Pancreatic Haemangioma: An Unusual Case of Massive Upper Gastrointestinal Bleeding with Clinical and Radiological Correlation of the Literature and Recommendations

Dear Editor,

Pancreatic haemangiomas in adults are exceedingly rare. Most cases reported have been diagnosed on postoperative histology, underlining the difficulty in preoperative diagnosis. We report the first adult patient with undiagnosed pancreatic haemangioma, presenting with massive upper gastrointestinal (GI) tract bleeding, necessitating an emergency Whipple's procedure to arrest the bleeding.

Case Report

A 62-year-old lady had presented to the Singapore General Hospital emergency department with haematemesis, haematochezia and sudden onset abdominal pain. She had previous history of malignant thymoma in remission, myasthenia gravis and connective tissue disease on oral steroids and immunosuppressants (mycophenolate mofetil).

On arrival in the emergency department, she was in hypovolaemic shock, with hypotension and tachycardia. Aggressive resuscitation with fluids and blood products was initiated. Examination found mild epigastric tenderness but no palpable mass. She was brought to the operating theatre for an emergency oesophagogastroduodenoscopy to attempt endoscopic haemostasis. Initial resuscitative efforts included a total of 9 units of packed red blood cells via a rapid transfuser.

The bleeding was localised to the third segment of the duodenum. However, the torrential bleeding made endoscopic haemostasis impossible. A laparotomy was thus performed for haemostasis. A 3 cm x 3 cm firm head of pancreas mass was found, with erosion into the third segment of the duodenum (Fig. 1). Active spurting from the erosion was seen after duodenotomy was performed. The bleeding point was controlled with a Prolene 2/0 stitch, and the decision was made to resect the mass as it was mobile, and free of adjacent structures, including the superior mesenteric artery and vein.

The final histology reports a pancreatic haemangioma, 4.5 cm x 5.5 cm, with focal erosion into overlying duodenum. Immunohistochemistry revealed CD31, CD34 and ERG-positive, supporting the diagnosis of a haemangioma.

She is currently 1-year postoperation and has recovered well.

Discussion

Vascular tumours of the pancreas (including haemangiomas, lymphangiomas, haemolymphangiomas, haemangioendothelioma, haemangiopericytoma, haemangioblastoma and angiosarcomas) are rare, and account for only 0.1% of pancreatic tumours.[1] Pancreatic haemangiomas as a subset are thus exceedingly rare. Pancreatic haemangiomas are more common in the paediatric age group, but these do not persist into adulthood (instead undergo involution and regress over several years).[2]

A total of 22 case reports describe pancreatic haemangioma in adults; the earliest by Ranstrom in 1939 (Table 1).[3-10] The most common presenting symptom of these reported cases was abdominal pain (accounting for more than half of these cases). This is the first reported case of pancreatic haemangioma presenting with massive bleeding, and requiring an emergency Whipple's procedure.

Although Ringoir et al described a case in 1961 who presented with bleeding,[4] the blood loss was not torrential as in our case. Their case had presented with 2 episodes of coffee grounds vomiting and melaena, and was haemodynamically stable on arrival. They reported a 15 cm pancreatic haemangioma, which we expect to have

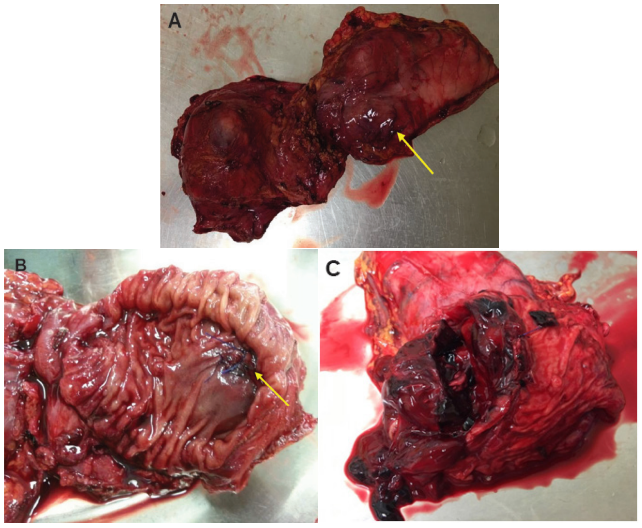

Fig. 1. A) Whipple's specimen showing mass at head of pancreas. B) Stitch haemostasis via duodenotomy, over area of erosion. C) Pancreatic lesion cut open, revealing clots and blood within.

