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Diagnostic challenges of intraabdominal schwannoma in victorian tertiary centre

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Abstract

A 62-year-old lady with a known segment 8 hepatic hemangioma was referred to our general surgical unit with an incidental CT finding of new rounded soft tissue density lesion either in the segment 3 of the liver or in the anterior wall of the stomach, measuring 21 x 21 x 27 mm. Based on the CT appearance, the lesion was initially thought to be a gastrointestinal stromal tumor. A further endoscopic ultrasound elucidated an oval-shaped, hypoechoic submucosal lesion in the body of the stomach. The FNA performed during the endoscopy was nondiagnostic. The patient subsequently underwent a laparoscopic transgastric resection of the lesion. The lesion was completely excised with an uneventful perioperative course. The histopathology revealed a completely excised 25 x 20 x 17mm well-circumscribed, but unencapsulated nodular spindle cell tumor, consistent with a gastric schwannoma.

A 48-year-old lady presented with a several month history of constipation and bloating on a background of hypertension and obstructive sleep apnea. Initial abdominal ultrasound revealed a large hypoechoic mass in the head of the pancreas. Further CT revealed a 51 x 37 x 37mm, hypodense, ovoid mass between the head of the pancreas and portal vein ventrally. The lesion was nonenhancing, and hypodense relative to the surrounding organs, including the liver and the pancreas. Subsequent endoscopic ultrasound revealed a well-circumscribed, solid mass lesion, measuring 48mm x 42mm in maximal cross-sectional diameter. It was abutting the first part of the duodenum, but was clearly separate from it and the other surrounding organs. The FNA was performed under ultrasound guidance. The histopathology revealed a mixture of cytologically bland spindle cells of intermediate size and foamy macrophages, together with small numbers of infiltrating lymphocytes. The appearance was consistent with a retroperitoneal schwannoma. The patient opted to be monitored on serial imaging.

both cases, the histological specimens were further examined with immunohistochemical testing. Both schwannomas were strongly immunopositive for S100 protein, but were negative for c-kit, DOG-1 and desmin.

Keywords

intraabdominal schwannoma; gastrointestinal stromal tumor; diagnostic challenge; computer tomography; endoscopic ultrasound; immunohistochemistry nazole

Abbreviations

CT: Computer tomography; FNA: Fine needle aspiration; GIST: Gastrointestinal stromal tumor

Introduction

Schwannoma is a slow-growing benign tumor that arises from nerve sheath composed of Schwann cells, which produce the insulating myelin sheath of peripheral nerves. The most frequently affected sites in the body include the flexor aspects of extremities, head and neck regions. Other sites are rarely affected, and of them all, the intraabdominal schwannomas are by far the most uncommon form of schwannomas.

Intraabdominal schwannomas are most commonly diagnosed as incidental findings or investigated for due to their mass effect. They usually present as painless lumps that gradually increase in size over a period of months to years without any constitutional symptoms, however, they can sometime present with vague abdominal or back pain [1]. Intraabdominal schwannomas are usually non tender, well demarcated and non-tethering as they are completely benign lesions. They have also been shown to demonstrate unique histological and immunohistochemical features from their soft tissue and central nervous system counterparts [2,3].

However, owing to their rarity, there is only limited experience and understanding of intraabdominal schwannomas across the globe. We herein present 2 cases of intraabdominal schwannoma at a large tertiary referral centre. To the best of our knowledge, this is the first published case report of such tumors from a Victorian tertiary referral centre.

Case Report 1

A 62-year-old lady with past medical history of a known large hepatic hemangioma in segment 8 under surveillance, hysterectomy and previous trauma laparotomy for duodenal injury from a motor vehicle accident, was referred to us for opinion and management of an incidental gastric wall mass on CT performed as an investigation for deranged liver function test.

