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Large Retroperitoneal Schwannoma- A Rare Case Report

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ABSTRACT

Schwannomas are also called as Neurilemmoma and are benign neoplasm derived from Schwann cells of the peripheral nerve sheath. Occurrences of retroperitoneal schwannomas are uncommon and are presented with non-specific symptoms such as abdominal pain and distension. Radiological and clinical diagnosis of retroperitoneal schwannomas is challenging and therefore surgical excision and histopathological examination are more helpful modalities. We report a case of a 46 year old woman with retroperitoneal mass. This case is reported due to its rare presentation.

Keywords: retroperitoneum, benign schwannoma, Schwann cells.

INTRODUCTION

Schwannoma is a benign tumor that arises from the Schwann cells of the peripheral nerve sheath.[1] It is also known as 'Neurilemmoma'. It is a neurogenic tumor that occurs usually between the 3rd and the 6th decade and has an equal predilection for men and women.[2]Schwannoma is composed of Antoni A that are compact areas of high cellularity and Antoni B areas that are loose hypocellular areas.[3] 1-10% of primary schwannomas occur in the retroperitoneal region and among them 0.7% of benign and 1.7% of malignant nature have been reported till date.[4] Since there are no typical features on radiological imaging to distinguish retroperitoneal schwannomas and since the patient usually presents with non specific symptoms, clinical and radiological diagnosis may not be possible in every case and therefore USG guided FNAC can help to give a preliminary diagnosis. Further surgical excision of the mass with histopathological examination should be done for the confirmation of the diagnosis.[3]

We herein report a case of a 46 years old female diagnosed to have a large benign schwannoma occurring in the retroperitoneal region.

CASE REPORT

A 46 years old female, housewife, came to the surgery OPD with complains of pain in lower abdomen since 5 days. Pain is of moderate intensity. Patient had no history of nausea, vomiting, loss of appetite, loss of weight, burning micturition, fever, diarrhoea and constipation. Patient had surgical history of lower abdomen Caesarean section 20 years back and had got tubectomy done 14 years ago. Abdomen examination revealed a firm immobile mass in the hypogastric region. Other systemic examinations showed no abnormality. Computed tomography of the abdomen and pelvis was done and the imaging findings showed a large well defined heterogeneously enhancing soft tissue density mass lesion of size 13x11x 9.5cm located predominantly in retroperitoneal, presacral space. The findings were suggestive of neurogenic tumor of sacrum (type 2).

USG guided fine needle aspiration from the presacral mass was done. Smears

studied were sparsely cellular and comprised of few clusters of spindle shaped cells with bland elongated nuclei against a hemorrhagic background, suggestive of benign spindle cell lesion. (Figure 1)

The patient was taken up for surgery and the tumor was resected with meticulous dissection of the surrounding structures. Post-operative course was uneventful. The specimen was sent for histopathological examination.

Grossly the specimen received was a globular, encapsulated soft tissue mass that measured 15x11x10cm. External surface was grey white with areas of congestion. Cut surface was grey white with focal yellow areas and a uniloculated cyst that measured 6cm in greatest diameter filled with blood. (Figure 2)

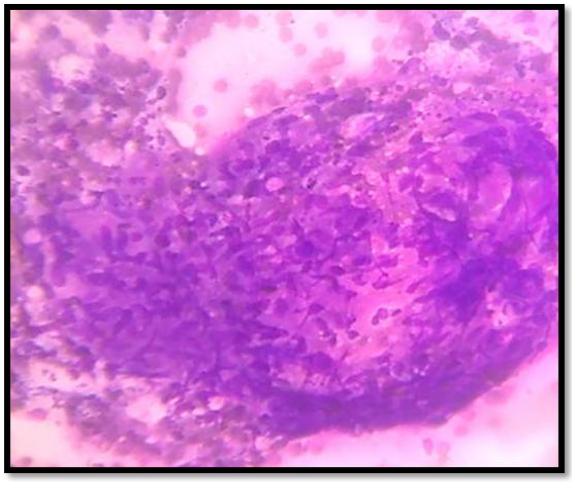

Figure 1: Benign spindle cells noted on smear, (Giemsa, x400)

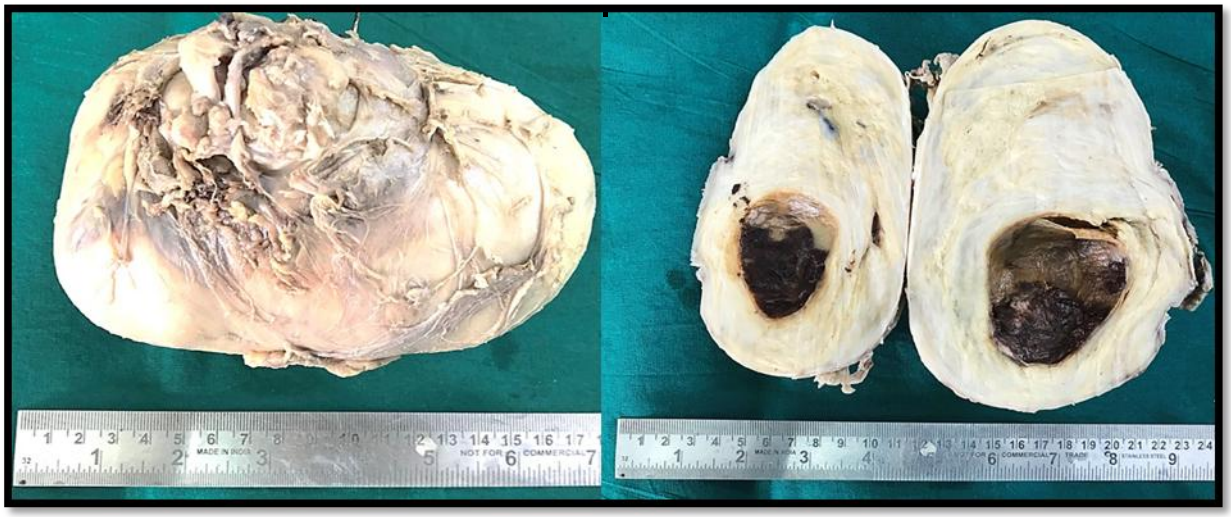

Figure 2: Grossly external surface of the globular mass was capsulated with areas of congestion, cut surface shows a grey white lesion with focal yellow areas and a uniloculated cyst.