Table 1. Summary of Previously Described Cases* of Pancreatic Haemangioma

No.	Author	Country	Year	Age/Gender	Site	Size (cm)	Presentation	Treatment
1	Ranstrom	-	1939	61/F	Head	7	Autopsy	NA
2	Derom	France	1960	-		-	Unknown	Surgery
3	Ringoir	France	1961	71/F	Head	15	Haemetemesis/ melaena	Gastroenterostomy and vagotomy
4	Colardyn	France	1972	42/F	Body		Abdominal pain	Fat-free diet and anticholinergics
5	Mangin	France	1985	62/F	Head to tail	20	Malaise, nausea, thrombocytopaenia	Laparotomy and observation
6	Kobayashi	Japan	1991	30/M	Head	20	Abdominal distension	Pancreatico-duodenectomy
7	Dageforde	Germany	1991	79/F	Body to tail	6	Abdominal pain	Observation
8	Chang	Taiwan	2003	70/F	Body to tail	4	Abdominal pain	Subtotal pancreatectomy
9	Plank	Austria	2006	36/M	Head	3	Abdominal pain	Laparotomy and observation
10	Xu	China	2008				3 cases	
11	Mundinger	United States	2009	45/F	Head	5.5	Abdominal pain	Pylorus preserving pancreatico-duodenectomy
12	Jarboui	Tunisia	2010	60/F	Body	2	Abdominal pain	Distal pancreatectomy
13	Weidenfeld	Israel	2011	73/F	Head	5	Abdominal pain	Pancreatico-duodenectomy
14	Lee	Malaysia	2011	49/F	Neck	5	Incidental US finding, non-specific dizziness	Central partial pancreatectomy and gastrostomy
15	Kersting	Germany	2012	53/M	Head	8	Asymptomatic	Extirpation of tumour
16	Zhi-hua	China	2013	23/F	Head	5.4	Incidental US finding	Subtotal pancreatectomy
17	Malik	United Kingdom	2013	70/F	Head	8	Abdominal pain	Pylorus preserving pancreatico-duodenectomy
18	Williamson	United Kingdom	2014	78/F	Head	4	Abdominal pain	Observation
19	Naito	Japan	2014	40/F	Body to tail	10	Abdominal pain	Pancreatectomy
20	Mondal	United States	2015	18/F	Head	6	Abdominal pain	Pylorus preserving pancreatico-duodenectomy
21	Liu	China	2015	28/F	Body to tail	8.8	Abdominal pain	Distal pancreatectomy
22	Kim	South Korea	2016	68/F	Tail	0.5	Incidental CT finding	Distal pancreatectomy

CT: Computed tomography; F: Female; M: Male; NA: Not applicable; US: Ultrasound

*Derom F, Ringoir S, Marlier R. [Two cases of intraabdominal hemangioma: liver and pancreas]. Acta Chir Belg 1960;59:172-82; Ringoir S, Derom F, Colle R, Mortier G. Hemangioma of the pancreas. Report of a case. Gastroenterology 1961;41:43-5; Kobayashi H, Itoh T, Murata R, Tanabe M. Pancreatic cavernous hemangioma: CT, MRI, US and angiography characteristics. Gastrointest Radiol 1991;16:307-10; Plank C, Niederle B, Ba-Ssalamah A, Schima W. Pancreatic hemangioma: imaging features with contrast-enhanced CT and with gadolinium- and mangafodipir-enhanced MRI. Eur J Radiol 2006;57:59-62; Williamson JM, Finch-Jones M, Pope I. Endoscopic ultrasonography allowing expectant management of pancreatic haemangioma. Ann R Coll Surg Engl 2014;96:e1-2; Mondal U, Henkes N, Henkes D, Rosenkranz L. Cavernous hemangioma of adult pancreas: a case report and literature review. World J Gastroenterol 2015;21:9793-802; Lu T, Yang C. Rare case of adult pancreatic hemangioma and review of the literature. World J Gastroenterol 2015;21:9228-32; Kim SH, Kim JY, Choi JY, Choi YD, Kim KS. Incidental detection of pancreatic hemangioma mimicking a metastatic tumor of renal cell carcinoma. Korean J Hepatobiliary Pancreat Surg 2016;20:93-6.

massive bleeding, if ruptured. This leads us to speculate whether the bleeding in Ringoir's case arose from erosions in a narrowed duodenum, which led to incidental discovery of an asymptomatic mass.

