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Case report

A rare case of ancient schwannoma of the lesser sac

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Abstract:
We report a case of ancient schwannoma of the lesser sac in a 35 year old man who presented with a 6 months history of pain in upper abdomen and early satiety. On examination he had a vague lump in the epigastrium. CT scan revealed a solid mass lesion located in the lesser sac. The mass was surgically resected and our patient has been well since. Histopathology and immunohistochemistry were suggestive of ancient schwannoma. Postoperatively, Patient is asymptomatic after 3 months. This case draws the reader's attention to an extremely rare condition of ancient schwannoma presenting as an intra abdominal mass located in the lesser sac and 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. Ancient schwannoma has to be differentiated from gastro intestinal stromal tumor since its clinical course and management differs.

Keywords: *Ancient schwannoma, lesser sac, Epigastrium, Histopathology and immunohistochemistry*

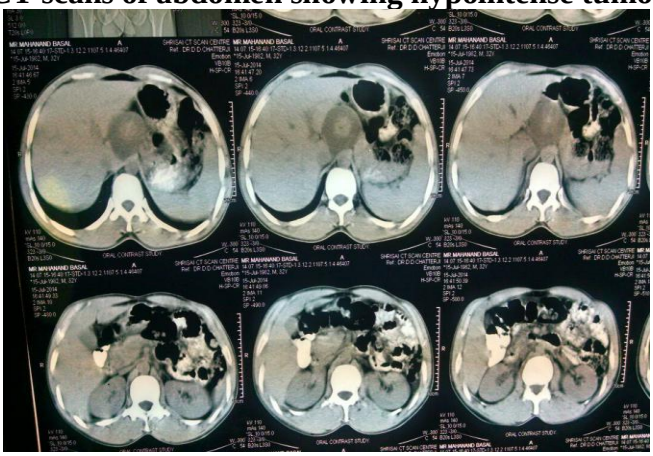
Introduction

Schwannoma is a benign neurogenic tumour arising from Schwann cells of the peripheral nerve sheath, which is commonly seen in women in the 5th decade. 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]. Ancient schwannoma is a variant of schwannoma which grows very slowly, deeply seated in the body and shows degenerative changes. It is seen in the middle age and elderly age groups. Ancient schwannoma of the lesser sac is a very rare occurrence. There have been only 3 reported cases so far to the best of our knowledge [2-4]. Several other reported cases of lesser sac schwannoma actually originated from the gastric mucosa. Here, we report a rare case of ancient schwannoma of the lesser sac, which was successfully diagnosed preoperatively and completely excised by laparoscopic approaches.

Case Report

A 35 years old male presented to our outpatient department (OPD) with a 6 months history of pain in upper abdomen and early satiety. There was no history of vomiting and jaundice. On physical examination there was an ill defined lump in the epigastrium. Initial abdominal ultrasonography revealed a well defined round hypoechoic lesion of size 6.5x5 cm in epigastric region anterior to body of pancreas and posterior to stomach showing minimal vascularity. Intravenous contrast-enhanced abdominal computed tomography (CT) scan (Figures 1) showed hypodense soft tissue attenuation with well defined round mass lesion in epigastric region with hyperdense central area of size 5.5x5.2x5.0cms.

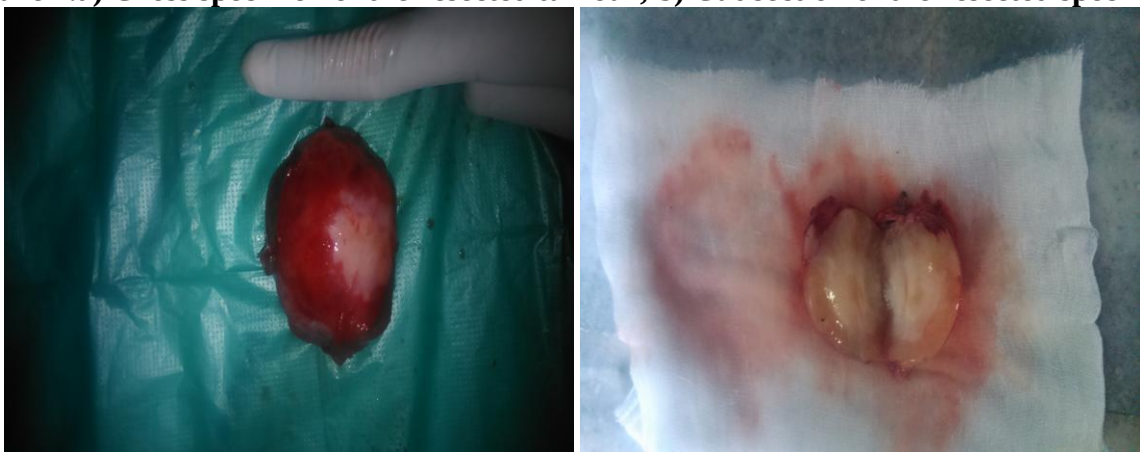
Figure 1. CT scans of abdomen showing hypointense tumor in the lesser sac.



Posteriorly the mass was abutting pancreatic body and located between lesser curvature of stomach and left lobe of liver. The differential diagnoses leapt were extra luminal gastro intestinal stromal tumor, lipoma and lymphoma.

