Original Research Paper

VOLUME-6 | ISSUE-4 | APRIL - 2017 • ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46

RETROPERITONEAL PARARENAL NEUROFIBROMA: A CASE REPORT

Urology

KEYWORDS: neurofibroma; retroperitoneal

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ABSTRACT

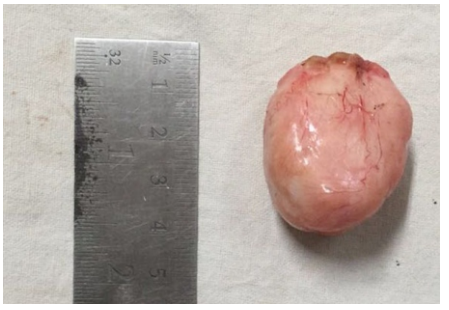
Neurofibroma is a tumour of neural origin. This kind of neoplasm, though, is generally skin located but rare cases in deep organs or in the peritoneal cavity are also reported in the literature. There are two types of neurofibromas, localized and diffuse; the latter is associated with von Recklinghausen disease and always occurs together with skin neurofibromas. Here we report the case of a 58 year old female affected by a solitary retroperitoneal neurofibroma, but not associated with von Recklinghausen disease. A MRI abdomen scan described a retroperitoneal Lt pararenal lesion with no clear involvement of adjacent viscera and suggested possibility of Non functioning phaeochromocytoma /extra adrenal paraganglioma/ spleniculus. Tumor was completely excised and histopathology revealed Neurofibroma.

INTRODUCTION: Nerve sheath tumours (NST) are a subclass of soft tissue neoplasms that include benign and malignant schwannomas and neurofibromas. Presentation as retroperitoneal tumors is uncommon and neurofibromas still rarer. Solitary neurofibroma without von Recklinghausen's disease is the rarest presentation of all neurofibromas. Most nerve sheath tumors are small solitary and benign and rarely exceed 6 cm in diameter(1).Symptoms do not appear until they have attained gigantic dimension or start compressing surrounding structure. We describe a case of solitary neurofibroma which was diagnosed by histopathology after complete excision.

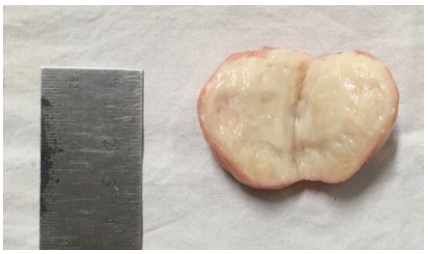
CASE REPORT: A 58 year old Indian female presented with complaint of vague abdominal pain for one month. Pain was of moderate intensity, localised to left flank and episodic in nature. She was diabetic and hypertensive since 2 years and was on medication. Family history was negative for Von Recklinghausen's disease. Physical examination did not contribute much. Haematological and biochemical investigations were normal. Ultrasound showed a mass lesion of size 37 x 31 mm, hypoechoic lesion in upper pole of left kidney suggestive of adrenal adenoma / neurogenic tumour/ spleniculus. A diagnosis of adrenal adenoma was considered. MRI abdomen demonstratedwell defined soft tissue lesion seen lateral to left adrenal gland and anterior to upper pole of left kidney, lesion measures 33 x 32 x 31 mm suggestive of Non functioning phaeochromocytoma /extra adrenal paraganglioma/ spleniculus. Laparoscopic excision of the mass was done. Well circumscribed mass measuring 3.5 x 3 cm in retroperitoneum close to upper pole of left kidney and pancreas, but separate from both these organs was noted. Tumour was resected in its entirety. Cut surface was yellowish brown with solid consistency. On histopathological examination it revealed benign retroperitoneal neurofibroma. Postoperative period was uneventful.

