography of the neck in (A) coronal view and (B) axial view (2 arrows indicate the tumour).

Findings and diagnosis. Neck CT showed a focal defect in the left mylohyoid muscle, and the left sublingual gland was not seen in the sublingual space. The clinical diagnosis of a sublingual tumour herniating through a mylohyoid boutonnière was made, and the possibility of metastatic pleomorphic adenoma from the adjacent left submandibular gland was considered. Hence, the patient underwent excision of the left submandibular tumour, level 1A and left level 1B neck dissection via a transcervical approach.

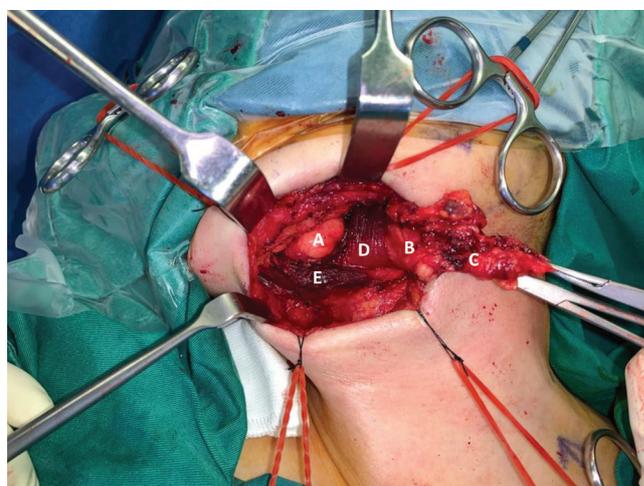


Fig. 2. Intraoperative image.

A: pleomorphic adenoma of the sublingual gland; B: submandibular gland; C: level 1 lymph nodes; D: mylohyoid muscle; E: anterior belly of the digastric muscle

Intraoperatively, the tumour was arising from the left sublingual gland and herniated through a mylohyoid boutonnière (Fig. 2). Following the principle of extracapsular dissection, we resected the tumour with a cuff of grossly normal sublingual gland attached to it superiorly. This was performed under 2.5x magnification using surgical loupes and we did not encounter a grossly visible branch of the lingual or the hypoglossal nerve in the vicinity of the tumour. Intraoperative frozen section of this tumour was suggestive of pleomorphic adenoma, and formalin-fixed paraffin-embedded histology confirmed salivary gland tissue containing a pleomorphic adenoma. The surgical margins were clear although the closest margin measured less than 1mm. Histology of the level 1 lymph nodes showed reactive changes and

Answer: D

the submandibular gland was normal. The patient recovered uneventfully except for a transient marginal mandibular palsy. Lingual sensation to cold touch was symmetrical postoperatively, suggesting normal function of the lingual nerve. Tongue protrusion was midline, suggesting normal function of the hypoglossal nerve. Patient was advised on a yearly follow-up to monitor for recurrence of the pleomorphic adenoma.

Discussion. A submandibular swelling can possibly arise from the submandibular gland, cervical lymph node or the sublingual gland. The submandibular gland has a superficial lobe that lies in the neck inferior to the mylohyoid muscle, and a deep lobe that hooks around the posterior margin of the mylohyoid to enter the floor of mouth. This makes ballotability a sign of a submandibular tumour. The sublingual gland sits superior to the mylohyoid and under the mucosa of the floor of the mouth. Thus, tumours of the sublingual gland typically present as a lump under the tongue instead of in the neck. Cervical lymphadenopathy is the usual clinical diagnosis when a submandibular swelling is not ballotable between the floor of the mouth and the neck. However, cytology from the mass in this patient suggested a salivary gland tumour. This made the diagnosis of a submandibular gland tumour the most likely, yet CT showed a normal submandibular gland. This conundrum raised the possibility of a nodal metastasis from an occult primary (Option B) in the major or minor salivary glands of the head and neck. Thus, a complete mucosal examination including nasolaryngoscopy was also performed. However, unknown primary carcinomas of salivary gland origin and metastatic pleomorphic adenoma (Option A)—possible diagnoses given the cytology—are very rare.¹

Following this, a tumour of the sublingual gland invading or herniating through the mylohyoid muscle (Options C and D) becomes plausible. As the majority of sublingual tumours are malignant, a sublingual carcinoma invading through the mylohyoid should be carefully considered. However, the well-circumscribed margin of this tumour on the CT indicates that gross invasion of the mylohyoid is unlikely. Despite this, a low-grade sublingual carcinoma is a differential that requires histology to rule out.

Chondroid syringoma (Option E), also known as pleomorphic adenoma of the skin, is a rare tumour arising from the eccrine or apocrine sweat glands. It typically presents as a solitary asymptomatic swelling that histologically resembles pleomorphic adenoma, but is confined within the cutaneous or subcutaneous tissue.

Mylohyoid boutonnières (buttonholes), ranging from 5mm to 2cm in size, are found in up to 77% of individuals and are usually clinically insignificant.² They are usually seen along the lateral margin of the mylohyoid muscle closer to the mandible than to the midline raphe.^{2,3} In many cases, the anterior belly of the digastric muscle was found totally or partially covering the herniation.³ Given this knowledge, a careful bimanual palpation of a mass in the anterior submandibular or submental triangles may confirm its connection with sublingual glands through a boutonnière. That the tumour was not palpable in the floor of mouth of our patient could be because we did not push up the tumour adequately during bimanual palpation. Overall, a sublingual gland tumour rarely presents exclusively as a neck mass. We found no report of such cases on PubMed. A more commonly reported entity is hypertrophied sublingual gland herniating through a mylohyoid boutonnière. This entity appears to be associated with hypoplasia of the submandibular glands.⁴

Soft-tissue CT of the neck can distinguish between submandibular gland tumours, sublingual hypertrophy and sublingual gland tumours. Performing a Valsalva manoeuvre during sonography has also been suggested to aid the diagnosis of herniating sublingual glands and mylohyoid boutonnière.⁵ Awareness of mylohyoid boutonnière is important for a clinical diagnosis of a sublingual tumour herniating into the neck. Surgeons should be aware of a herniating sublingual tumour when faced with a salivary gland tumour that is palpable in the submandibular or submental region, but not arising from the submandibular gland. Transcervical resection was able to achieve a clear pathological margin in this patient.

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