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CASE REPORT: Giant Retroperitoneal Presacral Ancient Schwannoma

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ABSTRACT

Summary: Ancient schwannoma, is a rare variant of schwannoma with characterization of degenerative changes and diffuse hypercellularity. Retroperitoneal presacral form is often found incidentally, because they present with vague symptoms or symptomless. Schwannoma occurring in this area occasionally presents with enormous dimensions, known as a giant schwannoma. The tumor removal is a surgical challenge due to the difficult approach and abundant vascularity. In this report we describe a 61 –year old female presented to ER with vaginal bleeding and lower abdominal pain. The case diagnosed on clinical, CT and MRI findings to be a fibroma of the left ovary. Exploration by the gynecology team revealed a huge retroperitoneal presacral tumor compressing the left external iliac vessels and displacing the left ureter; they took a biopsy and closed the abdomen. Histopathological result was benign schwannoma. The patient were referred to our hospital (Al Hada Armed Forces Hospital, Taif, Saudi Arabia) to be managed from postoperative DVT when her family asked our department of surgery for further management and signed a high risk consent. We explored the case after insertion of IVC filter and ureteric catheter. A 20x20 cm mass was thoroughly dissected and resected with part of sacrum. The final histopathological result was benign nerve sheath tumor with features consistent with degenerated (ancient) schwannoma and the tumor was completely resected. The patient was discharged from the hospital without complications and follow up for three years revealed no recurrence. The clinical, radiological, and pathological features of this disease are discussed in this report.

To conclude, retroperitoneal giant ancient schwannomas are a rare variant of the benign schwannoma and often present as unrecognized slow growing masses. Keep in mind potentially severe bleeding and neurological deficit risk of surgical intervention without away from oncologic principle. Therefore, careful preoperative evaluations and postoperative monitoring are necessary.

Keywords: ancient schwannomas, retroperitoneal, vaginal bleeding, masses, tumors.

Introduction

Schwannomas are usually benign tumors arising from the Schwann cells of the peripheral nerve sheath (1). The benign schwannomas produce symptoms, which are nonspecific and depend on the location and size of the mass and results in compression and irritation of the nerve roots, gastrointestinal system, urinary

system, impingement of the major vessels, and even severe destruction of the adjacent bone. Most common symptoms are abdominal distention and abdominal ache/pain (2). Retroperitoneal schwannomas occur most commonly between 40 and 60 years of age, with a male/female ratio of 2:3 (3).

The diagnosis of patients with retroperitoneal schwannoma is usually delayed, because the retroperitoneal cavity is flexible, unless the tumor becomes a substantial size, symptoms are not developed (4). At the time of diagnosis, the size of a retroperitoneal schwannoma is significantly larger than 8 cm in diameter (5). Though schwannomas found in peripheral nerve fibers in the limbs, head, and neck, they are the cause of 0.5 to 12% of retroperitoneal masses (3). Grossly, schwannomas are solitary (except in the hereditary form, multiple Schwannomatosis), well circumscribed, firm, smooth-surfaced tumors. Because of their large size, these tumors are likely to manifest degenerative changes such as cysts and calcification (6). Microscopically, the benign variant has a variation of Antoni A and B areas, with a diffuse positivity for S100 protein in the cytoplasm of the tumor cells (4). Most schwannomas are benign and malignant degeneration of schwannomas is extremely rare. Malignant degeneration particularly occurs in association with Von Recklinghausen‘s disease (7).

Treatment depends mainly on surgery (8). Conventional approach is commonly used but laparoscopic resection may be useful (9). Complete excision with negative margins is the recommendation. Simple enucleation or partial excisions are controversial (10). Radiotherapy may be of value in malignant conditions, however, these tumors do not respond very well to chemotherapy (11). This report presents a case of giant presacral retroperitoneal schwannoma. The tumor characteristics and clinical pictures are reviewed. The problems associated with the surgical removal of this tumor are also discussed.

CASE REPORT:

Presentation: A 61 year-old postmenopausal woman, presented to ER at January 2009, with irregular vaginal bleeding for one-month duration. The bleeding was minimal and associated with lower abdominal pain. The pain was dull aching in nature, mild to moderate in severity with no reference or radiation. There is no other abdominal complaint and the systemic review was unremarkable. General examination showed no abnormality and abdominal examination revealed huge pelvi-abdominal mass of irregular borders and limited mobility. Vaginal examination detected no abnormalities.