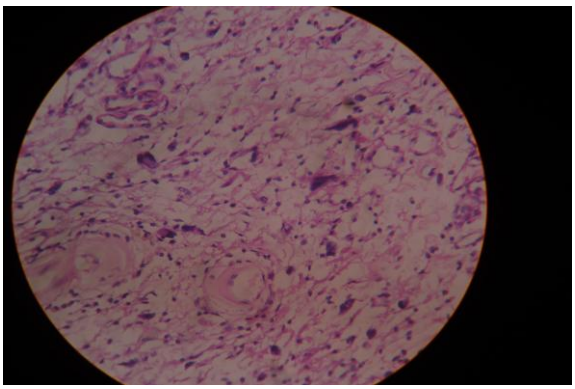
Following these investigative findings, an exploratory laparotomy was performed, upon exploration the mass was seen arising from the lesser sac. It was separate from the adjacent liver, stomach and pancreas. The tumor was excised in toto. Figure 2 shows that the gross specimen and cut section of the resected tumour.

Figure 2.a) Gross specimen of the resected tumour, b) Cut section of the resected specimen



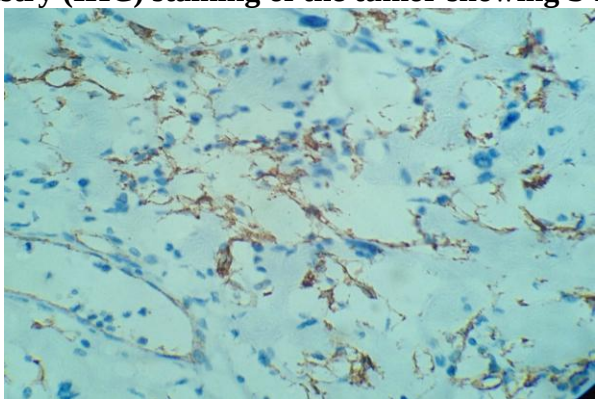
Histopathologic examination reported a benign schwannoma of ancient variety with densely hyalinised spindle cell neoplasm showing degenerative changes i.e. wavy nuclei showing hyperchromasia, pleomorphism with bizarre nuclei (Figure 3).

Figure 3. Showed H and E stained microscopic image of the tumor showing densely hyalinised spindle cells



Diagnosis was confirmed with immunohistochemistry, which revealed neoplastic cells, which were diffusely positive for S-100 protein (Figure 4) and negative for CD117/CD34/smooth muscle actins. Hence a diagnosis of ancient schwannoma was made.

Figure 4. Imuunohistochemistry (IHC) staining of the tumor showing S 100 protein positivity



Postoperatively, recovery was uneventful and patient was discharged after 7 days. Patient was followed up on regular basis clinically at 3 months post surgery. After 3 months of surgery, the patient is asymptomatic and doing well.

Discussion

Abdomen is a pandora's box. The differential diagnoses that are to be kept in mind are numerous. Neurogenic tumors are one such group of tumors that are to be considered while evaluating a mass per abdomen case. These tumors give rise to vague symptoms and can present late due to their benign nature. Schwannoma is a well-defined, benign tumor, which arises from neural crest cells and surrounds the nerve sheath. Ancient schwannoma are typically asymptomatic and are therefore difficult to diagnose in the absence of clinical symptoms. On their own, schwannoma are not known to cause pain. The pressure they exert on adjacent structures or nerves usually brings these tumors to clinical attention.

In the normal work-up for an abdominal mass, ultrasonography is often the first-line imaging modality, as it can indicate the presence of mass in the mid-abdomen. Ultrasonography is inexpensive but very non-specific in the detection of these tumors. In our case, the primary site of the tumor 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 tumor site. CT scan may also demonstrate displacement or compression of adjacent organs.

Definitive diagnosis of schwannoma is based on histological analysis. Histologically schwannoma are composed of bipolar spindle cells with a focal nuclear palisading pattern. There are areas of high cellularity named Antoni A and with low cellularity and myxoid matrix named Antoni B [5]. When schwannomas show pronounced degenerative changes including cyst formation, hemorrhage, calcification, and hyalinization these are termed "ancient" schwannomas [6]. "Ancient variety" is a rare form of schwannoma, which was first described by Ackerman and Taylor in 1951[7]. Ancient schwannoma is a variant of peripheral nerve sheath tumors which is characterized by diffuse hypocellular areas, relative loss of Antoni type A tissue, focal accumulations of hyaline material, calcification, cystic necrosis, haemorrhage and fatty degeneration [8]. These degenerative features are attributed to the growth and "aging" of the tumour, histologically characterised by nuclear hyperchromasia, mild nuclear pleomorphism, stromal oedema, fibrosis and xanthomatous changes leading to a misdiagnosis of malignancy in the aspirates [9]. Immunohistochemically, these tumors show diffuse positivity for S100 protein in the cytoplasm of the tumor cells [9].

Conclusion

In 35 year old male, ancient schwannoma occurring in a relatively small to medium sized in the epigastric region with degenerative changes was indeed a rare finding. It is important therefore to accurately assess and investigate such patients, who present with non-specific symptoms and despite

radiological and cytological investigations, pose a preoperative diagnostic dilemma due to degenerative changes. Surgical excision of the entire tumor is the treatment of choice.

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