The risk of rupture and consequent morbidity and mortality of pancreatic haemangiomas are difficult to delineate, as they are exceedingly rare. As such, references to hepatic haemangiomas have frequently been made. Rates of spontaneous hepatic haemangioma ruptures stand at 1%-4%, mainly in giant haemangiomas larger than 6 cm.[11] The average size in cases presenting with rupture was about 11 cm, with a very high mortality rate of 35%.[12]

The use of steroids has also been shown to increase the size of haemangiomas.[12] We postulate that the steroid

treatment for our patient's connective tissue disease could have caused an enlarging haemangioma. Correlating with the data from hepatic haemangiomas, she therefore faced an increased risk of rupture with potential mortality.

Also, since the lesion is located in the head of the pancreas and abutting the duodenum, endoluminal foreign body injury and erosions into the haemangioma could have also resulted in the rupture.

Our patient was at risk of death from exsanguination from a ruptured pancreatic haemangioma. This prompted us to perform a retrospective review of our patient's computed tomography (CT) scans that had been previously done for follow-up of her malignant thymoma.

In view of the clinical diagnostic difficulty faced in characterising this pancreatic haemangioma, we retrospectively reviewed the patient's earlier scans to determine if the pancreatic haemangioma could have been definitively diagnosed earlier. On review, the pancreatic lesion was already present in 2011, then measuring almost 35 mm (Fig. 2A). The lesion was isodense in relation to the pancreas on subsequent follow-up scans, making it difficult to perceive. A tri-phasic CT scan that was performed in 2013 demonstrated draping of vessels around the isodense mass (Fig. 2B). A later CT scan performed in 2016 showed a mild increase in size to 44 mm (Fig. 2C). We postulate that it may be the slight difference in timing of the single-phase portal venous study in 2011 that rendered the mass slightly more conspicuous compared to subsequent single-phase studies.

The difficulty in diagnosis of pancreatic haemangiomas is not unique to our case. A variety of imaging modalities have been previously applied. Trans-abdominal ultrasound seems effective for larger lesions, having clinched the diagnosis in 9 of the cases, albeit all larger than 5 cm in size. One case described the use of intraoperative ultrasound for diagnosis of a smaller 4 cm haemangioma.[6] The use of endoscopic ultrasound was later used in 3 cases, but only one correctly diagnosed pancreas haemangioma.[7] Contrast-enhanced CT was performed in 12 of the cases, with 6 showing poor arterial enhancement, and the other 6 showing hyper-enhancement. Four of the reported cases underwent magnetic resonance imaging (MRI), but only 1 case by Kobayashi[5] showed classical hypo-intensity in T1-weighted images and moderate hyper-intensity signal in T2-weighted images with marked enhancement post-gadolinium. Overall, there seems to be no superior modality in the diagnosis of pancreatic haemangiomas.

In our case, on CT alone, it was difficult to definitively diagnose pancreatic haemangioma. Had the lesion been seen, perhaps further evaluation with MRI may have aided its diagnosis.

While the general consensus for management of haemangiomas is conservative in view of its benign nature, we now present this rare case with a life-threatening massive bleeding from a pancreatic haemangioma. This might open doors to consideration of surgical resection for cases deemed to be at increased risk of rupture.

Conclusion

Haemangiomas are rare lesions of the pancreas. Diagnosis with imaging remains a challenge, and a high index

