

A Case of Cystic Schwannoma of the Lesser Sac

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Abstract

Introduction: Benign cystic schwannoma of the lesser sac is an extremely rare condition and only 3 reports were found in our review of the English literature. **Clinical Picture:** We describe a 58-year-old man with a large (5.2 x 6.7 x 7.6 cm) epigastric mass with solid and cystic components detected on sonography and computed tomography. Close association with the pancreas, stomach and liver led to a mistaken diagnosis of pancreatic cystadenoma/cystadenocarcinoma. **Treatment and Outcome:** The mass was surgically resected and our patient has been well since. **Conclusion:** This case draws the reader's attention to a rare condition that may mimic other sinister lesions, and highlights the pitfalls of diagnosis. Depending on the size and extent of the lesion, imaging may assist in characterisation of a schwannoma of the lesser sac. Surgical resection would still be the end point of management, especially when mass effect causes debilitating symptoms. Surgery is usually associated with good outcome, and tumour recurrence is unusual, provided adequate margins are resected.

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Introduction

Schwannoma or neurilemmoma (NL) is a well-defined, usually benign, tumour arising from the nerve sheath (Schwann cell). It may present as a solitary mass in any part of the body, but is more commonly seen in the head and neck, the extremities and on the trunk.¹ It is rarely found in the abdomen, although cases involving the small bowel, pancreas, pelvis and retroperitoneum have previously been reported.

In our literature review of English language journals, it was found that lesser sac schwannomas were reported by Noonan et al in 1975,² Sakai et al in 1988,³ and most recently by Muhammad in 1997.⁴

Clinical Picture

A 58-year-old man presented with non-specific symptoms of weight loss, bloatedness of the abdomen and increased swelling of the face over a period of 2 months. There was no significant appetite loss or fever. No chest discomfort or breathlessness was experienced. Physical examination was unremarkable. Significantly, there was no palpable abdominal mass.

Initial abdominal sonography (Figs. 1a and b) revealed a 5.2 x 6.7 x 7.6 cm left epigastric mass, closely related to the pancreatic tail, abutting the inferior aspect of the left hepatic lobe and stomach. It contained multiple hypoechoic cysts with mixed internal septations and solid areas. Intravenous contrast-enhanced abdominal computed tomography (CT) scan (Figs. 2a and b) documented a well-defined, mixed solid-cystic lesion, composed mainly of septated regions of fluid attenuation amid solid areas that showed heterogeneous enhancement. It appeared closely related to the body of the pancreas and was diagnosed as pancreatic cystadenoma/cystadenocarcinoma.

Treatment

The elective laparoscopic excision of the lesion which followed revealed a well encapsulated, 8-cm diameter cystic mass arising from the lesser sac. The adjacent pancreas, stomach, liver and spleen were not involved. Cut sections of the mass contained mucoid material. Gross appearance was compatible with gastric duplication cyst or mesenteric cyst.

Histological examination, however, confirmed the mass

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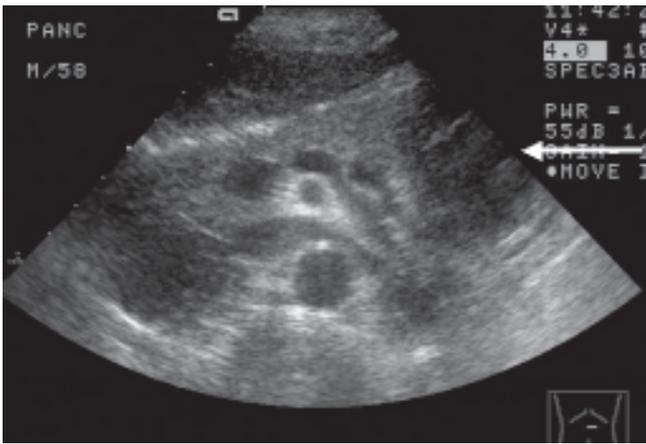


Fig. 1a.

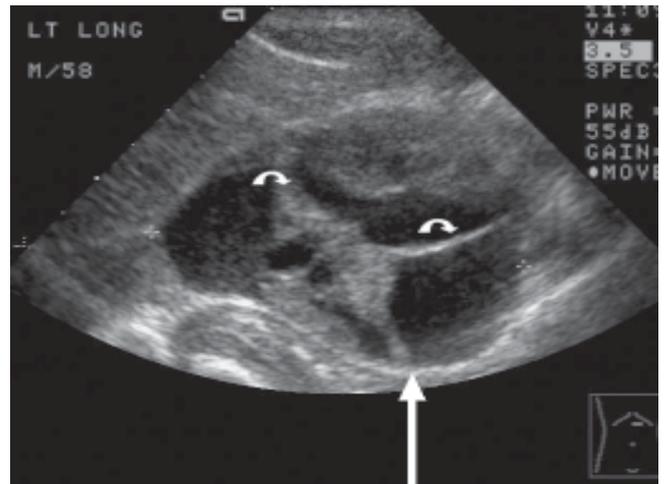


Fig. 1b.

