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RESEARCH ARTICLE

AN UNCOMMON CASE OF TWIN RETROPERITONEAL LUMPS: A CASE REPORT

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ABSTRACT

A retroperitoneal tumors is uncommon and neurofibromas still rarer. Solitary neurofibromas are frequently detected in the skin, appendix, stomach and jejunum. However, it has rarely been reported as occurring in the retroperitoneum. Most NST's are small solitary and benign and rarely exceed 6 cm in diameter. Symptoms do not appear until they have attained gigantic dimension or start compressing surrounding structure. A 17yr old male patient presented with a painless lump in the left lumbar region for last 6 months. A lump is palpable in the left lumbar region of about (10x8) cm. The lump is retroperitoneal in type and not mobile. Another lump of about (5x3) cm. is palpable in the left hypochondriac region. This lump is also not mobile and retroperitoneal in type. Laparotomy followed by Excision of the lumps done. Histopathology shows Neurofibroma; diffuse type.

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INTRODUCTION

Nerve sheath tumours (NST) are a subclass of soft tissue neoplasms that include benign and malignant schwannomas and neurofibromas. There are two types of neurofibromas, localized and diffuse; the latter is associated with von Recklinghausen disease and always occurs together with skin neurofibromas. Presentation as retroperitoneal tumors is uncommon and neurofibromas still rarer. Solitary neurofibromas are frequently detected in the skin, appendix, stomach and jejunum (Rutgeerts et al., 1981). However, it has rarely been reported as occurring in the retroperitoneum (Ladouce, 1978; Takahashi, 1977). Most NST's are small solitary and benign and rarely exceed 6 cm in diameter (Stout, 1935). Symptoms do not appear until they have attained gigantic dimension or start compressing surrounding structure.

CASE REPORT

HISTORY: A 17 yr old Indian male patient presented with a lump in the left lumbar region for last 6 months.

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The lump is insidious in onset, gradually increasing in size and now reached to a size of about (8x6) cm. There is no history of any pain abdomen or urinary symptoms. There is no history of any systemic symptoms and fever. Family history was negative for Von Recklinghausen's disease. His medical history did not reveal any diseases. There is no history of any addiction or allergy.

PHYSICAL EXAMINATION: General survey is essentially normal. On local examination of abdomen, on inspection, the shape of the abdomen is normal. The umbilicus is in the midline and inverted. All quadrants of abdomen are moving simultaneously with respiration. All hernia sites are normal along with the external genitalia. On palpation, a lump is palpable in the left lumbar region of about (10x8) cm., globular in shape, smooth surface, hard in consistency, ill defined margin, not moving with respiration and the lump disappears with head/leg raising. Another lump of about (5x3) cm. is palpable in the left hypochondrium with similar characteristics. Liver and spleen are not palpable. The abdomen is tympanitic on percussion. All systemic examination is normal.

INVESTIGATIONS: Ultrasonography reveals, two well defined heterogeneously hypoechoic mass lesion measuring about (80x69) mm and (47x46)mm found in left side of upper retroperitoneum.



Image 1. Ultrasonographic feature of retroperitoneal SOLs

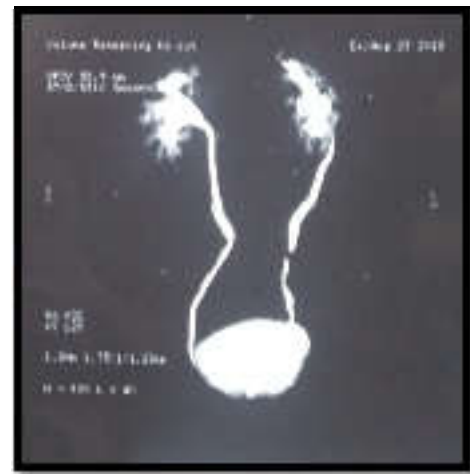
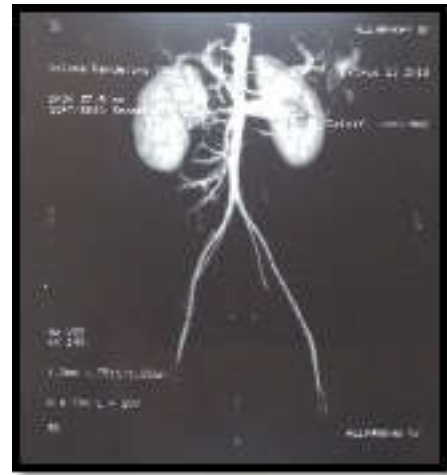


