Retroperitoneal Ancient Schwannoma: A Benign Tumor

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Abstract

Schwannomas are uncommon benign neoplasms arising from the Schwann cells lining the nerve sheath. Retroperitoneal schwannomas are extremely rare constituting about 0.3-3.2% of all retroperitoneal masses. Pancreatic schwannomas are obsolete and less than 50 cases reported until date in the literature. Ancient schwannomas is a histological variety in which there would be degenerative changes occurring with increasing duration of these tumors. We report 4 cases of retroperitoneal ancient schwannomas with one of pancreatic origin. Patients underwent pre-operative imaging studies (computed tomography/magnetic resonance imaging). All cases underwent exploration and en bloc excision of the tumors. The diagnosis was confirmed on both histopathological and immunohistochemical analysis. Pre-operative diagnostic dilemma in retroperitoneal schwannomas arises as they are either asymptomatic or have vague symptoms and signs. The radiological findings are not characteristic in the majority of the time. Fine-needle aspiration cytology is inconclusive and accessibility is always challenging due to its location. Pancreatic ancient schwannoma described here is unique due to the extreme rarity of location and was diagnosed intraoperatively on frozen section. As most of these tumors are in the paravertebral position in close proximity to important neurovascular structures it causes a technical challenge for a surgeon to operate. Resection of the tumor in toto is the treatment of choice for all tumors.

Key words: Benign, Excision, Schwannoma, Tumor

INTRODUCTION

Schwannoma is an uncommon benign neoplasm arising from the Schwann cells lining the nerve sheath. They are usually found in extremities, head, and neck region. Retroperitoneal schwannomas are quite rare accounting to 0.3-3.2% of all schwannomas. Pancreatic schwannomas are very rare and only 50 cases being reported in the literature. Some Schwannomas are termed “ancient” because of the degenerative changes which occur with increasing duration of these tumors. We present 4 cases of retroperitoneal ancient schwannomas with one of pancreatic origin.

CASE REPORTS

Case 1

A 48-year-old female presented with a mass in the right lower abdomen of 6 years duration which was painless, slowly progressive in size with no co-morbidities. On abdominal examination, fullness was noted in the right iliac fossa with no other findings on inspection of the abdomen. On palpation, a 10 cm × 6 cm vertically oval shaped non-tender mass was felt in the right iliac fossa which had a smooth surface and hard consistency. The mass was immobile, the plane being retroperitoneum.

Her routine blood investigations were within normal limits. Contrast enhanced abdominal computed tomography (CT) showed a smooth well-defined mass measuring 10 cm × 8 cm in the right retroperitoneum, displacing the right ureter and the iliac vessels with no local infiltration.

The mass was approached through a midline abdominal incision. The ileocecal junction was mobilized to expose...
the peritoneum overlying the tumor in the retroperitoneum. The peritoneum was incised, and the tumor capsule was exposed. A 12 cm × 8 cm tumor was noted extending from the level of the right renal artery to the bifurcation of common iliac artery and the tumor was found to be pushing the common iliac vein anteroinferiorly. The right ureter was coursing inferiorly over the tumor. On further dissection, the tumor was noted to be arising from the right lumbar sympathetic trunk (Figure 1a). The tumor was excised into sparing the nerves.

Gross examination showed the tumor to be well encapsulated and on cut section; there was heterogeneous yellow cheesy xanthomatous degeneration (Figure 1b).

Post-operative period was uneventful (Figure 1c) and at 1 year follow-up; there were no clinical or radiological evidence of recurrence.

Case 2
A 35-year-old female presented with a history of mass and vague pain in the lower abdomen since 1 year. She was asymptomatic otherwise. On abdominal examination, lower abdominal distension was noted with no visible peristalsis or prominent veins. A 20 cm × 15 cm mass was palpable in the lower abdomen, the lower border of which was extending into the pelvis. Per vaginally, no mass was palpable in the fornices.

She underwent contrast enhanced CT of abdomen and pelvis which revealed a heterogeneous mass in the left retroperitoneum measuring 20 cm × 10 cm. A midline laparotomy incision was performed, and a huge mass was found to be occupying the lower abdomen from umbilicus to the pelvis (Figure 2a). The left ureter was coursing over the mass. The mass was expected to be originating from the organ of Zukercandl. The tumor was excised with meticulous dissection and thorough hemostasis.