Figs. 1a and b. Sonogram shows a multicystic lesion with internal septations (∞) and solid areas in the left epigastric region (white arrow).

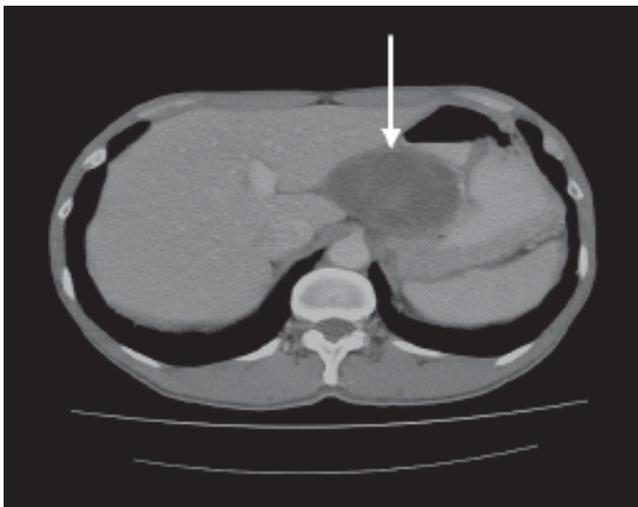


Fig. 2a.

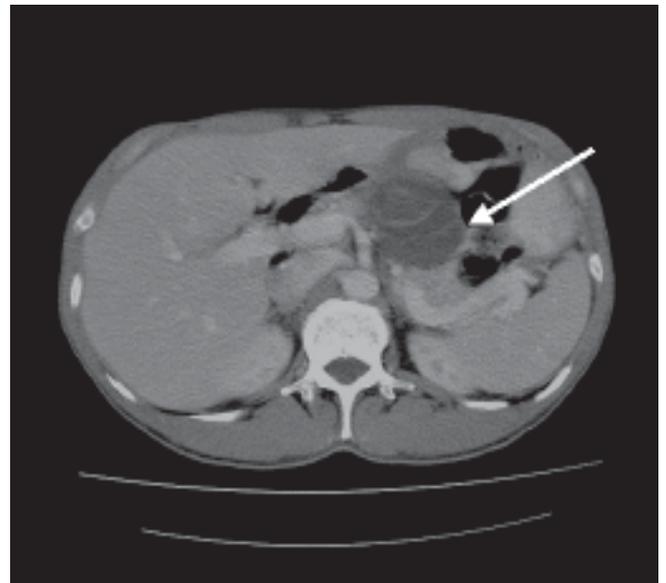


Fig. 2b.

Figs. 2a and b. Computed tomography shows a heterogeneous enhancing mass, mixed solid-cystic, closely related to the pancreas, liver and stomach (white arrow).

to be a schwannoma composed of Antoni A and B areas, with cystic change and degenerative nuclear atypia present. Staining for S-100 protein strongly highlighted the spindle cells. There was no evidence of malignant cells. Postoperatively, convalescence was uneventful, and the patient remains well and asymptomatic.

Discussion

Schwannomas/neurilemmomas are benign, slow-growing, solitary and well-encapsulated lesions that arise from the Schwann cells, not incorporating the nerve root. There is a 5% association with neurofibromatosis type I.

Extracranial schwannomas may take the form of a solitary mass anywhere in the body. Common sites include the head and neck,^{5,6} the flexor surfaces of the upper and lower extremities,^{7,8} the posterior mediastinum in the thorax^{9,10} and on the trunk.¹¹ The spinal and sympathetic nerve roots are often involved.

Intra-abdominal presentation is rare. However, cases involving the retroperitoneum,^{10,11} small bowel, extrahepatic biliary tree, pancreas, pelvis⁷ and sacrum have been reported as rare sites of occurrence. Schwannomas exclusively involving the lesser sac are extremely rare, with 3 reports published in the English literature.²⁻⁴ Several other reported

cases of lesser sac schwannoma actually originated from the gastric mucosa.

First described by Ackerman and Taylor in 1951,¹² ancient schwannoma is a rare variant that consists almost entirely of Antoni type B tissue. It is characterised by degenerative changes – cyst formation, calcification, haemorrhage and fibrosis – and cytologic atypia. They are typically asymptomatic and are therefore difficult to diagnose in the absence of clinical symptoms. On their own, schwannomas are not known to cause pain. The pressure they exert on adjacent structures or nerves usually brings these tumours to clinical attention. In our patient, it is likely that the lesion originated from a wandering branch of the vagus nerve.