Microscopically the sections studied showed a capsulated lesion composed of Antoni A and Antoni B areas in a loose matrix. Antoni A areas were composed of compact benign spindle shaped cells with wavy nuclei arranged in interlacing fascicles and bundles. Verocay bodies were noted. Antoni B areas were less cellular. Mitotic figures were inconspicuous. According to the histopathological examination the diagnosis given was 'Schwannoma'. (Figure 3&4)

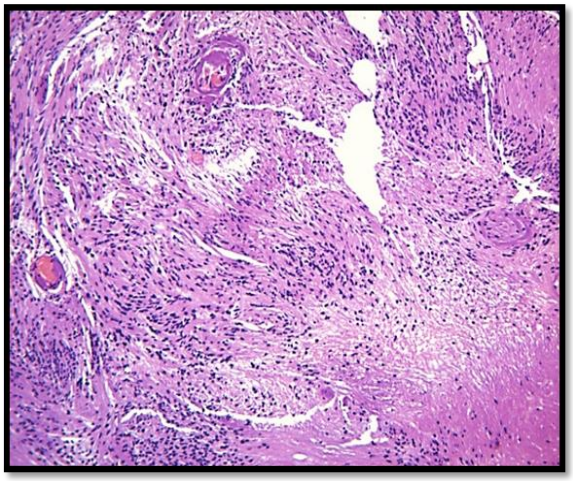

Figure 3: Microphotograph showing spindle shaped cells arranged in interlacing fascicles and bundles. Antony A & Antony B areas, (H&E, 100 x)

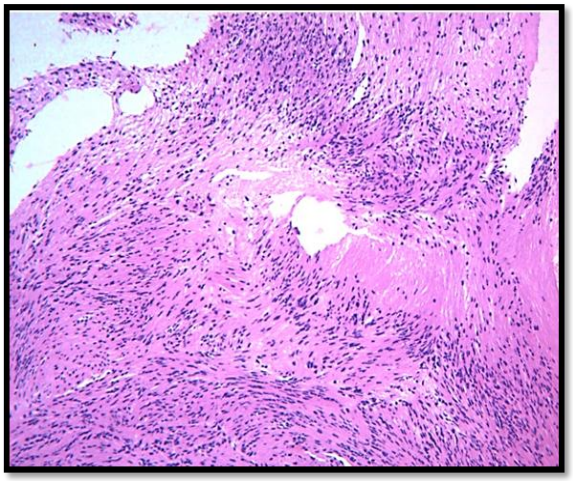

Figure 4: Antoni A areas with Verocay bodies, (H & E x400)

DISCUSSION

Schwannomas are benign, slow growing, solitary and well encapsulated lesion arising from the Schwann cells. Common location where schwannomas usually occur are head and neck, flexor surface of the upper and the lower extremities and posterior mediastinum and trunk.[3] Schwannomas account for 0.5% - 12% of all the retroperitoneal tumors.[5]Retroperitoneal schwannomas are rare and account for 0.3-3.2% of the benign schwannomas.[3]In retroperitoneal region majority of them are seen associated with patients having Von Recklinghausen disease.[2] Diagnosis of the retroperitoneal schwannomas are difficult since they are usually asysmptomatic or they present at a later stage.[3] Most patients with schwannoma are between 25-55 years of age but they can occur at any age and they show no sex predilection.[2] Major clinical symptoms are abdominal distension and abdominal pain.[2]

Pre-operative diagnosis of retroperitoneal schwannomas is difficult even radiologically due to lack of typical imaging features that distinguish schwannomas from other retroperitoneal tumors.[6] Surgical excision is the treatment of choice. Schwannomas respond poorly to chemotherapy and radiotherapy.[2]

Microscopically schwannomas are biphasic and are composed of Antoni A and Antoni B areas. Cystic changes are more common in the retroperitoneal schwannomas than other retroperitoneal tumors.[3] Heterogeneity due to cystic degeneration is termed as 'ancient' schwannoma.[3] Degeneration is caused by central tumor necrosis that occur if the tumor has increased in size beyond the capacity of its blood supply.[3]

Differential diagnosis for retroperitoneal schwannomas are other neurogenic tumors such as paraganglioma and pheochromocytoma, and liposarcoma and malignant fibrous histiocytoma. If the retroperitoneal schwannoma shows cystic degeneration, cystic masses that occur in the retroperitoneal region such as hematoma and lymphangioma should also be considered.[4]

Malignant schwannomas show features like irregular contours and invasion into the adjacent structures radiologically.[4] Prognosis of a benign retroperitoneal schwannoma is generally good but malignant transformation has been noted and therefore careful monitoring and follow up of the patient is a must.

CONCLUSION

Preoperative diagnosis of retroperitoneal schwannoma has been challenging. Surgical excision and histopathological examination is necessary to give an accurate diagnosis since preoperative diagnosis may not be possible each time.

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