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Case Report

Adrenal Schwannoma: a rare adrenal incidentaloma

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ABSTRACT

Adrenal schwannoma is a rare type of adrenal incidentaloma and is found in the medulla. There have been only 33 reported cases worldwide. We present a 56-year-old female referred to our institution for a history of abdominal discomfort and a left adrenal solid mass incidentally discovered in USG. Patient underwent surgical excision of the tumour. Histopathologic examination showed a neoplasm composed of cells arranged in interlacing fascicles with alternating hyper and hypo cellular areas, Immunohistochemistry (IHC) showed positive for S100 compatible with adrenal schwannoma.

Keywords: Adrenal gland tumour, Adrenal schwannoma, Incidentaloma

INTRODUCTION

Schwannoma (neurilemmoma) is an uncommon, benign, encapsulated tumour arising from nerve sheath cells.[1] They can occur anywhere in neural tissue where Schwann cells are present but are most common in the head and neck region and upper and lower extremities. Schwannomas rarely develop in the retroperitoneum but if they do, they can grow to a large size, sometimes displacing, and more rarely invading, surrounding structures and organs before becoming clinical apparent.[2] Visceral schwannomas are extremely rare and are usually discovered serendipitously.[3] Adrenal schwannoma is a rare type of adrenal incidentaloma and is found in the medulla. There have been only 33 reported cases worldwide.[4] We present a patient with a schwannoma of the left adrenal gland. To our knowledge this is the first report of adrenal schwannoma in an adult patient from India in the English literature.

CASE REPORT

A 56-year-old female with no known comorbidities presented with complaints of abdominal pain for 2 years. Physical examination, routine blood and urine tests were normal.

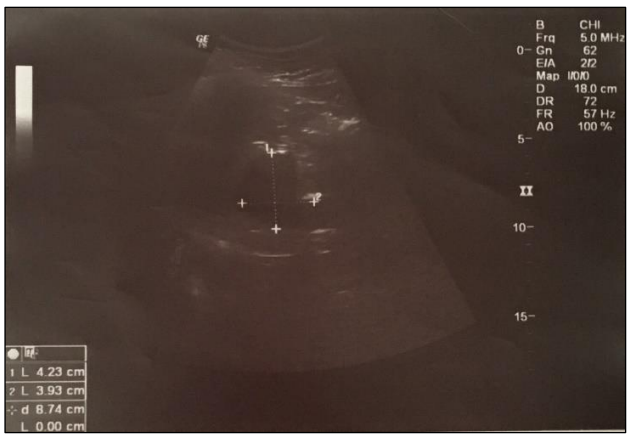

Figure 1: USG abdomen showing a hypoechoic lesion in left suprarenal region.

On evaluation ultrasonography (USG) abdomen showed a hypoechoic lesion of size 4.2 × 3.9 cm in left suprarenal region probably an adrenal adenoma (Figure 1).

Patient was well built and nourished. Blood pressure was within normal limits. Complete preoperative hormonal evaluation was carried out to evaluate functionality and the lesion was considered a non-secreting tumour. Contrast enhanced computed tomography (CECT) abdomen showed a well-defined, left suprarenal mass (5.3 × 4.4 × 5.1 cm) with smooth margins, no calcifications, heterogeneous enhancement with few small hypodense areas and progressive enhancement on delayed scan (Figure 2) and the provisional diagnosis was that of an adrenal carcinoma.

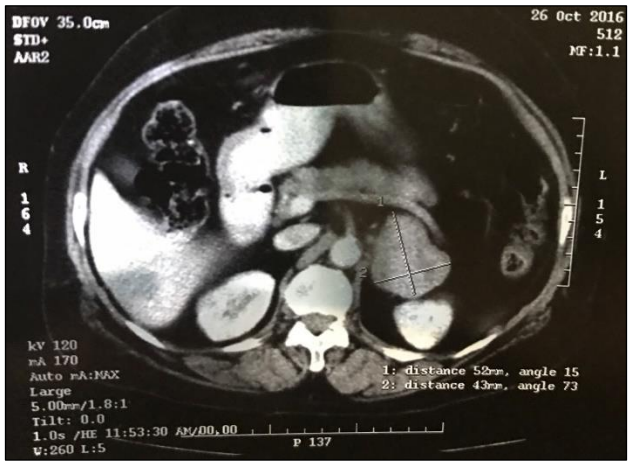

Figure 2: CECT abdomen showing a well-defined left suprarenal mass.

After a multidisciplinary committee evaluation, surgical resection was recommended. We proceeded with open left adrenalectomy. Intraoperatively there was a 5 × 5 cm firm nodular left adrenal mass with no infiltration to surrounding tissue. Rest of the abdomen was normal. Postoperative period was uneventful, and the patient was discharged on postoperative day five.