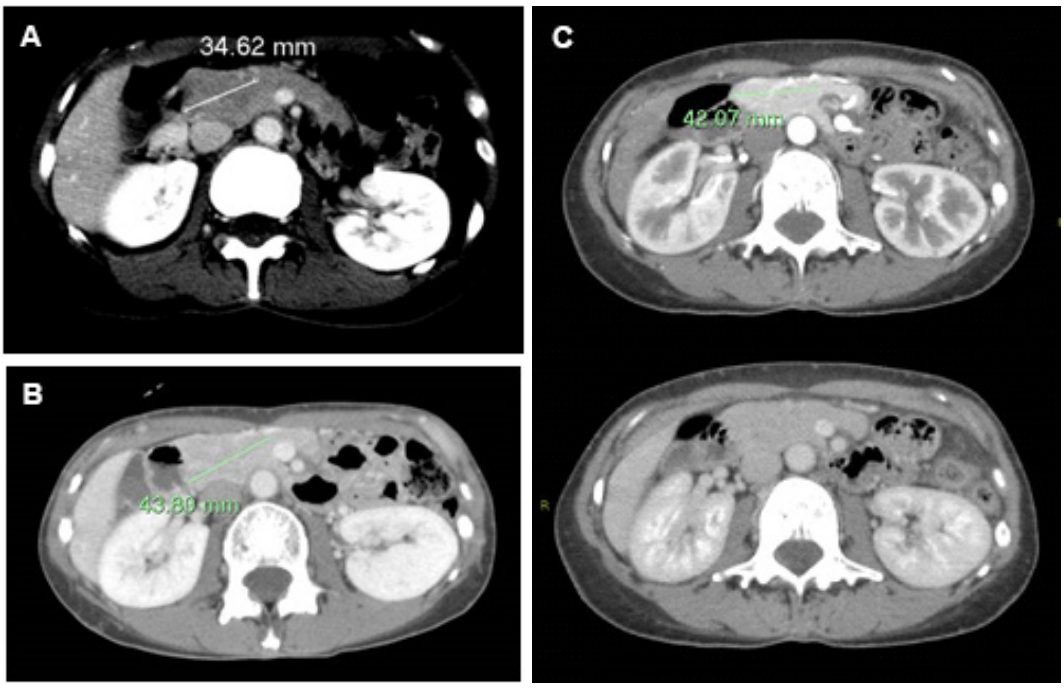

Fig. 2. Computed tomography (CT) scans of our patient. A) Scan in 2011 showing lesion in head of pancreas about 35 mm in size. B) Arterial and delayed phase scan in 2013, showing draping of vessels around isodense lesion in head of pancreas. C) Latest scan in 2016, again showing isodense lesion about 44 mm in size.

of suspicion is needed. We described the first ruptured pancreatic haemangioma presenting with massive bleeding into the GI tract, for which an emergency Whipple's procedure was performed. We urge the consideration of surgical management of pancreatic haemangiomas, if deemed at high risk of rupture.

REFERENCES

1. Le Borgne J, de Calan L, Partensky C. Cystadenomas and cystadenocarcinomas of the pancreas: a multiinstitutional retrospective study of 398 cases. Ann Surg 1999;230:152-61.
2. Takahashi K, Mulliken JB, Kozakewich HP, Rogers RA, Folkman J, Ezekowitz RA. Cellular markers that distinguish the phases of hemangioma during infancy and childhood. J Clin Invest 1994;93:2357-64.
3. Derom F, Ringoir S, Marlier R. [Two cases of intraabdominal hemangioma: liver and pancreas]. Acta Chir Belg 1960;59:172-82.
4. Ringoir S, Derom F, Colle R, Mortier G. Hemangioma of the pancreas. Report of a case. Gastroenterology 1961;41:43-5.
5. Kobayashi H, Itoh T, Murata R, Tanabe M. Pancreatic cavernous hemangioma: CT, MRI, US and angiography characteristics. Gastrointest Radiol 1991;16:307-10.
6. Plank C, Niederle B, Ba-Ssalamah A, Schima W. Pancreatic hemangioma: imaging features with contrast-enhanced CT and with gadolinium- and mangafodipir-enhanced MRI. Eur J Radiol 2006;57:59-62.
7. Williamson JM, Finch-Jones M, Pope I. Endoscopic ultrasonography allowing expectant management of pancreatic haemangioma. Ann R Coll Surg Engl 2014;96:e1-2.
8. Mondal U, Henkes N, Henkes D, Rosenkranz L. Cavernous hemangioma of adult pancreas: a case report and literature review. World J Gastroenterol 2015;21:9793-802.
9. Lu T, Yang C. Rare case of adult pancreatic hemangioma and review of the literature. World J Gastroenterol 2015;21:9228-32.
10. Kim SH, Kim JY, Choi JY, Choi YD, Kim KS. Incidental detection of pancreatic hemangioma mimicking a metastatic tumor of renal cell carcinoma. Korean J Hepatobiliary Pancreat Surg 2016;20:93-6.
11. Chen ZY, Qi QH, Dong ZL. Etiology and management of hemmorrhage in spontaneous liver rupture: a report of 70 cases. World J Gastroenterol 2002;8:1063-6.
12. Donati M, Stavrou GA, Donati A, Oldhafer KJ. The risk of spontaneous rupture of liver hemangiomas: a critical review of the literature. J Hepatobiliary Pancreat Sci 2011;18:797-805.

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