

Retroperitoneal Schwannoma, Incidental Detection - Case Report King Hamad University Hospital Bahrain

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ABSTRACT

Schwannomas are benign nerve sheath tumors that mostly occur in the head & neck, extremities, and mediastinum. These rarely present as retroperitoneal masses. A case was reported of an incidentally detected retroperitoneal schwannoma in a 32-year-old female. Schwannomas, the benign nerve sheath tumors, grow slowly but they may cause pain, weakness, numbness, or stay asymptomatic. Symptoms depend on the location and size of the tumor. Sometimes the patients, who present to the Accidents & Emergency with vague abdominal pain. Imaging incidentally may detect a pelvic lesion that is always diagnosed on histopathology after complete resection.

Key words: Schwannoma, Retroperitoneum, S-100, Vimentin, Cytokeratin AE1/AE3, HMB-45, desmin, smooth muscle actin and CD56.

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INTRODUCTION

Schwannomas (also termed neurilemmomas or neurinomas) are encapsulated nerve sheath tumor arising from Schwann cells, hence the name. Most of schwannomas are benign. These tumors are commonly found in flexor surfaces of the extremities, neck and head. Its location in the retroperitoneum is very rare comprising 0.5% to 1.2% of all retroperitoneal tumors.^{1,2} They usually arise from a single nerve bundle and are characterized by slow growth.³

CASE

A 32-year-old woman presented with vague nonspecific abdominal pain to the Accidents and Emergency. She had no history of fever, anorexia or weight loss. Abdominal examination was unremarkable. Blood investigations revealed no abnormalities.

Ultrasonography was done and showed an ovoid hypo-echoic area seen in the cul de sac to the right of the midline. It had a cystic component and was non-vascular and not attached to the ovaries (Fig.1 A).

CT showed a well-defined oval lesion measuring 4 x 3.8 x 6 cm in transverse, antero-posterior, and crano-caudal dimensions respectively. It was seen

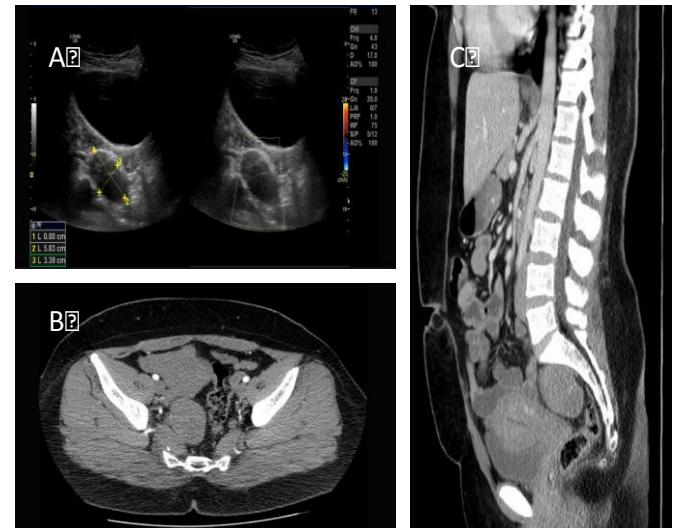


Figure 1: Photograph of a mass A, Ultrasonography was done and showing an ovoid hypo-echoic area seen in the cul de sac to the right of the midline. It had a cystic component and was non-vascular and not attached to the ovaries, B and C; CT showing a well-defined oval lesion measuring 4 x 3.8 x 6 cm in transverse, antero-posterior, and crano-caudal dimensions respectively. It was seen in the pre-sacral space

in the pelvis in the pre-sacral space. The right internal iliac vessels were seen lateral to this mass without any vascular encasement. The mass demonstrated mild heterogeneous enhancement after intravenous contrast. (Fig.1 B and C).