On gross examination, the tumor was nodular with a breach in the capsule (Figure 2b). On cut section, the tumor showed white cheesy areas with cystic degeneration. The histopathological examination showed malignant schwannoma. Her post-operative period was uneventful. During her follow-up, she was clinically and radiologically disease free until 1 year.

Case 3
A 27-year-old female presented with 1 month history of pain abdomen in the right hypochondrium and epigastrium. The pain was insidious in onset, dull aching in nature and associated with nausea, decreased appetite and early satiety. There was no significant past history. Abdominal examination revealed tenderness in the epigastrium and right hypochondrium with no palpable mass or organomegaly.

All laboratory investigations including complete blood count, liver function and renal function profiles, coagulation profile, serum amylase, serum lipase, carcinoembryonic antigen and cancer antigen 19-9 were within normal limits.

Contrast-enhanced CT abdomen and pelvis (plain and contrast) showed a well-defined round to oval heterogeneously enhancing lesion with areas of central calcification in the region of the head and uncinate process of pancreas measuring 38 mm × 43 mm × 49 mm. The pancreatic duct appeared normal measuring 2 mm. Anteromedially, the lesion was abutting the gastric antrum, gall bladder, portal vein. Posteriorly the lesion was abutting the right kidney and inferior vena cava. Cranially the lesion was abutting the caudate lobe of the liver (Figure 3a).

At laparotomy, a globular firm to hard mass measuring 4.5 cm × 3.5 cm × 3.2 cm was noted in the region of head and uncinate process of pancreas which was abutting the second part of duodenum (Figure 3b). The lesion appeared...
encapsulated and well circumscribed. A single 1 × 1 cm lymph node was noted along the right gastroepiploic vessels. Rest of the viscer was normal. The patient underwent wide local excision of the mass (Figure 3c) and excision of the lymph node with frozen section analysis.

Frozen section of pancreatic mass was suggestive of benign spindle cell lesion, and the lymph node showed reactive changes. Hence, in view of the benign nature of the pancreatic lesion, Whipple’s procedure was deferred. Complete hemostasis confirmed and abdomen closed with 28 Fr abdominal drain in the lesser sac.

Until the 3rd post-operative day, her drain output was 50 ml/day. Next day onward, the drain output increased to 300-500 ml/day and remained persistently elevated until the day of discharge. Pancreatic fistula was expected in this case because during the surgery it was hardly possible to identify and avoid injury to the undilated main pancreatic duct. Abdominopelvic sonography revealed no obvious collection. Enteral feeding was initiated on 7th post-operative period and she was discharged with drain in situ. On the 15th post-operative period, she got admitted in view of accidental removal of the abdominal drain. Since abdominopelvic sonography revealed no significant collection in the abdominal cavity, she was managed with stoma bag around the drain site (Figure 3d), prophylactic intravenous antibiotics, appropriate nutritional support, and regular dressings. The patient got discharged after 2 weeks. She is on regular follow-up monthly and on the 10th month of follow-up, she is asymptomatic and has gained weight.

**Case 4**

A 72-year-old female presented with pain in the lower abdomen of 3 months duration. She did not have any other gastrointestinal symptoms. She was a known diabetic and hypertensive on regular medications. Per abdominal examination revealed a vague mass in the left lumbar region, with the plane being retroperitoneum. CT abdomen and pelvis showed a central hypodense lesion with partial enhancement and calcification displacing the kidney laterally (Figure 4).

Abdomen was explored with a left paramedical incision. By a retroperitoneal approach, descending colon was mobilized. Left ureter was identified and mobilized laterally. Tumor was found to be arising from the left sympathetic trunk displacing the aorta medially. Tumor was excised sparing the nerve trunks. Postoperatively, the patient developed abdominal distension. A diagnosis of paralytic colon was made, and colonoscopic decompression was done. The patient is asymptomatic and free of disease both clinically and radiologically after 1 year of follow-up.

