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Case Report

A case of retroperitoneal schwannoma

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ABSTRACT

Schwannoma are benign tumours of nerve sheath. They are also called as neurilemmoma. They are rare tumours arising from peripheral nerves, though they can occur anywhere on the body. Schwannoma can arise from any peripheral nerves or spinal nerves. Since they arise from nerve sheath forming schwannoma cells, they are never seen in brain or spinal cord. Schwannoma are often associated with neurofibromatosis-2, schwannomatosis and Carney complex where it is inherited in an autosomal dominant manner. They care often asymptomatic and diagnosed incidentally. Symptoms of schwannoma are related to pressure effects over the surrounding structures, such as limb pain, tingling and numbness, deafness and facial weakness as in vestibular schwannoma. However, most schwannoma are detected incidentally as in the case reported below. A 48 years female was diagnosed with an asymptomatic left para renal lump, which was later confirmed to be a retroperitoneal schwannoma. The patient had no complaints with respect to the lump. The patient underwent a successful surgical excision and had an uneventful recovery.

Keywords: Schwannoma, Retroperitoneum, Neurofibromatosis, Carney complex, Nerve sheath tumour

INTRODUCTION

Schwannoma is a benign tumour arising from schwann cells.[1] Schwann cells form a covering over the peripheral nerves. Schwannomas can arise anywhere in the body. The most common site is the inner ear, where they arise from the vestibular nerve, more commonly when associated with neurofibromatosis-2. These are often bilateral. Malignant schwannoma, though rarer, are seen arising from sciatic nerve, brachial plexus and sacral plexus. Of all the schwannoma, only about 0.5% are seen in the retroperitoneum. Majority of the retroperitoneal schwannomas are benign, though malignant ones are also reported. Malignant transformation of benign tumour is commonly seen in neurofibromatosis-2. They are often diagnosed incidentally during routine check-up or imaging done for apparently unrelated symptom. They lack specific diagnostic sign on ultrasound scan, computed tomography or magnetic resonance imaging, thus making the diagnosis difficult.[2] Treatment of benign schwannomas is complete surgical excision. Schwannomas are resistant to chemotherapy and radiotherapy. Recurrences are rare but need close follow up after surgical excision.

CASE REPORT

A 48 years old female undergoing an abdominal ultrasound for complaint of menorrhagia was diagnosed to have a large retroperitoneal mass from the left kidney. The patient was referred to our centre for further management. Patient had no history of abdominal pain, abdominal distension, constipation, diarrhoea, weight loss, difficulty in micturition, blood in urine, per rectal bleed or icterus. The patient had no past history of an abdominal surgery, general physical examination was unremarkable. Per abdominal palpation revealed an ill-defined lump in left hypochondrium. It was non-tender,

smooth, non-ballotable and finger could not be insinuated between the lump and the left coastal margin. A contrast enhanced computed tomography of abdomen and pelvis showed a well-defined heterogeneously echotextured lesion with solid and cystic components in left paravertebral region at the level of D-12 to L-4 vertebrae. The tumour was seen separately from the left kidney and ureter. It had minimal internal vascularity, suggesting a benign retroperitoneal tumour, a schwannoma more likely. A US guided fine needle aspiration also confirmed the same.

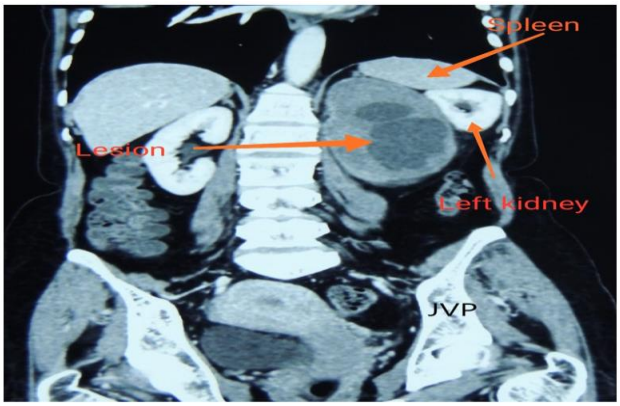

Figure 1: CT image of the lesion.