Histologically, schwannomas may demonstrate a biphasic pattern with areas of highly cellular (Antoni type A) and myxoid matrix (Antoni type B) predominance.^{13,14} S-100 protein is demonstrated via immunohistochemical staining, particularly in Antoni type A areas.¹⁴ Despite sometimes striking cytologic atypia, mitotic figures are rare.¹⁵ It is postulated that degenerative change occurs due to the long period of time over which large schwannomas develop.¹⁶ Waxing and waning of the tumour size, when noted, is attributed to fluctuations in the amount of cystic change within the neoplasm.¹⁵

Incompletely excised lesions, usually due to anatomically challenging locations, are capable of slow recurrence. As such, higher recurrence rates are noted with intraspinal, sacral, and intracranial schwannomas, as well as with the plexiform variety and large tumours (e.g., giant sacral schwannomas).^{12,14,15} However, a recent clinicopathologic study found that patients with asymptomatic schwannomas occurring in association with neurofibromatosis type 2 (NF2) not only had more severe neurologic deficits but also experienced little postoperative improvement and a higher rate of tumour recurrence.¹⁵ In exceedingly rare circumstances, locally aggressive behaviour was observed in tumours with increased cellularity, higher mitotic rates (mean, 4 per 10 high-power fields), and underlying bone extension (observed in occasional cases of orbital NLs).

In the normal work-up for an abdominal mass, ultrasonography is often the first-line imaging modality, as it can indicate the presence of a mass in the mid-abdomen and can differentiate cystic from solid tumours. Unfortunately, in our case, the primary site of the tumour and its characteristics were difficult to determine. Therefore, an abdominal CT scan was deemed the study of choice for diagnosis. CT scan provides anatomic details and, depending on lesion size, can often identify the primary tumour site. CT may also demonstrate displacement or compression of adjacent organs.

On sonography, CT and magnetic resonance imaging

(MRI), the appearance of mixed solid content with cystic areas often results in mistaken diagnosis for other tumour types.^{14,15} Pancreatic tumours (cystadenoma, cystadenocarcinoma), smooth muscle tumours (leiomyoma, leiomyosarcoma), fat tumours (lipoma, liposarcoma), fibrous tumours (fibroma, fibrosarcoma), vascular tumours (haemangiopericytoma), omental metastases and neural sheath tumours are known mimics with similar imaging appearances. The ability to delineate margins of the tumour is helpful in localising the organ of origin, and therefore in the narrowing of the long list of possible differential diagnoses of the lesser sac mass.

Multiphase reconstruction, possible on both CT and MRI, can further aid in the determination of the origin of the mass. Characterisation of the mass on various triplanar MRI gradient echo sequences, including in and out phase algorithms, might have further contributed to diagnosis. However, the likelihood of image degradation from motion artefact, and the sheer size of the mass, cast doubts on any additional information that might have been obtained and thus, the value of MRI in our patient. Many of the possible differential masses in the region can also have varying contents of fluid, fat, haemorrhage, mucin and protein, and produce an image of mixed/heterogeneous appearance on MRI.^{14,15} A point to note in our case is that the sheer size of the tumour, with compression of adjacent structures, not only prevented the identification of the organ of tumour origin, but also resulted in the associated symptoms and made it impossible to exclude malignancy. It was therefore necessary to resect the lesion.

The proximity to the pancreas, stomach and liver, amongst other abdominal structures adjacent to the lesser sac made diagnosis a challenge, even at surgical excision. Depending on the site and size of the lesion, histological correlation via image-guided biopsy may be an option, potentially sparing the patient major surgery. However, the potential for needle track seeding from malignant lesions and the possibility of haemorrhage, and other possible complications, need be factored in before proceeding. In our case, this option was not a consideration due to the size of the lesion, and the symptoms experienced by the patient.

Conclusion

While schwannoma rarely occurs in the abdomen, cases involving the retroperitoneum, greater omentum, small bowel and pancreas have been reported. This case illustrates that they can occur anywhere in the body, including the lesser sac. On diagnostic abdominal imaging, lesser sac schwannoma may easily be mistaken for more sinister pathology due to the imaging features of heterogeneous solid and cystic components commonly seen in malignant lesions. Multimodality imaging such as sonography and

CT is often required for assessment of abdominal masses. MRI may also be useful but the high cost and possible motion degradation of the images, may limit its value. Image-guided biopsy is an alternative to major surgery, depending on the accessibility and size of the lesion.

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