Workup: The laboratory findings were within normal. Her abdominal computed tomography showed a large pelvi-abdominal mass measuring 19x16x20

compressing the uterus and seems to arise from the left ovary (Fig 1). The mass had an irregular surface and consisted of solid and necrotic areas. The magnetic resonance imaging confirmed the left ovarian origin with areas of degeneration and without evidence of lymphadenopathy (Fig 2, 3).

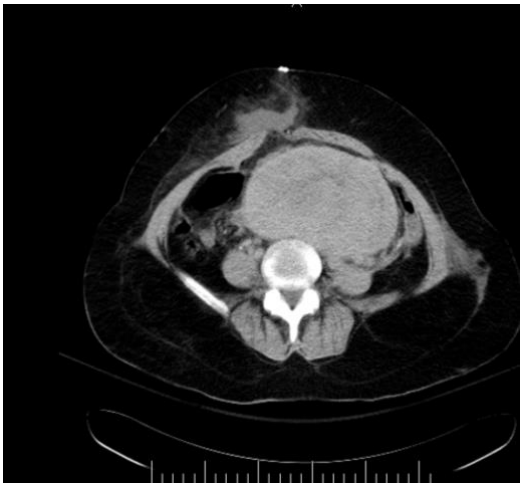

Figure 1: CT scan; showing the mass as it arises from the left ovary.

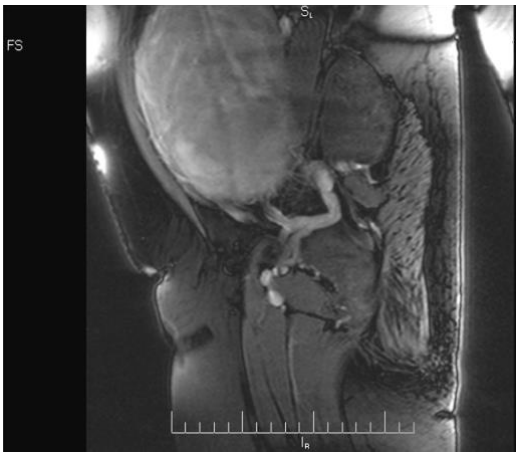

Figure 2: MRI shows the large size of the mass and its pelvi-abdominal nature.

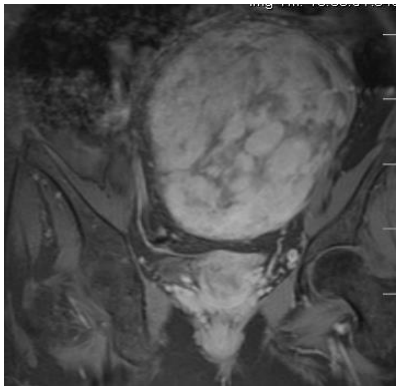

Figure 3: MRI showing areas of degenerations and necrosis.

The provisional diagnosis: was an ovarian fibroma and the gynecology team took a decision of total abdominal hysterectomy.

In the OR: a midline incision was done. The omentum was covering the mass so, a partial omentectomy was done. They started dissection of the mass, which was fixed inferiorly to the sacrum. The lower pole could not be reached. They reported that the mass was mainly uterine and fixed to the great vessels and to the retroperitoneal area. No further dissection was attempted to avoid injury of these vessels. Biopsy of the mass was taken. Abdomen was closed. Patient tolerated the procedure well. Histopathological examination of the specimen revealed a degenerated peripheral nerve sheath tumor with features consistent with benign schwannoma. The omentum was free from malignant deposits.

Postoperative: Unfortunately, on the second post operative day the patient started to complain of painful left leg swelling extending to the thigh. An ultrasound performed on the patient

showed evidence of deep venous thrombosis (DVT), in the left leg. The patient was referred to our hospital (Al Hada Armed Forces Hospital) for management of the DVT.

