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Retroperitoneal Schwannoma: A Rare Presentation

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ABSTRACT

The Schwannoma is nerve sheath tumor originating from Schwann cell of peripheral nerve and is commonly found in head,neck and flexor surface of extremities. Only few cases have been reported to be found in retroperitoneum. Here, we describe a 21 year old female lady who presented to our centre with lump and pain in right iliac fossa and was found to have Schwannoma by CT scan and Fine needle aspiration cytology.She underwent laparotomy and excision of tumor andwas confirmedto have Schwannoma on histopathological examination.

Keywords: neural sheat; retroperitoneal; schwannoma.

INTRODUCTION

Schwannoma are neural sheath tumor that originate from Schwann cells of peripheral nerve fibre and are usually found in the head, neck and flexor surface of extremities.[1]The majority of them are benign with few cases of malignant transformation. Schwannoma are associated with von Recklinghausen's disease in 5% to 8% of cases.[2] In absence of von Recklinghausen's disease,retroperitoneal cavity is rare location for schwannoma.Only 0.7% of benign and 1.7% of malignant schwannoma are reported to be found in retroperitoneal area.[1,3]

Here we describe a retroperitoneal schwannoma detected in a women presented to our centre with complaint of pain and swelling in right iliac fossa.

CASE REPORT

A 21 year female was admitted to our centre with complaints of swelling in right iliac fossa for 1 month and abdominal pain in right iliac fossa for 5 days.There was no history of trauma, melena, hematemesis and weight loss.There was no history of consumption of alcohol and smoking.Physcial examination revealed 4cm by 3 cm tender,firm and immobile retroperitoneal lump in right iliac fossa.

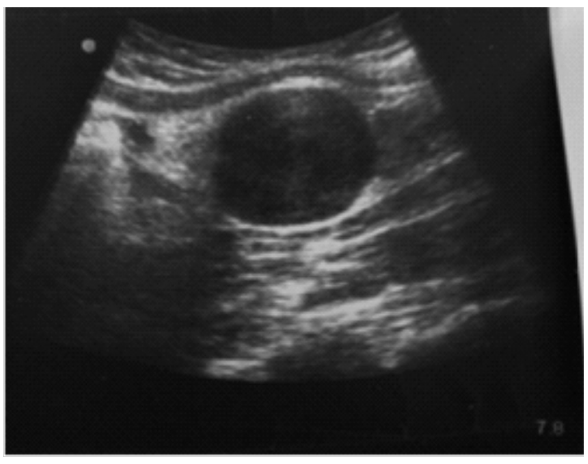

Figure 1. USG pelvis showing hypoechoic mass in right paraumblical area.

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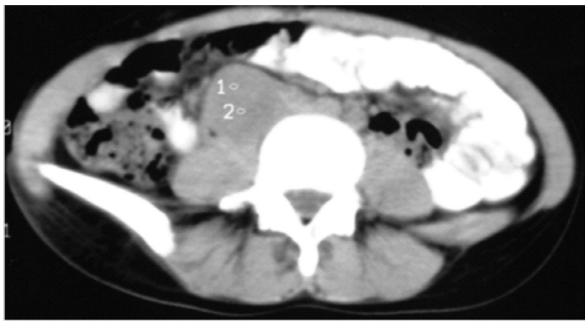

Figure 2. Plain CT pelvis showing homogenous mass at right paraspinal region, displacing midureter anteriorly.

