

Case Report

Management of giant adrenal schwannoma

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ABSTRACT

Adrenal schwannoma is a rare differential of adrenal incidentaloma with scarce literature available at present. Adrenal schwannomas are usually non-secreting tumours but can very rarely secrete catecholamines. Specimen of our patient was 14 cm in diameter and weighed 900 gm which is the largest adrenal schwannoma in literature. Although it is a benign lesion but there are no standard treatment guidelines because of rarity of lesion.

Keywords: Adrenal incidentaloma, Adrenal schwannoma, Adrenalectomy

INTRODUCTION

Adrenal incidentaloma (AI) has a prevalence of 4-6% in general population.¹ Schwannoma is a rare differential of adrenal incidentaloma with scarce literature available at present. They are slow growing nerve sheath tumours with no evidence of hormone secretion. Patient usually present with vague abdominal complaints. Management includes adrenalectomy via open or minimal invasive approach. Diagnosis is confirmed on histopathology and immunohistochemistry (IHC).² Herein we describe a case of a very large non-secreting adrenal mass detected incidentally which had large area of contact with renal vessels, aorta and surrounding viscera. Adrenalectomy was done with careful dissection and histopathology with IHC confirmed a diagnosis of conventional schwannoma.

CASE REPORT

An Asian female aged 50 years (BMI- 30.2 kg/m²) presented to surgical OPD for left adrenal mass detected incidentally on ultrasound abdomen done for an unrelated fever episode 6 months ago. On examination her vitals were normal, per abdomen examination revealed a 12×10 cms firm, non-tender mass with smooth surface in the left

hypochondrium extending till left lumbar region. Upper margin of the mass could not be felt.

The mass was ballotable and moving with respiration. Ultrasound abdomen revealed 13.4×12.5 cms round to oval hypoechoic mass at upper pole of left kidney. Contrast enhanced CT scan (Fig. 1A, 1B, 1C) revealed well defined heterogeneously enhancing 10.4×11 cms mass in the left suprarenal location. Mass had >180 degrees area of contact with left renal artery and vein and 90 degrees area of contact with the left lateral aspect of descending abdominal aorta at the level of branching of left gonadal artery but with no evidence of vascular invasion (Figure 1B, 1C, 1D). Mass displaced the surrounding organs, but fat planes were maintained. MIBG SPECT/CT revealed no radiotracer uptake (Figure 2). A provisional diagnosis of adrenocortical carcinoma was made.

Blood investigations including complete blood count (CBC), liver function test, kidney function test, serum cortisol, cortisol post dexamethasone suppression test, DHEAS, serum testosterone, 24 hours urinary VMA were normal. Patient underwent open transabdominal left adrenalectomy. Intraoperatively adrenal mass was

adhered to renal vessels and descending aorta. Mass was carefully dissected, and vessels were spared. On gross pathological examination (Figure 3A) a globular well encapsulated 14x10x9 cms mass weighing 900 gm was found. Cut section (Figure 3B) revealed grey white tumour with variegated appearance. No areas of cystic degeneration or haemorrhage were seen. Histopathological examination (Figure 4) revealed conventional adrenal schwannoma with normal adrenal tissue at the periphery. Tumour cells were immunopositive for S-100. Post-operative period was uneventful and discharged on post-operative day 3. On 18 months follow up patient is in preserved state of health with no recurrence detected clinically.