Al Hada Armed Forces Hospital: In our hospital, the patient received anticoagulant therapy under supervision of the internists. The patient's family consulted our department of surgery for a second opinion, regarding the further management of the mass. After discussing the case with the radiologists and neurosurgeons, we reached to conclusion that the tumor is huge but resectable, with high risk. The patient and her family informed about the operation with all the expected risks and possible complications, they agreed and a full informed consent was signed. Prior to the operation, IVC filter was applied.

In the OR: The wound was reopened and extended. Exploration revealed a mass of 20x20 cm in diameter. The mass was attached to the sacral bone, compressing the iliac vein, and pushing the left iliac artery and ureter laterally. The posterior peritoneum was opened and there was marked adhesions around the tumor. It was dissected meticulously from the abdominal wall and the surrounding structures preserving the ureter and the iliac vessels. Dissection was completed to the bony attachment with difficulty, but the mass was totally resected with its capsule and a small pad from the sacral foramen was also excised and sent for histopathology. During dissection, the colon was completely mobilized, so appendectomy was done to avoid clinical misleading, if appendicitis develops in its new position. Hemostasis was completed and the blood loss was less than 500 cc, the abdomen was closed.

Postoperative: The patient tolerated the procedure very well and transferred to the recovery room in a good condition, and spent the first postoperative day in the ICU. The internal medicine team followed the patient for the DVT. The patient passed the postoperative period smoothly, and she was discharged on 17th postoperative day without developing any complications. Follow up of patient for more than 3 years; revealed complete resolution of the DVT and she was doing well with no recurrence until her last visit on the 15th of February 2012.

The final histopathology report: A well-defined lesion surrounded by fibrous capsule showing residual nerve fibers. The lesion shows spindle cells proliferation with two patterns of growths, the first was Antoni type A areas, characterized by compact proliferation of spindle cells that have twisted nuclei and indistinct cytoplasmic borders (Fig 4). They are arranged in between interlacing fascicules of Antoni type B areas, which was less cellular and composed of spindled cells and oval cells arranged haphazardly within loosely textured matrix. Micro cystic changes, inflammatory cells, and delicate collagen fibers punctuated the matrix. The lesion also showed large, irregularly

spaced vessels with thick hyalinized walls. There are focal areas of degenerative changes including hemorrhage, hyalinization, infiltration by macrophages and lymphocytes, and focal mild cellular atypia. No hypercellularity, mitoses, necrosis, nor pleomorphism, were noted. The omentum shows hemorrhage, fat necrosis, and inflammation. The lesion was diffusely and strongly positive for S-100 and focally positive for NSE and negative for Actin, CD34, and CD117 (C-kit) (Fig 5). The lumen of the appendix mildly and focally dilated and occupied by mucinous material. There was a focal lining by atypical mucinous epithelium; however, the epithelium was mostly denuded. There was atrophy, fibrosis, and chronic inflammation of the underlying wall. No invasion of the wall was noted and no omental deposits were detected.

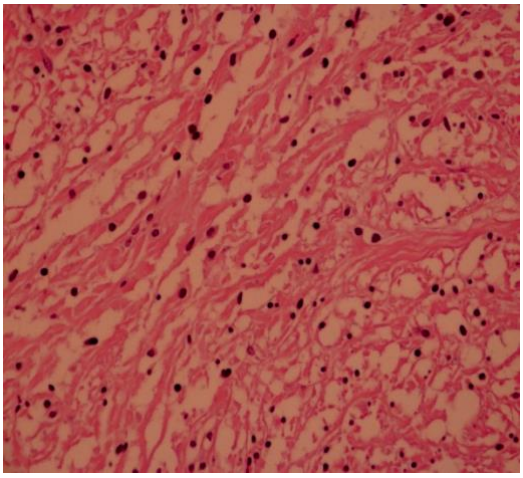

Figure 4: Shows the spindle cell nature of the tumor.

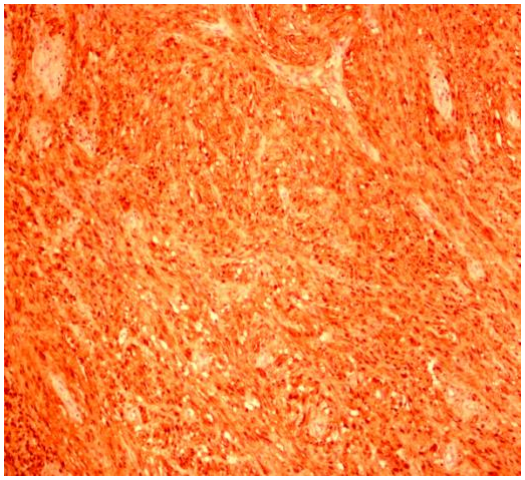

Figure 5: the specimen is diffusely positive for S100.