**DISCUSSION**

Schwannoma belongs to the benign peripheral nerve sheath tumors which also includes neurofibroma.1 The neoplastic cells of both neurofibroma and schwannoma are closely related to the normal Schwann cells which are derived from the neural crest.2 Schwannoma is less common compared to neurofibroma accounting for approximately 5% of all benign soft tissue tumors.3 Schwannomas are usually located in the head, neck, and flexor aspect of the extremities. They are rarely situated in mediastenum, pelvis and rectum. Up to 20% of these cases are associated with Neurofibromatosis Type 1.4
Retroperitoneal schwannomas are rare, and they account for 0.3-3.2% of all schwannomas. Most of the retroperitoneal tumors are considered malignant and retroperitoneal schwannomas mimick retroperitoneal sarcomas. Even though pancreas is richly supplied by both sympathetic and parasympathetic nerves, schwannomas arising from the nerves supplying the pancreas are extremely rare.

Retroperitoneal tumors constitute a difficult management problem due to their anatomic location, late presentation and proximity to adjacent vital structures thus making resection difficult or even impossible. Retroperitoneal schwannoma often attains a large dimension due to the presence of loose areolar tissue in the retroperitoneum, with non-specific vague clinical symptoms leading to a delay in diagnosis. They usually arise in women between the ages of 20 and 50 years. Patients present with poorly localized pain and discomfort, accompanied by non-specific digestive disturbances. Atypical presentations include flank pain and hematuria, headache, varicose veins of lower extremities, secondary hypertension, and recurrent renal colic pain.

Pancreatic schwannomas are usually diagnosed at the age ranging from 20 to 87 years (mean of 56 years) with equal sex distribution. In 40% of the cases, the tumor is located in the head of the pancreas followed by its body (20%). The size of the tumor ranges from 1 to 20 cm. Schwannoma usually occurs as a solitary lesion in the majority of cases, but can occasionally be multiple when associated with von Recklinghausen's disease. Patients present with nonspecific symptoms, usually with upper abdominal pain and discomfort (60%). Recent reports have shown that in 70% of cases, it can be asymptomatic. Other symptoms include nausea, vomiting, malena, abdominal mass, jaundice, and gastrointestinal bleeding.

Upon presentation, patients usually show clinical symptoms, which range from vague to predominant. The most common symptoms include intermittent abdominal pain (60%), nausea, vomiting, malena, abdominal mass, jaundice, and recurrent renal colic pain.

Laboratory investigations will not provide any assistance for the diagnosis of pancreatic schwannomas. Macroscopically, schwannomas are characteristically cystic, thin-walled, and hemorrhagic masses. However, solid and mixed tumors have also been reported. CT section will show well-demarcated, encapsulated mass with or without myxomatous and hemorrhagic areas. Microscopically, there will be two characteristic components, a highly ordered cellular component (Antoni A areas) consisting of closely packed spindle cells arranged in palisading and interlacing fashions (Figure 5a), and a loose myxoid/hypocellular component with degenerative changes (Antoni B areas) (Figure 5b). Verocay bodies without mitotic figures can also be noted.

Ancient schwannoma is a rare variant of benign peripheral nerve sheath tumor. Ackermann and Taylor coined the term “ancient schwannoma” in 1951. It morphologically resembles malignancy but is, in fact, a benign tumor. They are deep-seated and can attain large size. There will be less Antoni A areas and more hypocellular areas with areas of secondary degenerations such as hyalinization, calcification, necrosis, hemorrhage, cystic, and fatty degenerations. All these changes are believed to be due to the aging of the tumor, hence the term “ancient.” Sometimes because of the presence of nuclear atypia and pleomorphism, such tumors are mistakenly diagnosed as malignant. In such cases, mitotic count will provide the benign nature of these tumors. Ogren et al. described the use of flow cytometry to assess DNA ploidy and thus, helps in distinguishing benign schwannomas from malignant schwannomas. Immunohistochemically, the tumors are strongly immunoreactive for S-100 protein (Figure 5c), vimentin and CD56, and negative for cytokeratin AE1/3, CD34, CD117 (c-kit), desmin, and smooth muscle myosin.

CT of abdomen and pelvis usually show well-defined, round masses with multiple, low-attenuation, cystic necrotic areas. Antoni A areas (cellular component) in CT show inhomogeneous, hypodense, solid masses with contrast enhancement. Antoni B areas (loose myxoid) will be homogeneously cystic without significant contrast enhancement. Cystic changes occur more commonly in retroperitoneal schwannomas (66%) compared to other retroperitoneal tumors.