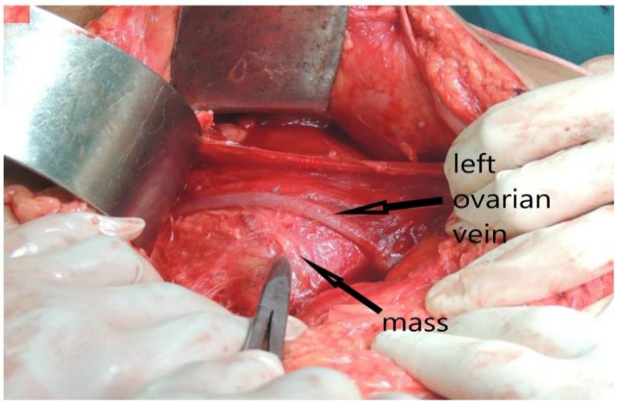

Figure 2: Intraoperative image of the mass.

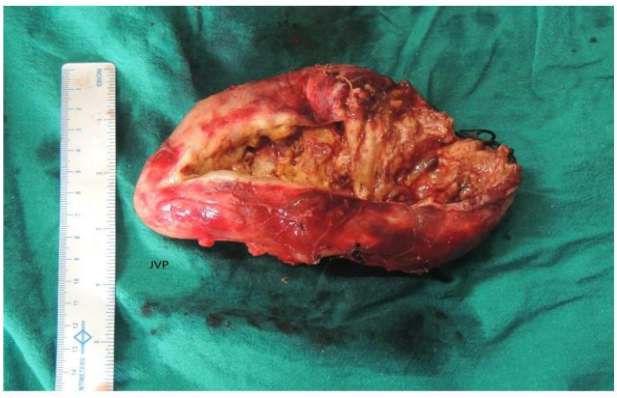

Figure 3: Specimen with cut section showing internal necrosis.

A planned laparotomy was done with intra operative frozen section suggesting a low grade fibromyxoid sarcoma. A complete surgical resection was done. Nephrectomy or bowel resection were not performed. A final biopsy report was confirmative of benign schwannoma. Patient had an uneventful post-operative period and was discharged after 7 days.

DISCUSSION

Schwannoma are predominantly benign tumors arising from Schwan cells which form the covering of neurons. Thus, they can be found in any part of the body or organ. They are usually seen in the age group of (20-50) years. Symptoms due to the tumor are mostly related to pressure effects in the part they arise from. Retroperitoneal location of these tumors is quite rare and attain a fairly large size before being noticed. Malignant schwannomas are extremely rare and most often associated with von Recklinghausen's disease.[3] The most common symptoms include abdominal distension and pain. They are well encapsulated and often undergo cystic degeneration. Pre operatively retroperitoneal schwannomas can be difficult to diagnose and high degree of suspicion is needed. Imaging done for these tumors can be supplementary but not confirmatory. The target sign is seen on MRI, where a hyper intense periphery is seen around a hypo intense centre. The fascicular sign seen, though nonspecific, shows bundle like arrangement of tumor. Characteristic histopathological characters include spindle shape cells with the myxoid background with Antoni A areas of dense cellular infiltration and Antoni B areas with few cells and more abundant myxoid cytoplasm.[4] On immunohistochemical staining, schwannoma show positive expression of S-100 and negative of CD 34.[5] Differential diagnoses of retroperitoneal schwannomas include pheochromocytoma, paraganglioma, liposarcoma and malignant fibrous histiocytoma. Local surgical resection is usually sufficient for benign schwannomas, but a long-term follow-up is usually preferred as recurrences are also noted.

CONCLUSION

Thus concluding, it is important to consider retroperitoneal tumors when one has ruled out ones arising from liver, spleen, pancreas and bowel by clinical examination. One should have a suspicious eye regarding retroperitoneal tumors though these are very rare. Schwannomas may not always be associated with defined syndromes and can present sporadically. Tissue diagnosis is important but a surgeon needs to correlate clinically in excluding various differentials. One also needs to remember that most schwannomas can be treated by way less aggressive modes and thus avoid unnecessary fear among the patients.

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