Final diagnosis: was benign nerve sheath tumor with features consistent with degenerated (ancient) schwannoma. The tumor was completely resected. The appendectomy specimen showed evidence of benign mucinous cystadenoma.

Discussion

Presacral tumors are uncommon and occur in approximately one of 40,000 hospital admissions (12). Schwannoma is one of the tumors that often occur in this area. Schwannoma also called neurinoma, neurilemmoma, and perineural fibroblastoma, and is a tumor originating from the Schwann's cells derived from the neuroectoderm (1). Masson, (13) has described the tumors as arising from Schwann's cells and termed as schwannoma. Schwannomatosis is a rare inherited condition, with multiple schwannomas of the different nerves (6). The majority of the schwannomas arise from cranial nerves or nerves of upper extremities but rarely they can arise from mediastinum and retroperitoneum (5). In

the present case, the tumor was retroperitoneal and presacral.

Pathologically, the tumor is grossly; a well-encapsulated lesion and well circumscribed, with firm and smooth-surface. Because of their large size, these tumors are likely to manifest degenerative changes such as cysts and calcification (6). In our case the tumor size was large, well encapsulated and with similar degenerative changes.

Microscopically, they demonstrate Antoni A areas, which are densely cellular, arranged in short bundles or interlacing fascicles and Antoni B areas, which characterized by being less cellular and organized, with great myxoid component. Characteristic immunochemical features are a positive S-100 and a negative CD-34 (14). The same findings were seen in the present case.

Most schwannomas are benign, malignant schwannoma is very uncommon and known to occur, in association with Von Recklinghausen's disease, which occurs in 5% to 18% of cases. In the absence of von Recklinghausen's disease, these tumors rarely occur in the retroperitoneum, comprising 0.5% to 5% of all schwannomas and 0.5% to 12% of all retroperitoneal tumors (7 & 15). In the present case, Von Recklinghausen's disease was not present but benign mucinous cystadenoma was accidentally discovered in the appendix, which was removed after mobilizing the colon. Malignant transformation of the benign lesions is also, exceedingly rare (1).

The retroperitoneal schwannoma occur most commonly between 40 and 60 years of age, with a male/female ratio of 2:3 (3). Our patient was 61-year old female, with benign form of retroperitoneal schwannoma.

At this site the tumor usually presents with slow growth and the symptoms are vague and nonspecific, such as vague abdominal pain and dull ache or low back pain, so, the diagnosis and treatment are often delayed until the tumor enlarges. In the present case, the presentation was by minimal intermittent vaginal bleeding and lower abdominal aching pain, a presentation that was vague and difficult to be interpreted.

The size of a retroperitoneal schwannoma is usually more than 8 cm in diameter at the time of presentation (4, 5 & 16). It may present by enormous dimensions, known as a giant schwannoma, which is difficult to manage (5 & 8). In this case, the size was 20x16x 20 cm, and at the first operation, the gynecology team failed to resect the mass.

Appropriate radiological evaluation is important both for diagnosis and for management (17). CT scanning, usually shows; well-defined mass with low or mixed attenuation. Retroperitoneal schwannomas show cystic degeneration in up to 60% of cases while calcification is seen in 23% of cases only (18). MRI findings usually show a low signal intensity

