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Case Report

A rare case of retroperitoneal tumor diagnosed as adrenal ancient schwannoma

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ABSTRACT

Retroperitoneal ancient schwannoma is a rare condition. Schwannomas are mostly seen arising from head and neck, flexor aspects of extremities, with an incidence 0.7–2.7%. Other sites of schwannoma are rare with few recorded cases. Retroperitoneal schwannomas (RPS) are relatively rare and constitute only 3% of all schwannomas and 4% of retroperitoneal tumours. Diagnosis is mostly made by histopathology and immunohistochemistry (IHC). Here we report a case of 60-year female with retroperitoneal ancient schwannoma, who presented with chronic abdominal pain.

Keywords: Ancient schwannoma, Retroperitoneal, Antoni A and B, S100

INTRODUCTION

Schwannomas are also called as neurilemoma. They are seen arising from Schwann cells that form neural sheath anywhere in the body. Most common location of schwannomas is head and neck region as acoustic schwannoma. Retroperitoneal schwannomas (RPS) most commonly occur in paravertebral and presacral region.[1-3] RPS are mostly asymptomatic unless until they enlarge to a larger size causing pressure symptoms. Adrenal schwannomas are very rare with 80 reported cases till date with incidence of 0.2% of adrenal tumours. Diagnosis is uncommon and based on clinicoanatomical location, pathological and immunohistochemical examination. We here report a case of 60-year-old female was diagnosed with RPS.

CASE REPORT

A 60-year-old female presented to surgical clinic with complaints of vague dull aching abdominal pain in epigastric, umbilical and back region for one week. She has similar episodes in past for a year on and off, with no associated symptoms. On physical examination patient is in good general condition and family history was unremarkable, blood pressure - 140/90 mm Hg, pulse rate - 78 beats/min, blood glucose was within normal range, abdomen was soft, no organomegaly and no cushingoid features. Blood and biochemistry analysis were within normal limits. Endocrinology evaluations serum electrolytes, serum cortisol, dexamethasone suppression test, urinary metanephrine, vanillylmandelic acid, plasma renin and aldosterone levels were all within normal range indicating a non-functioning tumour. Left adrenal mass was found on ultrasonography. Contrast computed tomography (CT) abdomen pelvis revealing a well-defined heterogeneously enhancing soft tissue mass with cystic component with thin septations in left adrenal gland measuring 10×9×11 cm. Lateral limb of adrenal gland is not seen separate from the lesion and mass is extending from D11 to L3, abutting aorta, pancreas and displacing left kidney suggestive of malignancy. No additional mesenteric/retroperitoneal lymphadenopathy was identified.

Laparoscopic approach of removal of tumour was attempted but the tumour being inaccessible on posteromedial aspect, conversion to laparotomy has been done revealing a mass of around 9×7 cm posterior to pancreas attached left adrenal gland displacing left kidney

with a feeding branch from aorta. The tumour mass has been excised, sent for histopathology. Gross examination revealed a well circumscribed, lobulated, partially encapsulated adrenal mass measuring 9×7.5×5 cm. Cut section shows focal cystic, mucoid areas. Histologically, tumour composed of cellular and hypocellular areas comprising of interlacing short fascicles and bundles of spindle cells with indistinct cell boundaries, mild pleomorphism, degenerative changes reporting it to be an ancient schwannoma with S100 positive, synaptophysin and chromogranin negative.

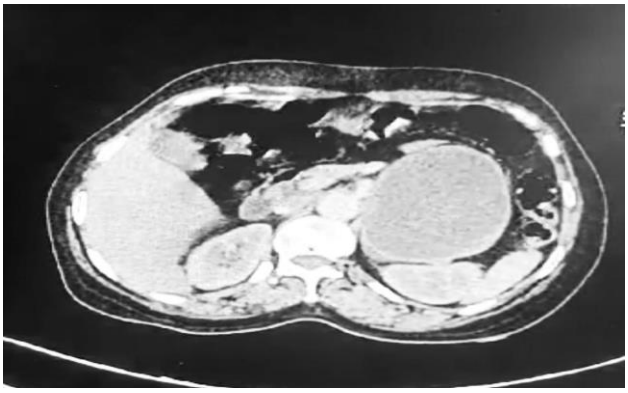

Figure 1: CT cross section (tumor highlighted).

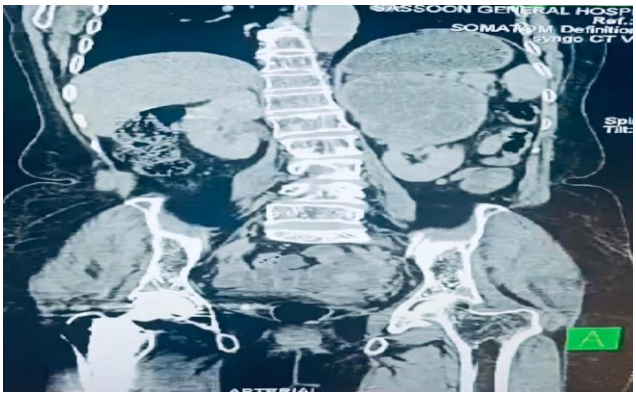

Figure 2: CT coronal section showing tumor above left kidney not separately visualised from left adrenal gland (tumour marked with an arrow).