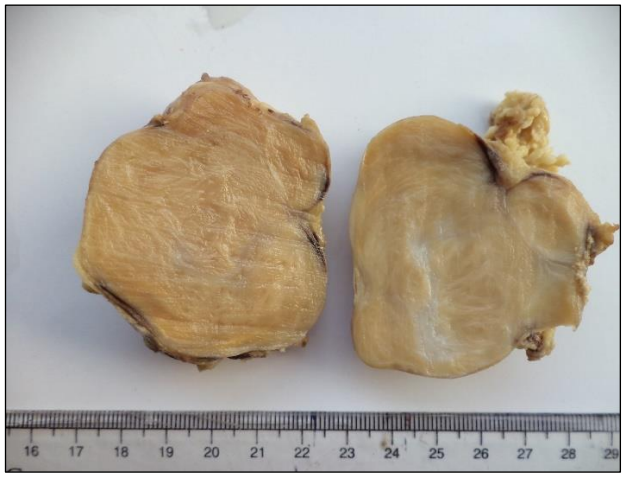

Figure 3: Gross examination showing a well encapsulated grey white nodular mass.

Gross examination of the resected specimen showed a well encapsulated grey white nodular mass (5.5 × 5 × 4 cm) with surface showing haemorrhage and attached fat (Figure 3). Cut section of the mass showed whitish and yellowish areas with faint whorling.

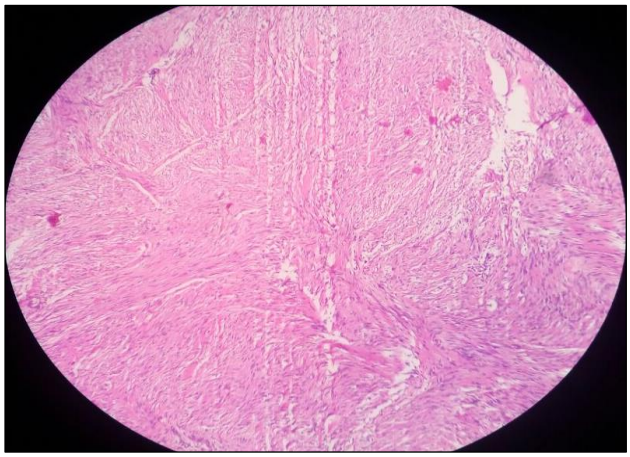

Figure 4: Haematoxylin and Eosin stained section of the tumor- low power (10X) neoplasm composed of cells in interlacing fascicles.

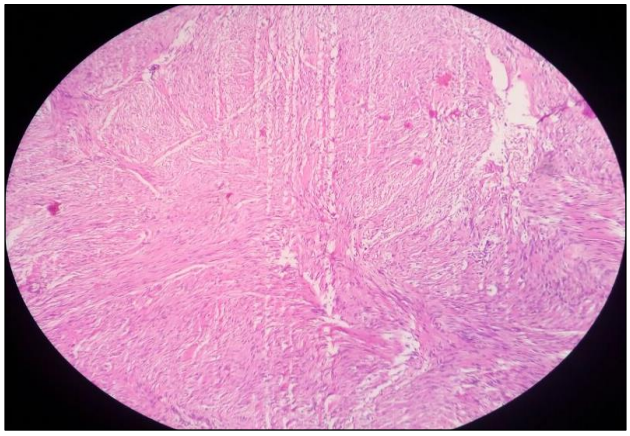

Figure 5: Haematoxylin and Eosin stained section of the tumor-high power view (40X) showing spindly cells, some with wavy nuclei

Histopathologic examination showed a neoplasm composed of cells arranged in interlacing fascicles with alternating hyper and hypo cellular areas (Figure 4 and 5). Immunohistochemistry showed positivity for S100, weakly positive for vimentin and negative for desmin. A diagnosis of adrenal schwannoma was made.

DISCUSSION

Adrenal Schwannoma is a rare incidentaloma found in the adrenal medulla and only less than 35 cases have been reported till date.[4,5] Schwannomas commonly originate in the myelin sheath of peripheral, motor, sensitive, sympathetic, or cranial nerves. They are most often found within the head, neck, upper and lower extremities, and with a lesser frequency on the trunk level, gastrointestinal tract and retroperitoneum.[6,7] Schwannomas are

encapsulated, usually benign, slow-growing nerve sheath tumours, containing Schwann cells in the stroma with minimal collagen.[3] It has been theorized that adrenal gland schwannomas originate from Schwann cells that insulate the nerve fibres innervating the adrenal medulla.[3]

It is difficult to diagnose adrenal schwannoma preoperatively by imaging alone and surgical resection is the primary management.[3] Adrenal schwannomas are typically found incidentally; however, patients may present with clinical symptoms secondary to the mass effect of the tumor.[3] Four of eight cases reviewed in the literature were discovered incidentally, with abdominal discomfort being the most frequent presentation in the other four cases.[3] CT-scan presentation is that of a well-circumscribed, homogeneous, round or oval mass, with slight enhancement.[8]

Cystic degeneration or calcification are described as a terminal stage of degeneration in a long-standing schwannoma and may suggest its diagnosis.[7,9] Magnetic resonance imaging (MRI) findings are nonspecific - adrenal schwannomas being included in the differential diagnosis of solid non-functional adrenal tumors.[6] Definitive diagnosis is possible only after histological examination of the operative specimen.[6] Almost all schwannomas show intense immuno-histochemical staining for S-100 protein, confirming the neurectodermal origin of the tumour cells.[2]

CONCLUSION

Adrenal schwannomas are rare tumours that are difficult to diagnose. Pre-operative workup is non-diagnostic and can only postulate a non-secreting adrenal mass. Hence complete resection is recommended.

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