DISCUSSION: Nerve sheath tumours contribute about 4% of retroperitoneal tumors(2).Neurofibromas are often multiple, plexiform and found in association with Von Recklinghausen's disease . It is an autosomal dominant disorder with multiple neurofibromas and schwannomas, along with cafe au lait spots and axillary freckling. In absence of this disease,neurofibromas are solitary and don't turn malignant even though 4-11% of neurofibroma associated with Von Recklinghausen's disease do undergo malignant degeneration(3). A solitary neurofibroma represents either a sporadic case or a carrier of defective gene with only mild clinical presentation(4).Therefore all patients should be examined for stigmata of neurofibromatosis. Solitary neurofibromas are slow growing tumours which lack a well-defined capsule and an identifiable parent nerve. These tumours generally have equal gender distribution and present between 3rd and 6th decade of life(5). Clinically they present with symptoms of compression rather than invasion. They can present as asymptomatic abdominal mass or can cause pain which can be colicky in case of obstruction of bowel, ureter, biliary system or neuralgia due to involvement of nerve and bone. Gastrointestinal symptoms like abdominal discomfort, nausea, vomiting, constipation,obstipation or neurological symptoms like neuralgic pain, hypoesthesias and paraesthesias can also be seen. Diagnosis of a retroperitoneal neurofibroma is difficult to establish preoperatively asneuroimaging is not specific. On CT,neurofibromas are smooth, rounded, homogeneously hypoattenuating masses. MRI shows a target like enhancement pattern with attenuation in the centre known as the central dot sign. Dot sign corresponds to edema, microcysts, foam cells, hyalinization of blood vessels, old hemorrhage and dystrophic calcification(6,7). Diagnosis can be confirmed only on histopathology. CT or USG guided aspiration biopsy is the only way to confirm diagnosis preoperatively. It is not advisable though, because aspirate usually consists of highly pleomorphic cells which are difficult to interpret and procedure itself is associated with complications like infection and haemorrhage .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(8).Surgical resection is challenging because sometimes there is nerve entrapment by the tumor and it becomes impossible to remove tumor without sacrificing the nerve. In case of huge masses, if malignancy cannot be ruled out, enbloc excision of margins or organs around the tumor should be done. Recurrence rate in wide excision of tumor is 11.7% as compared to 72% in only margin excision(9).

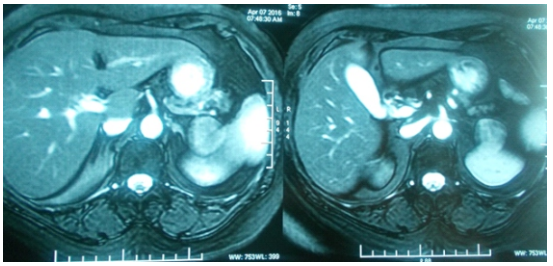
CONCLUSION: Retroperitoneal pararenal neurofibroma is a histopathological diagnosis. Because of its varied imaging appearance, interpretation of imaging results is often difficult. Differentiating between benign and malignant disease radiologically too is difficult. It is imperative that extensive surgery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

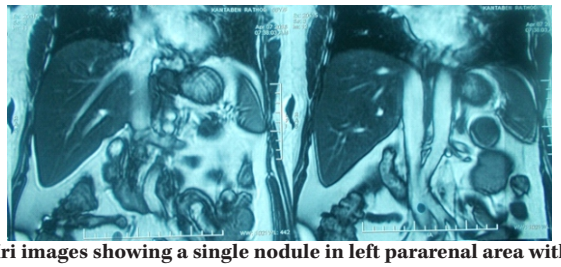


single nodule of size 4x3x2 cm with smooth surface and light pink in colour

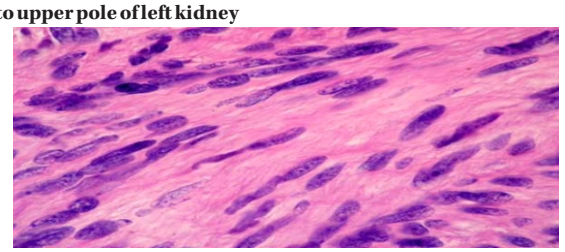


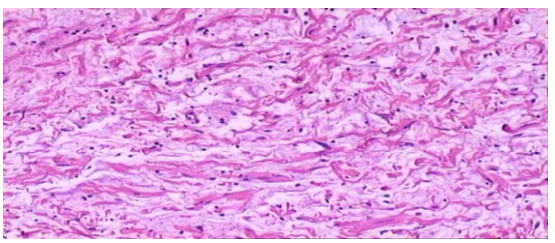
on cut section the nodule is tan yellow in colour with fibrous hard consistency



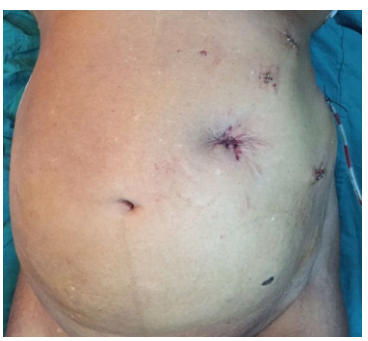


Mri images showing a single nodule in left pararenal area with size approx. 4x3x2 cm lateral to left adrenal gland and anterior to upper pole of left kidney





HPE- schwann cells seen as intersecting bundles of spindle with hperchromatic wavy nuclei. Wire like strands of collagen seen . background stroma contains lymphocytes and xanthoma cells



Post operative picture

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