on T1-weighted images and high signal intensity on T2-weighted images (10). Depending on the cell density, the signal intensity on T2-weighted images may vary. Tumors with microscopic findings of hypercellular Antoni type A tissue have intermediate signals, while tumors with Antoni type B tissue have a bright signal on T2-weighted images (18). Although target and fascicular signs are characteristic radiological features of schwannoma, these are not frequently seen in retroperitoneal schwannomas. However these radiologic findings are characteristic but not specific of schwannomas and was noted in only 57% of the cases and based solely on imaging, the percentage of misdiagnosis was found very high (84%) (17).In our case, the radiologist did not find these characteristics and diagnosed the case to be ovarian fibroma. However, in the literature, the differential diagnosis should include retroperitoneal lymph nodes and ovarian tumors (19 & 20). The former can be distinguished by evidence of other lymphadenopathy and the latter can sometimes be distinguished by identifying the normal separate ovaries. We have also to Keep in mind the other differential diagnoses for a retroperitoneal schwannoma include, epithelial cyst, abscess, plexiform malignant peripheral nerve sheath tumor (MPNST), sacral meningioma, ependymoma, chordoma, chondrosarcoma, giant cell tumor, aneurysmal bony cyst, osteoblastoma, sacrococcygeal teratoma, lymphoma, and the malignant transformation of a benign tumor. These disease categories can be distinguished from schwannoma by consideration of the clinical symptoms, age, the radiologic image, subcutaneous paracentesis, and chromosomal study (12). Ganglioneuroma is a neurogenic tumor, which should be considered in the differential diagnosis. It is shown more commonly in the retroperitoneum than schwannoma and has similar findings on CT and MRI (21). However, Histopathological diagnosis is the only accurate diagnostic tool and surgical resection is the only accurate approach for pathologic evaluation to enable diagnosis of retroperitoneal schwannoma (4, 5 & 12). This is what occurred in our case.

Ancient schwannoma, a degenerative neurilemmoma, is a schwannoma subtype, characterized by degeneration and diffuse hypercellular areas. These changes are believed to occur because it takes a long time for schwannomas to develop (22 & 23). Similar degenerative changes were found in our case and the diagnosis was benign ancient schwannoma.

Some authors found that laparoscopic biopsy may be helpful but most of them do not recommend CT-guided biopsy for the diagnosis of retroperitoneal schwannoma (5 & 9). They mentioned that this maneuver is unreliable, as areas of degeneration and the cellular pleomorphism can hinder the diagnosis and degenerative cells may be misinterpreted as malignancy. However, it may be helpful

only if the sample contains enough Schwann cells to be visualized microscopically (5). The procedure carries the risk of hemorrhage, infection, and tumor seedling. To date, no standard diagnostic criteria or radiologic feature of malignant schwannomas has been described. Malignant schwannomas are commonly larger and they act as high-grade sarcomas with the possibility of producing local recurrence and distant metastasis. Typically, malignant schwannomas are diagnosed histopatologically, after the surgical excision of a mass, with features of high mitotic rate, pleomorphism, and blood vessel infiltration (7 & 14). In our case, there was no evidence of malignancy.

Surgery is the only hope for cure. Complete excision with negative margins is recommended, sacrifice of adjacent tissues and viscera may be considered for complete surgical excision (8, 10 & 11). In this case, the tumor was completely resected in the second operation with a piece of sacral foramen, from its nerve, the tumor arises. All surrounding structures were carefully dissected and preserved.

Simple enucleation or partial excision of the tumor is not sufficient as local recurrence rate ranging from 16% to 54% after conservative intralesional enucleation has been reported (8). Giglio *et al* (10), proposed that even if the tumor were determined to be benign using frozen biopsy, the possibility of malignancy could not be excluded accurately. Therefore, maximal removal as completely as possible of the operable part of tumor without severe hemorrhage and anticipated neurological injury level after surgery are two factors considered during the surgery. In this case, the tumor was completely resected and the bleeding was less than 500 ml.

A few reported cases in which where metastases demonstrated after resection of the histologically benign schwannoma, leading to recommendations of regular follow up (11). We are following the patient for more than 3 years and no detected recurrence or evidence of metastases up until now.

Single therapy modality of malignant schwannomas have shown poor results, so adjuvant radiation therapy and/or chemotherapy are required beside surgery (2 & 11). The prognosis for retroperitoneal benign schwannoma is good (1 & 15). On the other hand, even in cases in which the appropriate treatments are administered, prognosis is poor with the malignant variant (10).

Conclusion

Retroperitoneal giant ancient schwannomas are rare variant of the benign schwannoma and often present as unrecognized slow growing masses. Keep in mind potentially severe bleeding and neurological deficit risk of surgical intervention without away from oncologic principle. Therefore, careful preoperative evaluations and postoperative monitoring is necessary.

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