On magnetic resonance imaging (MRI), they appear as masses of low signal intensity on T1-weighted images and of high signal intensity on T2-weighted images. The signal intensity on T2-weighted images may differ depending on cell density. These findings are characteristic but not specific for schwannomas and have been reported to be present in only 57% of the cases. Hence, pre-operative diagnosis of retroperitoneal schwannomas is unusual.
Pre-operative radiological diagnoses as pancreatic cystic neoplasms, hepatic tumors, ovarian tumors, and psoas abscess have been reported. Gadolinium enhanced MRI may provide enhancement of tissue inhomogeneities within the tumor. MRI is generally accepted as the imaging modality of choice for most soft tissue lesions but is incapable of distinguishing between benign and malignant tumors reliably.

Question will arise whether to perform pre-operative biopsy when the radiological investigations provide a suspicion of neoplasm. Making pre-operative histological diagnosis by fine-needle aspiration biopsy or core needle biopsy has been shown to be unnecessary. Fine-needle aspiration cytology (FNAC) from degenerated areas will provide cells with pleomorphism giving a false interpretation of malignancy. Such large tumors will have large engorged veins coursing over the surface or within the tumor substance and these tumors are very close to aorta and inferior vena cava. Hence performing FNAC can cause torrential hemorrhage, tumor seeding and can introduce infection.

It is advised to perform FNAC and core needle biopsy under CT guidance. Proponents argue that pre-operative histological diagnosis enables to plan the proper intervention. If the radiologically diagnosed asymptomatic lesion is benign, one may consider regular timely radiological follow-up, especially in elderly individuals and the exemption from surgery, which is associated with high risk of complications.

Wide surgical resection has been advocated as the treatment of choice in retroperitoneal schwannomas based on the belief that malignant transformation occurs very rarely (0.7–2.6%) and that pre-operative histological confirmation of malignancy is not possible most of the times. Because of the benign nature of most of the retroperitoneal schwannomas, some authors have recommended simple intralesional enucleation of the tumor and laparoscopic piecemeal excision. The proponents argue that because of the indolent nature of the tumor, local recurrence will take considerable time and the resection of involved viscera and neurovascular bundles to attain clear margin can be avoided, thus reducing the morbidity and mortality associated with the radical surgery. Opponents argue that the rate of local recurrence after enucleation ranges from 16% to 54%, whereas recurrence after resection with a wide surgical margin has been reported in only 11.7%. Concern regarding the margin status arises when the histology reveals malignant transformation if the tumor had been excised in piecemeal fashion.

Management of pancreatic schwannomas remains to be controversial. Most of the pancreatic schwannomas are benign and hence if the histopathological diagnosis is obtained during the surgery, local excision of these tumors is sufficient and safe. If the definitive histopathological diagnosis cannot be confirmed, or if the tumor is of malignant potential during surgery, radical resection of the pancreas is contemplated based on the region involved. Hence, intra-operative histopathological confirmation of the pancreatic schwannoma will become important to avoid morbidity and mortality associated with radical resection. In our case, based on the frozen section report, we deferred a radical surgery on the patient with pancreatic ancient schwannoma, avoiding major post-operative complications and hospital stay.

CONCLUSION

We have described 4 cases of retroperitoneal ancient schwannomas with varied ages and symptoms of presentations and the methodical approach to the diagnosis and expert management. The majority of the retroperitoneal schwannomas presented with vague symptoms and were diagnosed when they attained a large size. The pancreatic ancient schwannomas described here is unique because the ancient variety developing in such young age and in such a small tumor is very rare.

There will be a pre-operative diagnostic dilemma because there are no specific findings in the imaging studies and FNA may not be conclusive because sampling from the representative area is a mere chance. Resection of the tumor in toto is the treatment of choice for benign schwannomas and wide local excision in the case of malignant ones. Most of the retroperitoneal ancient schwannomas are paravertebral in a location with close proximity to iliac vessels, aorta, ureter and other vital structures. Hence, it is a challenge for the surgeons to have adequate margin during dissection.

REFERENCES


Source of Support: Nil, Conflict of Interest: None declared.