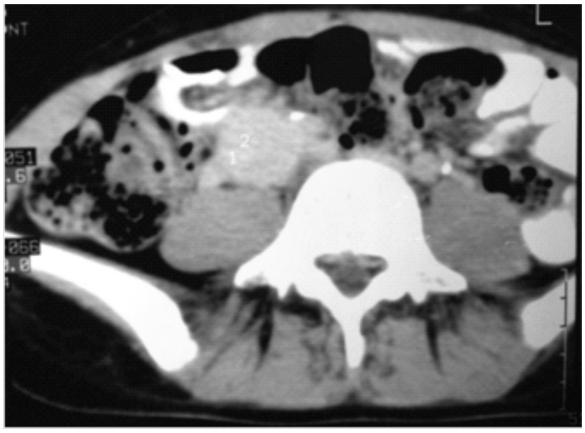

Figure 3. Contrast CT pelvis showing homogenous enhancement

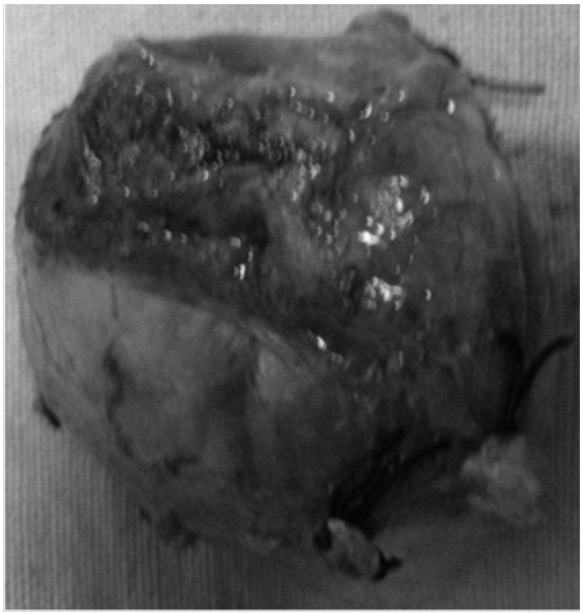

Figure 4. Resected specimen measuring 4x4 cm in size

Figure 5.Resected specimen with cut surface

Biochemical parameter revealed normal findings. Pelvic USG revealed well defined hypoechoic mass measuring 33.7 mmx40.2 mm in right paraumblical area lateral to common iliac vessels suggestive of lymphadenopathy with differential diagnosis of hemorrhagic cyst (Figure1). The CT scan of abdomen and pelvis was done which revealed well defined enhancing soft tissue density mass lesion measuring 41x40x31 mm at right paraspinal region likely to be benign lesion with differential diagnosis of GIST and neurogenic tumor (Figure 2, 3).

Fine Needle Aspiration Cytology from mass was done which showed benign spindle shaped tumor likely to be schwannoma.

Finally, the abdomen was explored with lower midline incision.There was 4x4 cm hard mass at fourth lumbar vertebral level pushing the iliac vessel and ureter medially which was excised and found to be hard and whitish in color on cut surface (Figure 4,5). Histopathology examination revealed encapsulated mass showing hypercellular Antoni A and hypocellular Antoni B suggestive to be benign schwannoma.

DISCUSSION

Schwannomas are nerve sheath tumors usually seen in adult population between 20 to 50 years of age. They are usually benign and affect head, neck and the flexor surface. The retroperitoneal area is rare location for schwannoma except in patients with Von Recklinghausen's disease.[1,2]

The patients with retroperitoneal Schwannoma often present late since the retroperitoneal space is large and

flexible. The most common symptoms are abdominal pain and distention.[4]

MRI is diagnostic modality of choice in evaluation of retroperitoneal tumors.It gives information about the origin, extent and internal composition of the lesion. The typical "target" and "fascicular" sign seen in schwnnoma are not found in case of retroperitoneal schwannoma which impose diagnostic challenge. The "target sign" is hypo -intense centre with hyper-intense periphery while 'fascicular sign" is presence of fascicular bundles.[5,6] Another important feature is destruction of adjacent bony structures. CT and Ultrasonography are used to guide tissue biopsy, monitor treatment and see features of malignant transformation.[7] Irregular margin and infiltrates in adjacent structures are seen in case of malignant transformation.[1]

Tissue diagnosis is last resort for diagnosis which can be achieved via FNAC or excisional biopsy. Schwannomas have true capsule derived from epineurium. Calcification and cystic degeneration is seen in 23% and 66 % of retroperitoneal schwannoma.[4] Microscopically, it consist of Antoni A areas-densely cellular, arranged in short bundles or interlacing fascicles and Antoni B areas-fewer cells,organized with great myxoid component.[5,8] Infiltrative margin with nuclear palisading is striking feature in case of malignant transformation.[9]

Surgery is treatment of choice for retroperitoneal schwannoma. Local resection can be done to remove tumor but current approach is endoscopic assisted minilaparotmy. Chemotherpay and radiotherapy have limited role excepted for marginal benefit in case of malignant shcwannoma.[1]

Metastasis have been reported after local resection, so followup is important.

A retroperitoneal schwannoma is rare tumorusually benign in nature.The tumor presents with vague symptoms and signs. So, the aid of radiology is important is diagnosis and monitoring of patient but the tissue diagnosis is last resort. It is important to rule out malignant transformation since it carries poor prognosis. Surgery is treatment of choice with limited role of chemotherapy and radiotherapy. Regular follow up after surgery is important to rule out metastasis.

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