The CT demonstrated stable 96mm hemangioma in segment 8, but a new rounded soft tissue density lesion either in segment 3 of the liver or from the anterior wall of the stomach measuring 21 x 21 x 27 mm was revealed. The lesion was more in keeping with a submucosal gastric wall mass rather than an exophytic hepatic lesion. The patient had no gastrointestinal symptoms such as abdominal pain, nausea, vomiting, dysphagia, hematemesis or melena. She did not report any weight loss nor had any stigmata of chronic liver disease.

She subsequently underwent a gastroscopy, which revealed a medium-sized, submucosal, non-circumferential mass found on the anterior wall of the gastric body. The mass was measuring 25mm in maximal diameter and did not show any evidence of active or previous bleeding. Its benign appearance was most consistent with a gastrointestinal stromal tumor (GIST).

A further endoscopic ultrasound was performed, which further elucidated an oval, intramural, subepithelial lesion in the body of the stomach. The lesion was hypoechoic and heterogeneous. Sonographically, the lesion appeared to originate from the muscularis propria. The lesion was measuring 17mm in maximal thickness, and 25mm in maximal diameter. The outer endosonographic borders were

well defined. Fine needle aspiration (FNA) was performed. Colour Doppler imaging was utilized prior to needle puncture to confirm a lack of significant vascular structures within the needle path. Three passes were made with the 19 gauge needle and with the 25 gauge needle using a transgastric approach. The appearance was, once again, consistent with a 25mm GIST. Unfortunately, the FNA was nondiagnostic.

She subsequently underwent a laparoscopic transgastric resection of stomach. The tumor was lying in the anterior body of the stomach. Postoperative course was uneventful. The histopathology was reported as gastric schwannoma which was completely excised with dimensions of 25 x 20 x 17mm. Sections showed a circumscribed but unencapsulated nodular spindle cell tumor, consistent with a schwannoma, expanding the muscularis propria. No malignant features were identified. The lesion was completely excised in planes of section. Immunostains were positive for S100 and SOX-10, but negative for c-kit, desmin, and DOG-1. These findings all confirmed the diagnosis of gastric schwannoma.

Case Report 2

A 48-year-old lady, from a Russian ethnic background, presented with a several month history of constipation and bloating on a background of hypertension and obstructive sleep apnea. Abdominal ultrasound by the primary care physician demonstrated a large hypoechoic mass in the head of the pancreas.

Further CT revealed a 51 x 37 x 37mm, hypodense, ovoid mass between the head of the pancreas and portal vein ventrally, IVC and diaphragmatic crus dorsally, liver laterally, and the celiac artery medially. The lesion was hypodense relative to the liver and the pancreas. No calcification or hyperenhancement were noted on the arterial phase, however, on portal venous phase, the mass demonstrated slight enhancement though it remained hypodense relative to the adjacent organs. The duct of the pancreas was not dilated, and therefore, the lesion was thought to be less likely to be within the pancreatic head but abutting the head. Both the IVC and portal vein appeared mildly compressed due to the mass effect by the lesion. The pancreas itself demonstrated a homogeneous parenchymal enhancement and did not show any calcifications or peripancreatic inflammation.

The patient did not report any abdominal pain, back pain, nausea, vomiting, jaundice or weight loss. Clinical examination was unremarkable. CA 19.9, chromogranin A, LDH, B2 microglobulin, and CEA were normal. There were no clinical symptoms suggestive of lymphoma such as fever, night-sweats, lethargy or weight loss. There was no clinical evidence of cervical, axillary or inguinal lymphadenopathy.

Subsequent endoscopic ultrasound revealed one solid mass lesion in the celiac region, just to the right of the celiac axis with the ultrasound probe 42cm from the incisors. It measured 48mm x 42mm in maximal cross-sectional diameter. It was also clearly adjacent to and abutting the first part of the duodenum. There was no vascular invasion noted. It was clearly separate from the pancreas and liver. The node was irregular and of mixed hypoechoic and isoechoic echogenicity, and had well defined margins. FNA was performed under ultrasound guidance. The tissue from FNA was composed of a mixture of cytologically bland spindle cells of intermediate size and foamy macrophages, together with small numbers of infiltrating lymphocytes. It was positive for S100 and negative for c-kit, DOG-1 and desmin. This was consistent with a retroperitoneal schwannoma.