The MRI showed the mass to the right of the rectum and measuring 3.9 x 3.7 x 5.8 on maximum dimensions. It was separable from the uterus, in close proximity to the right ovary with no definite connection and with a 4 mm line of cleavage with the adjacent rectum. (Fig.2 A and B)

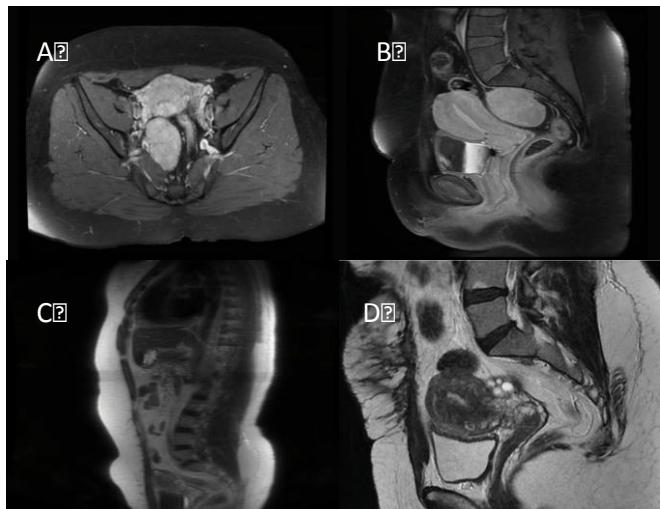


Figure 2: A; MRI showing the mass to the right of the rectum and measuring 3.9 x 3.7 x 5.8 on maximum dimensions Axial view. C MRI on sagittal view and D; The postoperative period was uneventful but no recurrence was seen on MRI.

The patient was prepared for surgery. Initially, laparoscopy was carried out followed by laparotomy, which revealed a pelvic retroperitoneal mass to the right of the midline in the pre-sacral space that was completely resected and sent to histopathology for a definite diagnosis.

Gross examination revealed a 65 x 30 x 30 mm well circumscribed lobulated soft yellowish grey mass weighing 45 gm. The outer surface was postulated however it was completely encapsulated. The cut surface was nodular with a focal area of yellowish discolouration. Histopathological examination showed an encapsulated tumor with moderate stromal cellularity, consisting mainly of spindle cells with clusters of stromal foamy macrophages

(Fig.3a, 3b). The tumor shows no nuclear atypia and no mitotic figures.

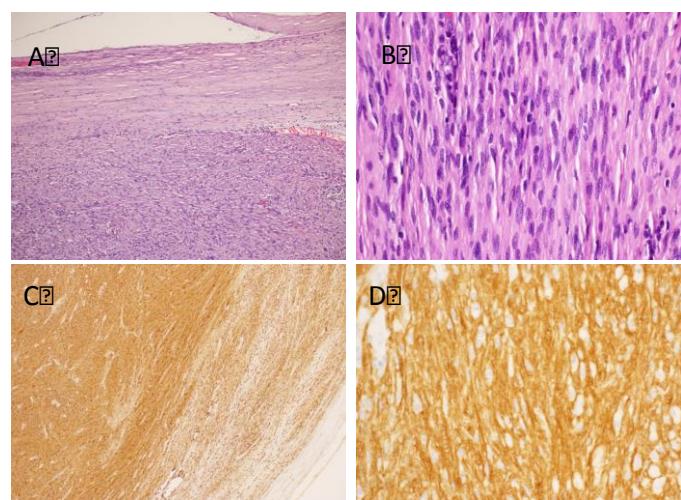


Figure 3: Photomicrographs of A; H&E: Cellular schwannoma characterized by highly cellular spindle cells B; H&E: Cellular schwannoma characterized by highly cellular spindle cells (40x) C, Tumor cells showing diffuse positivity for s100 (5x) and D; Tumor cells showing diffuse positivity for s100 (40x)

Immunohistochemical studies showed the tumor cells are positive with Vimentin (cytoplasmic) and S 100 (cytoplasmic and nuclear) confirming a neurogenic origin (Fig.3 C and D). Cytokeratin AE1/AE3, HMB-45, desmin, smooth muscle actin and CD56 are all negative. CD68 is positive at areas of highlight clusters of stromal foamy macrophages. The overall histological findings and the immunophenotype are those of cellular schwannoma. The postoperative period was uneventful. A follow up MRI after 6 months was done and revealed no recurrence (Fig.2 C and D).

