Research Paper

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Neurilemmoma – A Rare Presentation as a retrperitoneal cyst

Medical Science

KEYWORDS :

MOHAMMED RAZA	Professor,Department of General Surgery JSS Medical college,Mysore.
SHIVANAND REDDY K V	Post Graduates Department of General Surgery,JSS Medical College,Mysore.
DILEEP M	Post Graduates Department of General Surgery,JSS Medical College,Mysore.
MOHINDER VISHWANATH	Post Graduates Department of General Surgery,JSS Medical College,Mysore.
LALIT KUMAR V	Post Graduates Department of General Surgery,JSS Medical College,Mysore.

ABSTRACT

Neurilemmomas are benign, encapsulated tumors of the nerve sheath. Their cells of origin are thought to be Schwann cells derived from the neural crest. These masses usually arise from the side of a nerve, are well encapsulated, and have a unique histologic pattern.Cystic lesions of the retroperitoneum can be classified as either neoplastic or non-neoplastic.

Neoplastic lesions include cystic lymphangioma, mucinous cystadenoma, cystic teratoma, cystic mesothelioma, müllerian cyst, epidermoid cyst, tailgut cyst, bronchogenic cyst, cystic change in solid neoplasms, pseudomyxomaretroperitonei, and perianal mucinous carcinoma. Nonneoplastic lesions include pancreatic pseudocyst, nonpancreaticpseudocyst, lymphocele, urinoma, and hematoma.

Because the clinical implications of and therapeutic strategies for retroperitoneal cystic masses vary depending on the cause, the ability to noninvasively differentiate between masses is important. Although there is substantial overlap of computed tomographic (CT) findings in various retroperitoneal cysts, some CT features, along with clinical characteristics, may suggest a specific diagnosis. CT may provide important information regarding lesion location, size, and shape; the presence and thickness of a wall; the presence of septa, calcifications, or fat; and involvement of adjacent structures. The most important clinical parameters include patient gender, age, symptoms, and clinical history. Nuerilemmomas presenting as retroperitoneal cystic mass is extremely rare with incidence being 0.2 to 1 %.

Pathophysiology:

Their cells of origin are thought to be Schwann cells derived from the neural crest. These masses usually arise from the side of a nerve, are well encapsulated, and have a unique histologic pattern.The benign lesion essentially manifests itself with cosmetic deformity, a palpable mass, and/or symptoms similar to a compressive neuropathy. Neurologic symptoms tend to present late. Symptoms can be vague, and there is an average interval of up to 5 years before the diagnosis is established. Neurilemmoma is the most common neurogenic tumor.

Diagnosis:

No racial or sex predilection is recognized. Neurilemmomas affect persons aged 20-50 years. Common locations for the tumors are, in order of decreasing frequency, the head and flexor surfaces of the upper and lower extremities and the trunk. The mass is usually mobile in the transverse plane and tethered along the axis of the nerve from which it arises.

Tenderness to palpation is often present; secondary neurologic symptoms may occur if the tumor is large. When involving the C7 nerve root, neurilemmoma has been described as a cause of thoracic outlet syndrome. Lesions in the sciatic nerve can mimic discogenic low-back pain.

Because these tumors can present in many locations, the clinical presentation can be varied.[5] Some may involve the spinal nerve roots and present with symptoms that mimic those of herniated disk disease of the spine.[6, 7] In the extremities, neurilemmomas can present either as an asymptomatic mass or as mild, localized pain and paresthesia resulting from pressure on the nerve of origin. Masses are slow growing and can exist for months to years without producing symptoms. The average time from onset of symptoms to diagnosis is 5.5 years.

Lesions in proximal nerves may cause distal symptoms. If these masses occur in well-defined compartments (eg, wrist, ankle), they can present as either carpal tunnel or tarsal tunnel syndrome.

Case report:

A 34 yr old female patient presented to the JSS ER department on 10th of april 2014 with complaints of mass in the right lower abdomen associated with pain since 2 months.Mass of initially of the size of 4 × 4 cm which has presently increased to the size of 8 × 8 cm over two months period.Patient had previously shown to the local doctor for similar complaints but patient was treated symptomatically and patient didn't follow up for the condition.

Patient is P_2L_2 underwent caesarean section for second delivery 7 years back and tubectomy 4 years ago. Patient was admitted and routine investigations were done.