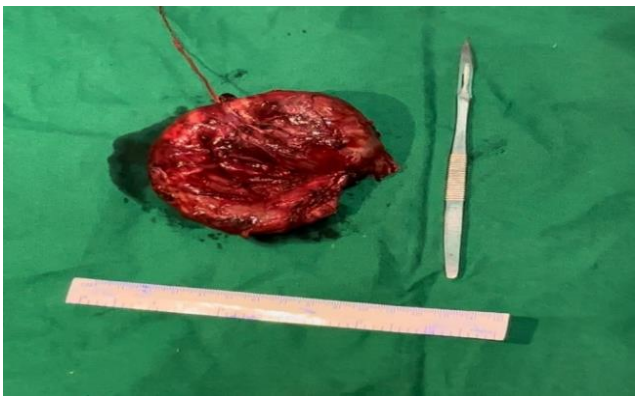

Figure 3: Gross appearance of tumour.

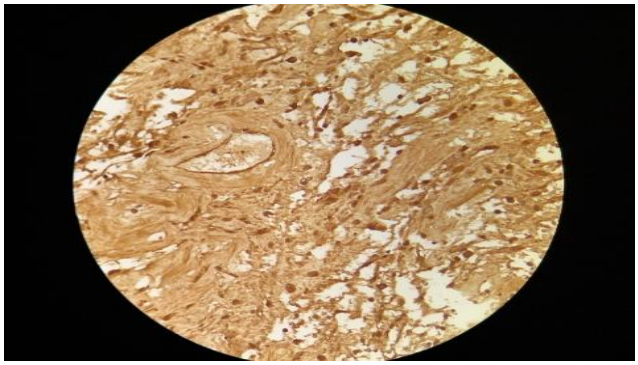

Figure 4: Slide showing S100 positivity.

DISCUSSION

Ancient schwannomas (AS) are a rare variant of schwannomas, originally described by Ackerman and Taylor in 1951.[4] Schwannoma is also known as neurilemmoma/neurinoma/perineural fibro blastoma.[5] AS commonly arise from neural crest cells with incidence of 0.7 to 2.7%.[6,7] It is a peripheral nerve sheath tumour which is benign in nature originating from Schwann cells forming the nerve sheath in cranial (except CN 1,2)/peripheral/autonomic nervous system.[5] It is mostly seen in head, neck, flexor regions presenting as vestibular schwannoma/acoustic neuroma which is a patho-gnomic feature of neurofibromatosis-2 (NF-2).[8] The term ancient schwannoma is derived from histological appearance of tumour attributing to its long-standing nature causing degenerative changes but behaves like a schwannoma.[5]

Retroperitoneal location of ancient schwannoma is rare, of which adrenal gland is rarely encountered and it is a non-functional adrenocortical tumour.[9] They are seen in a male to female ratio of 2:3 with mean age of presentation being 20 to 50 years.[2,6] They are solitary, benign in nature. If RPS are associated with neurofibramatosis-1 they are multiple and malignant in nature appearing at a younger age.[11]

The origin of the adrenal schwannoma is considered to be from either of the two myelinated nerve systems innervated to the adrenal medulla: one is the sympathetic nerve from the upper lumbar plexus and the other is the phrenic or vagus nerve.[4] They are usually detected when their large size produces compressive symptoms with masses less than 5cm are diagnosed incidentally on scans.[10] It has several differential diagnoses as pheochromocytoma, paraganglioma, incidentaloma, ganglioneuroma.[2,13]

Computed tomography (CT), and magnetic resonance imaging (MRI) scans are useful in diagnosis. Per cutaneous needle biopsy is not recommended due to risk of tumour dissemination/haemorrhage instead core needle biopsy through retroperitoneal route can be attempted.[2,12]

Definitive diagnosis is through histopathology showing a distinctive areas spindle cell proliferations arranged in

biphasic patterns of Antoni A (hypercellular areas of palisading pattern as verocay bodies) and Antoni B (loosely arranged haphazard cells with abundant cytoplasm) as seen in this case.[9] IHC stains like S100 and SOX10 show strong positivity, which is characteristic of schwannoma.[9,14]

Treatment is only complete surgical excision of tumour with clear margin. Less radical surgery (like enucleation) is preferred in these cases rather than complete excision with surrounding involved structures which is mostly believed to have high recurrence rate.[15] Recurrence of these tumours are less with good prognosis. In the case of incomplete excision of tumour, the incidence of recurrence is 5% to 10%. So, long-term follow-up is required for these patients.[10] Metastasis of these tumours are uncommon with most common site being liver with only 1 reported case. For locally advanced, malignant and metastatic conditions chemotherapy with doxorubicin, etoposide, ifosfamide have shown encouraging results.[1,16]

CONCLUSION

Ancient schwannomas are mostly benign and rare tumours. They have excellent prognosis with surgical resection. They are obliged with diagnostic difficulty, which needs CT and histopathology evidence for confirmation. They have to be considered as a differential diagnosis to soft tissue tumours. They have to be treated in a tertiary care centre with multidisciplinary approach due to lack of supporting literature.

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