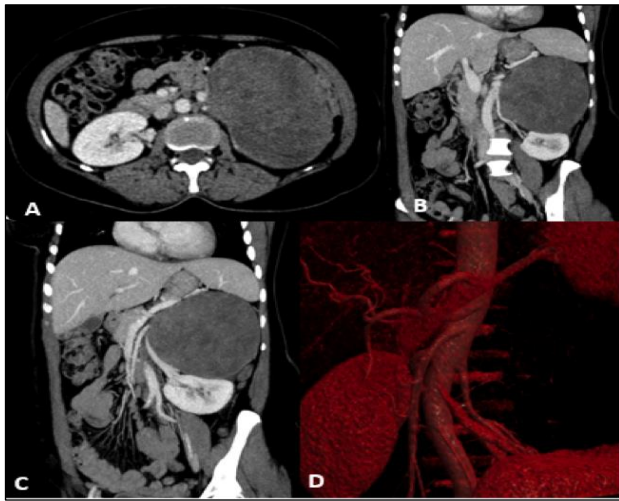


Figure 1: Axial (A) and coronal (B and C) CT angiogram images show 13.4x12.5 cm heterogeneously enhancing mass in left adrenal location. The mass has >180 degrees area of contact with left renal artery (B) and vein (C) and 90 degrees area of contact with the left lateral aspect of descending abdominal aorta at the level of branching of left gonadal artery. (D) VRT image shows no significant narrowing of the renal vessels.

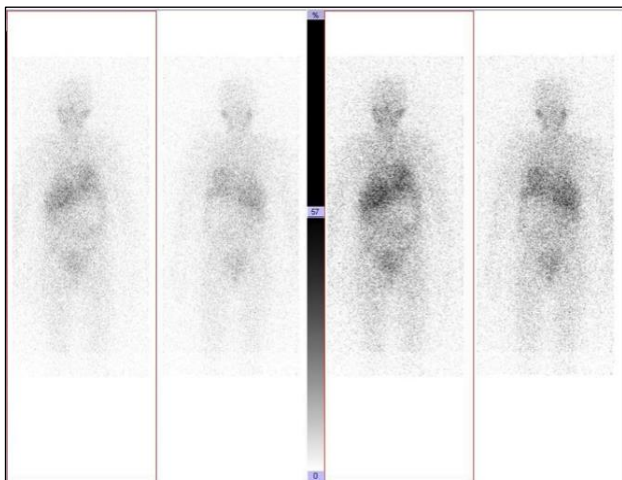


Figure 2: MIBG scan show no radiotracer uptake.



Figure 3: gross histopathology shows (A): globular mass measures 14x10x9 cm and weighing 900 gm and cut section (B): shows grey-white tumour with variegated appearance. No areas of cystic degeneration or haemorrhage were seen.

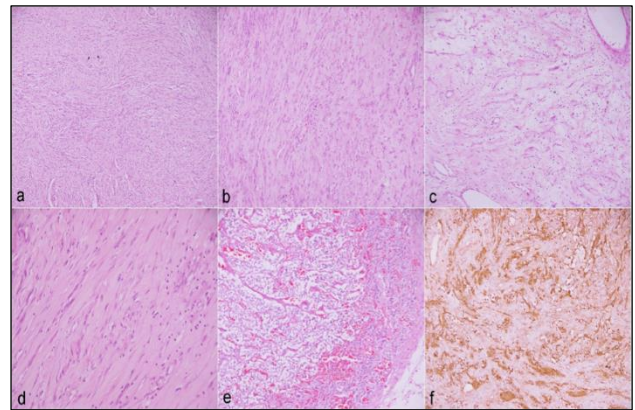


Figure 4: (a): The tumor shows spindle cells arranged in ill-defined fascicles in the cellular areas, X100, H&E. (b): At places, the spindle cells show palisading and Verocay body formation in the cellular (Antoni A) areas, X200, H&E. (c): The other less cellular (Antoni B) areas show fewer spindle cells scattered in abundant loose myxoid stroma, X100, H&E. (d): The spindle cells display thin wavy mildly pleomorphic nuclei and ill-defined eosinophilic cytoplasm, X200, H&E. (e): Normal adrenal parenchyma is seen at the periphery of the lesion, X100, H&E. (f): The tumor cells stain positive for S100, X100, IHC.