DISCUSSION

Despite ultrasound being a simple non-invasive method that can be used initially to detect masses, MRI remains the imaging modality of choice.² It is used for the purpose of clearly identifying the origin, borders, and composition of lesions. However, CT is usually done prior to MRI to clearly localize the lesion after sonographic detection. On CT, benign schwannomas appear as low density homogenous masses that are well circumscribed with no calcifications.⁴

Intraoperatively, benign schwannomas are found to be well demarcated and are freely resectable.

Treatment is surgical as complete resection has proven to be curative in all cases of benign schwannomas and post-surgical recurrences are unusual. Hence, benign schwannomas are known to have an excellent prognosis.¹

Review of cases from the literature show that schwannomas form firm greyish masses of variable sizes often grossly show cystic degeneration. Microscopically, schwannomas show two different patterns of growth and are designated as Antoni A and Antoni B. The former is being more cellular and formed by palisading spindle cells while the latter is hypocellular and sometimes shows cystic and myxoid formation. Since the origin of the tumor is neuronal, these tumors are immunoreactive for s-100, vimentin and calcineurin.^{5,6} In this study, we present a case of cellular schwannoma variant. Which as the name implies, is hypercellular and composed mainly of Antoni A areas and lacks the organized nuclear palisades of verocay bodies.

The most widely applied ancillary technique today is immunohistochemistry to help in the diagnosis of cellular schwannoma and to exclude the other similar-appearing spindle cell neoplasms such as neurogenic tumors (including malignant peripheral nerve sheath tumor), synovial sarcoma, leiomyosarcoma, fibrosarcoma and malignant fibrous histiocytoma. Malignant transformation is rare; however few cases have been reported and are supported by the presence of histological mitotic figures, cellularity, nuclear atypia, and tumor necrosis.⁷ Frequent mitotic figures are the most reliable factor in correlation with malignancy. Malignant schwannomas are of two types: Solitary malignant schwannomas (SMS) and schwannomas associated with von Recklinghausen's disease (VRMS). The latter have a poorer prognosis with a shorter five-year survival (23%) compared to the former (47%).⁸ Also, patients with VRMS are usually younger and have centrally located schwannomas rather than peripherally located (Fig. 8). On CT, malignant schwannomas appear hypodense with mixed attenuation, which results from areas of tumor cellularity and of tumor necrosis and hemorrhage being adjacent to each other.⁴ Histologically, VRMS have a collagenous appearance while SMS are poorly differentiated and highly cellular. Both are characterized by invasion of surrounding tissues, nuclear pleomorphism, necrosis and mitotic activity.² Prognosis is poor with multiple local recurrences and eventually

pulmonary metastasis.^{2,8} VRMS. Note the central distribution of tumors. The image on the right shows the primary site of tumors in patients with SMS. The tumors occur in all parts of the body. In addition to the above mentioned, differential diagnosis of a retroperitoneal schwannoma also includes paraganglioma, pheochromocytoma, liposarcoma, and fibrous tumors. Hematoma and lymphangioma can be among the differentials as well if cystic degeneration is appreciated on imaging.⁹ It is of great importance to mention that malignant schwannomas must be included in the differential diagnosis in a patient with a history of radiation exposure.⁸ This is explained in the literature by the cytoplasmic vacuolization cells undergo after exposure to radiation, followed by cell death. While the remaining surviving cells undergo genetic mutations which leads to the malignant transformation.¹⁰ It is also noteworthy to mention that Saito et. al have reported a case of benign schwannoma in the retroperitoneal space that had metastasized to the liver. Therefore, careful follow up is highly recommended after the excision of retroperitoneal schwannomas, including the benign variant.¹¹

CONCLUSION

Retroperitoneal schwannomas are rare and are detected incidentally unless they are large enough to be palpated or cause symptoms. Treatment is by resection and prognosis is excellent in cases of benign schwannomas, while malignant schwannomas carry a poor prognosis especially those associated with von Recklinghausen's disease.

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