Image 4: CT Urogram Plates

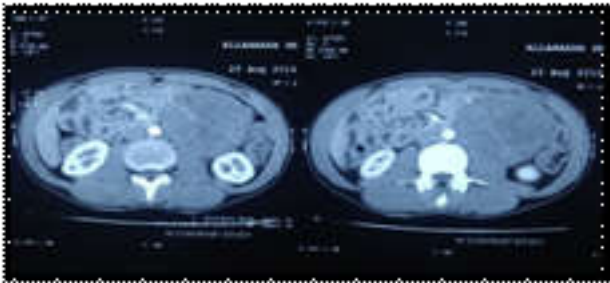


Image 2: CECT plates showing SOLs (Space Occupying Lesions)

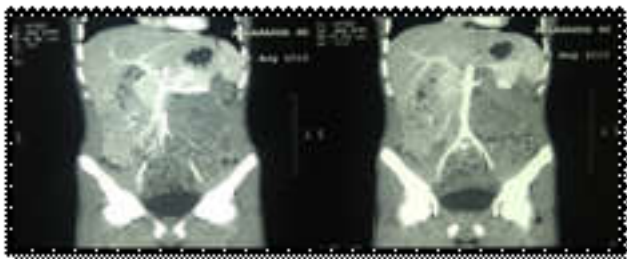
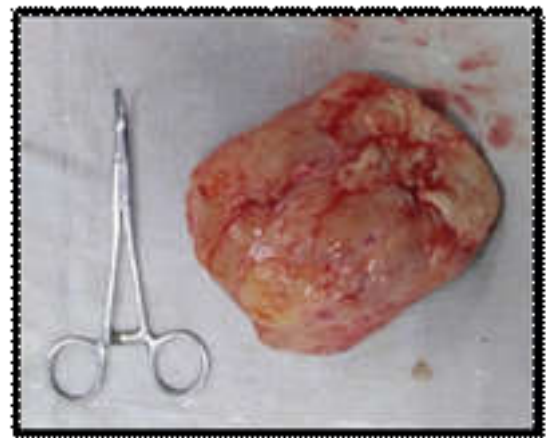


Image 3: CT Angio plates



The superior one is seen posterior to left renal vessels. On doppler lesions show internal flow. Contrast Enhanced CT (CECT) Scan of abdomen shows, heterogeneously enhancing retroperitoneal mass lesions on left side with indistinct fat planes with adjacent posterior diaphragm (upper mass) and left psoas muscle (lower mass) with mass effects and left mild hydronephrosis. No evidence of calcification or fat seen within the lesions.



Image 5: Resected retroperitoneal SOLs

PROCEDURE: At the intra-abdominal exploration, the lesion appeared displace the left kidney, the abdominal aorta and the left ureter. These structures were identified and preserved after a sharp dissection. The specimen was extracted with en-bloc resection.

HISTOPATHOLOGY: Section shows an ill defined poorly circumscribed tumor mainly composed of fascicles, bundles of spindle nerve fibres having wavy nuclei, fine chromatin and inconspicuous nucleoli. Pleomorphism and mitotic activities not found. The tumors are infiltrated with mixed inflammatory cells.

Suggestive of *Neurofibroma; diffuse type*.

DISCUSSION

Nerve sheath tumours contribute about 4% of retroperitoneal tumors⁽⁵⁾. Paraaortic-Pararenal Neurofibroma is an exceedingly rare tumour location (Kostakopoulos, 2003). Diagnosis of a retroperitoneal neurofibroma is difficult to establish preoperatively as neuroimaging is not specific. Complete surgical excision is the treatment of choice for retroperitoneal nerve sheath tumors. Residual disease might lead to recurrence of tumor and its symptoms or malignant transformation (Hurley, 1994). Recurrence rate in wide excision of tumor is 11.7% as compared to 72% in only margin excision (Bose, 1979).

Conclusion

Retroperitoneal neurofibroma is a histopathological diagnosis. Because of its varied imaging appearance, interpretation of imaging results is often difficult. Differentiating between benign and malignant disease radiologically too is difficult. It is imperative that extensive surgery for a benign disease is avoided and radical surgery for a malignant disease is undertaken.

Conflict of interest: None

Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images

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