DISCUSSION

Adrenal incidentalomas have an incidence of 4-6% in general population.¹ Incidence is further increasing with increase in awareness, health screening and advances in imaging modalities.³ Differential diagnosis to these depend on their location. Cortical lesions can be adenoma or carcinomas. Medullary lesions can be pheochromocytoma, neuroblastoma and gangliogliomas.⁴ Adrenal schwannoma is a rare differential of adrenal incidentaloma. To the best of our review of literature less than 60 cases have been described previously.³⁻⁵ Schwannomas are slow growing benign nerve sheath tumours which principally arise from neural crest cells. They are usually located in head, neck and

flexor surfaces of upper and lower limbs. Retroperitoneal schwannomas account for 1-3% of all schwannoma.⁶ Mohiuddin et al in a review of 33 patients with adrenal schwannomas found a slight female predominance, with most of them presenting as incidentalomas and only 13 patients presented with symptoms, abdominal pain and discomfort being most common. Adrenal schwannomas are usually non-secreting tumours but can very rarely secrete catecholamines and present as hypertension.^{4,5} Li et al, in series of 19 patients also found female predominance, 16 being females.⁷ Only 4 patients reported abdominal discomfort. All except two patients had normal catecholamines levels. rest two had low levels of catecholamines.

Computed tomography scan reveals a well-defined homogenous solid mass. Rarely if long standing, can also show heterogenous changes with calcifications and cystic changes. Characteristic enhancement patterns of primary adrenal schwannomas are mild heterogeneous enhancement on arterial phase and progressive enhancement during the portal venous and equilibrium phases which, are likely due to variable degree of tumour cellularity or degenerative changes such as cystic degeneration, necrosis, and haemorrhage.^{8,9} Minimal contrast enhancement may be seen in neoplasms with low cellularity and edema. Therefore, differential diagnosis of adrenal schwannomas includes variety of non-functioning solid tumours and cystic adrenal lesions.

Standard management of adrenal masses more than 6 cm is adrenalectomy, though appropriate approach, either open or laparoscopic, is yet to be studied in adrenal schwannomas.⁴ Actual diagnosis lies in the hands of pathologist. Moiuiddin et al, reported a median size of 5.5 cm (0.6-14.5 cm) and median weight of 84 gm (31.5-600 gm).⁴ Specimen of our patient was 14 cm in diameter and weighed 900 gm which is the largest adrenal schwannoma in literature. They are usually well encapsulated firm and round masses and are tan yellow to greyish white on cut section. Most are solids and homogenous but long-standing adrenal schwannomas may reveal areas of cystic degeneration, calcifications and haemorrhage.^{4,10}

Histopathological examination reveals presence of Antoni A and Antoni B areas with Verocay bodies.² Antoni A areas represent fascicles of spindle cells with cytologically bland nuclei, indistinct cell borders, and faintly eosinophilic cytoplasm in a collagenous stroma. Fascicles of spindle cells that are hypocellular are known as Antoni B areas. whereas Verocay bodies are nuclear-free zones in between regions of nuclear palisading. Schwannomas are divided into conventional or cellular schwannomas based on the presence or absence of these bodies. Cellular schwannomas consist entirely of Antoni A areas and are devoid of Antoni B areas and Verocay bodies. Whereas conventional schwannomas consist of all Antoni A, Antoni B and Verocay bodies. On immunohistochemistry, schwannomas are classically

positive for S-100, vimentin and CD 56 and are negative for synaptophysin, chromogranin, CD 34, CD 117 and HMB 45.^{1,2} Histopathology combined with immunohistochemistry differentiates it from other differential diagnosis such as pheochromocytoma, ganglioneuroma, solitary fibrous tumours, gastrointestinal stromal tumours and melanoma. Correct differentiation into conventional and cellular schwannoma is very important, as cellular schwannomas are very difficult to differentiate from close differentials of sustentaculoma and neurofibroma which share same microscopic and immunohistochemistry features and can be differentiated only on the basis of electron microscopy.¹¹ Follow up using ultrasound and CT scans have been done by different authors for a variable time period and none of them have shown any recurrence or metastasis, probably because of the known benign nature of the lesion. But accurate follow up protocols and preferred imaging is yet to be studied.^{4,12,13}

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