The decision was made to have the lesion under surveillance with serial imaging given largely

asymptomatic patient. Repeat CT 3 month later showed retroperitoneal schwannoma stable in size without any evidence of new lesions.

Discussion

Schwannomas usually have proclivity to affect the flexor surface of extremities, head and neck regions. However, their occurrence in the intraabdominal region is much rarer. In the study by Das Gupta et al, intraabdominal schwannoma only comprised of 2% of all benign schwannomas[4], and in the report of Goh et al, only 5.6% of mesenchymal tumors were intraabdominal schwannomas [5]. The intraabdmoinial schwannomas are most frequently found in the gastrointestinal tract, where the stomach is the most frequently affected site [5]. Extragastrointestinal intraabdominal schwannomas are even rarer and there have been only a handful of reported cases in the omentum, porta hepatis, adrenals, kidneys, abdominal wall and retroperitoneum [5,7].

Intraabdominal schwannomas are mostly benign, well demarcated and slowly progressive. Therefore, most patients are asymptomatic until they grow to a very large size to cause compression of surrounding structures. The majority is asymptomatic and often diagnosed incidentally on investigative imaging for nonspecific abdominal complaints. However, depending on their size and location they may present with symptoms of mass effect such as bowel obstruction, urinary or fecal incontinence as well as neurological symptoms of the lower extremities [5,8,9].

Intraabdominal schwannomas, especially the ones affecting the gastrointestinal tract, are frequently thought to be gastrointestinal stromal tumors (GISTs) prior to definitive tissue diagnosis. In Goh et al, 9 out 12 histologically confirmed intraabdominal schwannomas were initially thought to be GISTs [5]. This is because the intraabdominal schwannomas macroscopically appear as submucosal lesions, which is not a distinctive feature from other mesenchymal tumors. Furthermore, it is often difficult to obtain a representative sample of the deeper tissue, and even when the biopsy succeeds in obtaining deeper tissue samples most of them usually demonstrate nonspecific spindle cells, insufficient for a definite diagnosis [5].

On computed tomography (CT), gastrointestinal schwannomas are homogeneously attenuating on both non-contrast and contrast scans, and appear as well-defined mural masses [3]. The lack of low-attenuation hemorrhage, necrosis, and degeneration within the tumor may help distinguish these tumors from GISTs on CT, however, it is not a reliable radiological feature as some GISTs can appear as homogeneous tumors at times [10]. Hence, often the definitive diagnosis requires surgical excision as both radiological and endoscopic findings are nonspecific, and biopsies can be nondiagnostic, as demonstrated by our first patient and eight patients who underwent gastroscopic biopsy in Goh et al [5].

Once tissue specimen is obtained, immunohistochemical testing should be performed to aid in the diagnosis. Typically, schwannomas are composed of spindle cells with alternating areas of hyper- (Antoni A) and hypo- (Antoni B) cellularity [11]. S-100 protein is a key marker in the diagnosis of schwannoma [12] and is positive in almost all schwannomas. GIT schwannomas may rarely express CD34 cells, but CD117, SMA, and desmin are uniformly negative [13]. Therefore, the immunopositivity in CD117, CD 34, SMA and desmin is critical in distinguishing the other gastrointestinal mesenchymal tumors such as GISTs from GIT schwannomas [5,14]. Almost all GISTs are immunopositive for c-Kit and

DOG1, whereas schwannomas are negative for c-KIT and DOG1, and strongly immunopositive for S100 [12].