Patients HB was 12.6 gm and all other routine investigations were in the normal range.

Patient was advised **USG abdomen and pelvis** which revealed a well circumscribed lesion in the right iliac fossa ?ovarian cyst. **Usg guided FNAC** was done which showed features of only haemorrhage,no malignant cell were seen.

CT-SCAN abdomen was planned which revealed a large well defined complex cystic lesion arising in relation to right psoas muscle measuring approximately 27×31 cm .Multiple thickened septations noted within with few areas of calcification.Left common iliac artery is displaced and abutting the medial aspect. Right ovary was seen separately from the lesion in right adnexa .Left ovary is normal.

IMPRESSION:

? Retroperitoneal complex cyst

? Hydatid cyst

Patient was planned for laparotomy on 13 thapril after consent and fitness for surgery.

Per-op findings :

A 15 × 15 cm cystic mass was found in the right iliac fossa retroperitoneally very close to the right iliac vessels and the ureter and

the posterior surface was difficult to approach,but plane was created and dissected neatly and mass was separated from the vessels and there was no involvement of underlying muscle and ovaries.

There was not much of bleeding and mass of removed in toto. Retroperitoneum was closed and a drain was placed and abdomen wasclosed.Specimen was sent for histo-pathological examination.Drain was removed on 2nd post op day and patient recovered well post-operatively.

HISTOPATHOLOGY:
Sections showed tumor tissue with features of a**Neurilemmoma with secondary changes.**

DISCUSSION:
Retroperitoneal cysts are very rare, and most of the time they are discovered incidentally. Patients may be asymptomatic or present with abdominal pain, referred pain to the legs or weight loss. Imaging may help diagnose these lesions, but surgery is the keystone in confirming the diagnosis. This case is very rare and very educational as it highlights an unusual presentation of a benign retroperitoneal cyst. In our patient, the course of the disease was unique as there was no history of weight loss,rapid increase in size but later it turned out to be neurilemmoma which is extremely rare presentation as retroperitoneal cyst as incidence being 0.2 to 1%. This case report educates us about rare presentation of a rare disease.

PICTURES:

Fig 1: Clinical presentation of the patient.

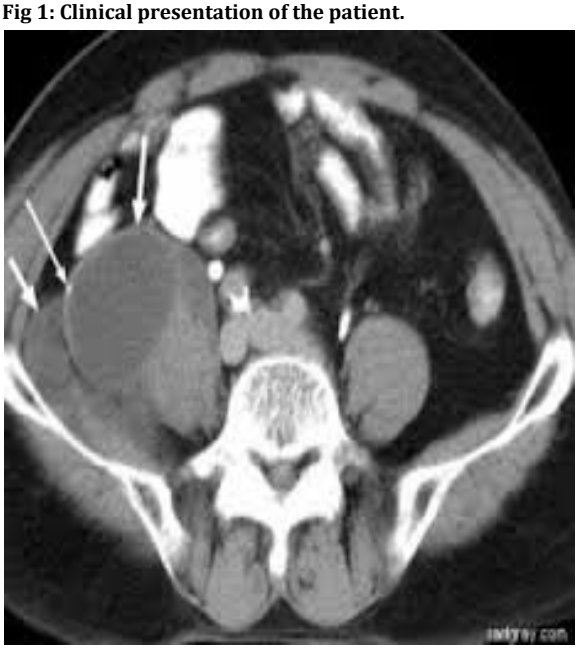


Fig 2 : CT –Abdomen showing a retroperitoneal mass in the right iliac fossa.

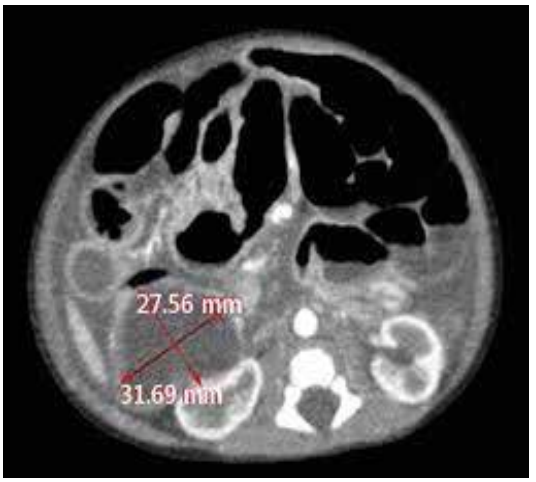


Fig 3 : CT film showing the dimensions of the mass.

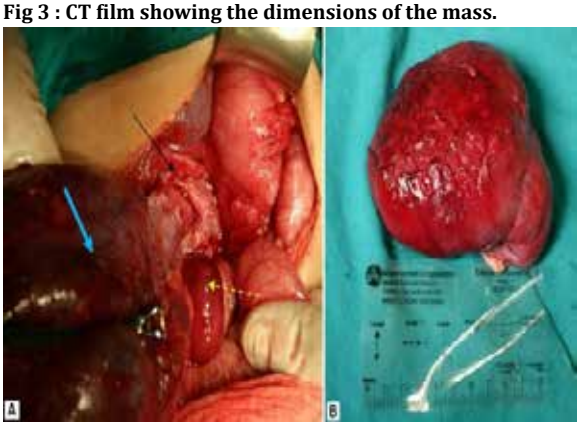


Fig 4A: Showing a retroperitoneal cyst with capsule.
Fig 4B: Showing specimen in toto.

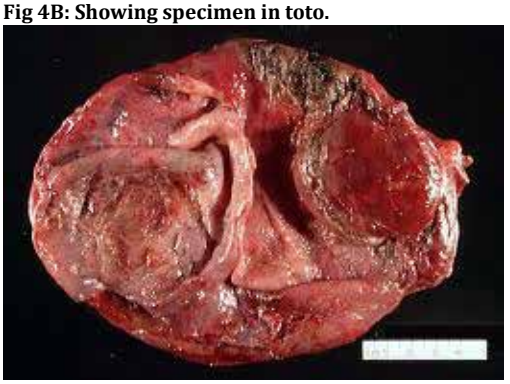
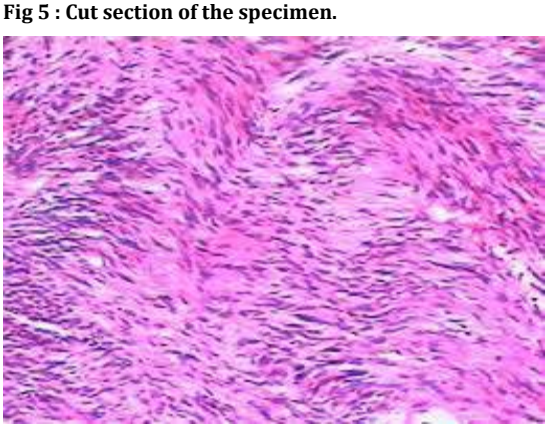


Fig 5 : Cut section of the specimen.



- **Fig 6: Histopathological slide showing cells which are narrow, elongate, wavy with tapered ends interspersed with collagen fibers suggestive of neurilemmoma.**

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REFERENCE

1. Guile M, Fagan M, Simopolous A, Ellerkman M: Retroperitoneal Cyst of Mullerian Origin: A case report and review of the literature. J of Pelvic Medicine and Surgery 2007,13(3):149–152. Handfield-Jones R: Retroperitoneal Cysts: Their Pathology, Diagnosis and Treatment. BJS 1942, 119–134. | 2. Felix EdwardL, Wood DonaldK, Das Gupta TapasK: Tumours of the Retroperitoneun. Cancer 1981,6(1):1–47. | 3. Yang D, Jung D, Kim H, Kang H, Kim S, Kim J, Hwang H: Retroperitoneal cystic masses: CT, Clinical and Pathological Findings and Literature Review. RG 2004,25(5):1353–1365. | 4. Kurtz R, Heimann T, Beck R, Holt J: Mesenteric and Retroperitoneal Cysts. Ann Surg 1986,203(1):109–112. | 5. HaysakaKazumasa, Yamada Tomonori, Saitoh Yasuhiro, Yoshikawa Daihei, et al.: CT Evaluation of Primary Benign Retropeitoneal Tumour. Diagnostic Radiology 1994,12(3):115–120. | 6. Konishi Eiichi, Nakashima Yasuaki, Iwasaki Takeki: Immunohistochemical analysis of Retroperitoneal Mullerian Cyst. Human Pathology2003,34(2):194–198. | 7. Ravo B, Metwally N, Pai B, Ger R: Developmental retroperitoneal Cysts of the Pelvis; A Review. Dis Col &Rect 1987,30(7):559–564. |