The treatment of choice depends on the size and the location of the lesion, but generally involves complete surgical excision. This is because it is often difficult to differentiate these tumors from other mesenchymal tumors, some of which are malignant [15]. Also, though uncommon, recurrence has been reported possibly due to incomplete resection in up to 10% of the reported cases [16]. However, the authors believes that it is not unreasonable in the cases of difficult tumor location or high risk surgical patients precluding complete surgical excision the lesion may be biopsied for tissue diagnosis and be monitored for any progression on serial imaging. In our second case, given diagnostic FNA and largely asymptomatic patient we have elected to monitor the lesion on serial imaging. In both of our cases at 3 month follow-up, there was no clinical or radiological evidence of recurrence or rapidly progressive lesions. Hence, the plan was made for a surveillance gastroscopy 1 year from the date of surgery for the first patient, and yearly surveillance CT for the second patient.

Intraabdominal schwannomas are mostly benign, however, there have been reported rare cases of malignant schwannomas [18,17], which are known to be refractory to chemotherapy and radiotherapy with poor prognosis [18]. Whether these malignant forms of schwannoma arise de novo or underwent malignant transformation remains debatable [19].

Figures

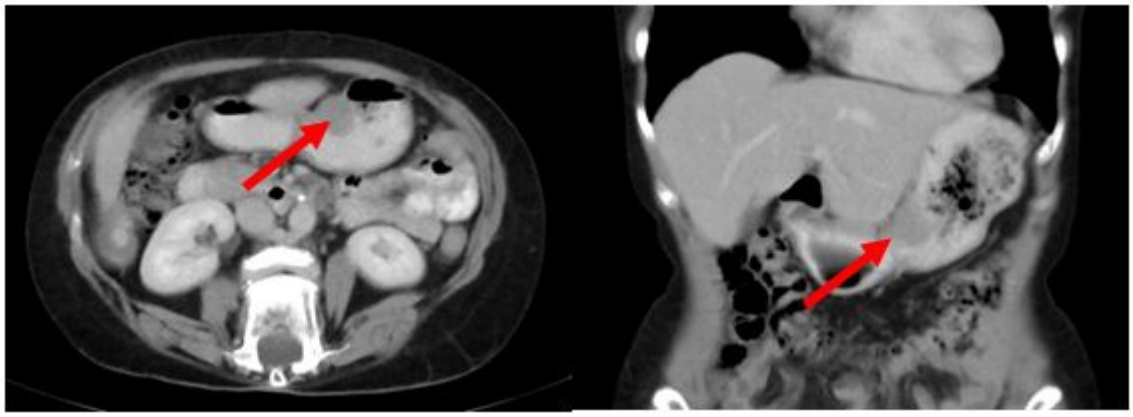

Figure 1: Portal Venous phase CT of Gastric Schwannoma

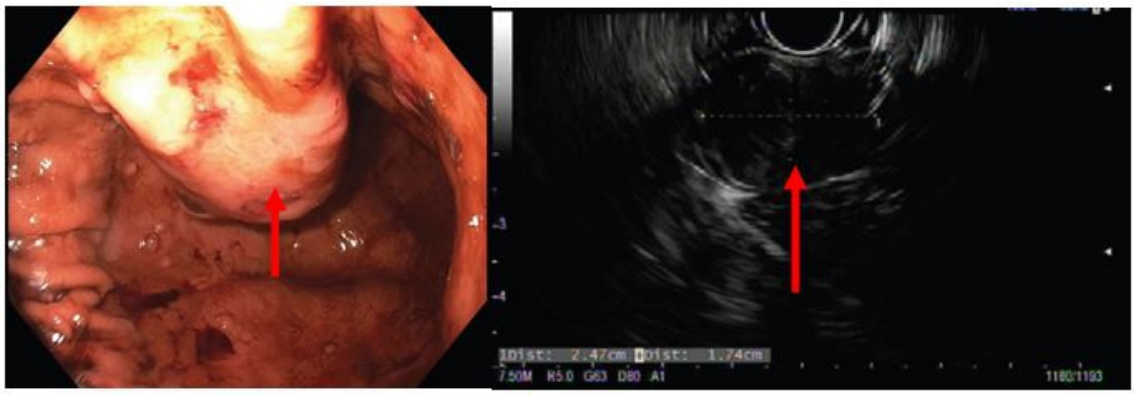

Figure 2: Endoscopic and endoscopic ultrasound appearance of Gastric Schwannoma

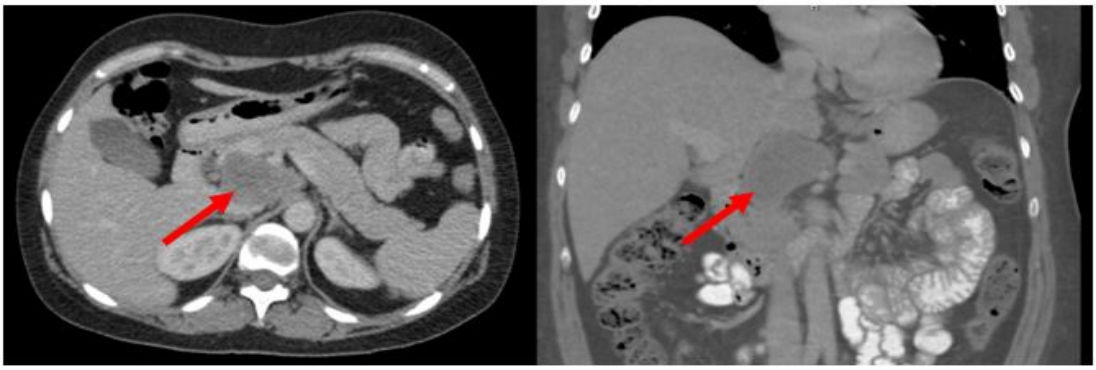

Figure 3: Portal Venous phase CT of Retroperitoneal Schwannoma

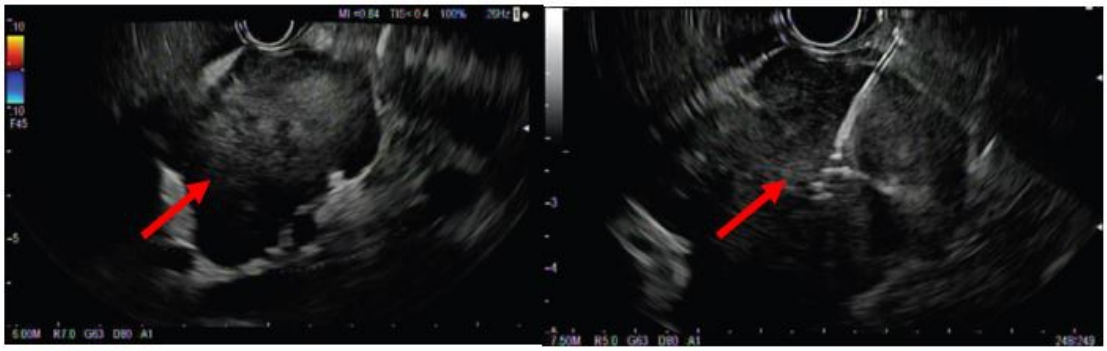

Figure 4: Endoscopic ultrasound appearance (second figure with biopsy needle) of Retroperitoneal Schwannoma

Conclusion

Intraabdominal schwannomas are rare forms of schwannoma, which are predominantly benign lesions. They are often asymptomatic and commonly diagnosed incidentally. They often provide diagnostic challenge as their endoscopic and radiological appearances are nonspecific, and share similar features with other mesenchymal tumors. It is only on immunohistochemical testing that we can reliably diagnose intraabdominal schwannomas as they demonstrate specific immunopositivity. Both diagnostic and treatment of choice is complete surgical excision as it offers most reliable diagnostic and therapeutic results.

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