



## Adrenal Schwannoma: A Case Report

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### Abstract

*Schwannoma is a benign peripheral nerve sheath tumor arising from the schwann cells. The usual location of Schwannoma is in the head & neck region and the extremities. Adrenal schwannoma is an extremely rare cause of incidentaloma accounting for 0.7% of all adrenal tumors. We report a case of benign right adrenal schwannoma detected in a 43 year old female presenting with right flank pain. There were no clinical features suggestive of Cushing disease, Pheochromocytoma or primary hyperaldosteronism. Computed Tomography (CT) of the abdomen showed a heterogeneously enhancing mass of size 9\*5cm. The biochemical and metabolic evaluations including endocrinology studies were unremarkable and consistent with a non functioning adrenal mass. Adrenalectomy was performed due to the large size of the tumor. Histopathological examination revealed a well circumscribed, partially encapsulated mass of size 9\*7.5\*5cm. Tumor was composed of fascicles of spindle cells with alternating hypercellular and hypocellular areas with mild atypia and no mitosis. Degenerative changes in the form of cystic and mucoid areas were seen. On the basis of histopathological examination the diagnosis of adrenal schwannoma was made. The diagnosis was supported by immunohistochemical positivity for S-100. Adrenal schwannoma is a rare tumor with less than 50 cases reported in the literature, and hence the significance of this case report.*

**Keywords:** Adrenal tumor, Schwannoma, Adrenal schwannoma.

### Introduction

Schwannoma, also known as neurilemmomas, is a rare benign peripheral nerve sheath tumor originating from the schwann cells, surrounding the nerve.<sup>1</sup> It is predominantly common in the head, neck and extremities.<sup>2</sup> It is the most common benign tumor of the peripheral nerve, accounting for 90% of all peripheral neural tumors and 8% of all benign soft tissue tumors.<sup>3</sup> 1-3% of all schwannomas are retroperitoneal in

origin.<sup>4</sup> Adrenal schwannomas are extremely rare and accounts for 0.7% all the adrenal tumors<sup>2</sup>. Adrenal schwannoma are mostly asymptomatic and difficult to diagnose due to its indistinct clinical and radiological features.<sup>4</sup> Histopathology and immunohistochemical staining are the gold standard in diagnosis of adrenal schwannoma.<sup>5</sup> Herein, we report a case of adrenal schwannoma incidentally discovered in a 43 year old female who presented with right lumbar pain.

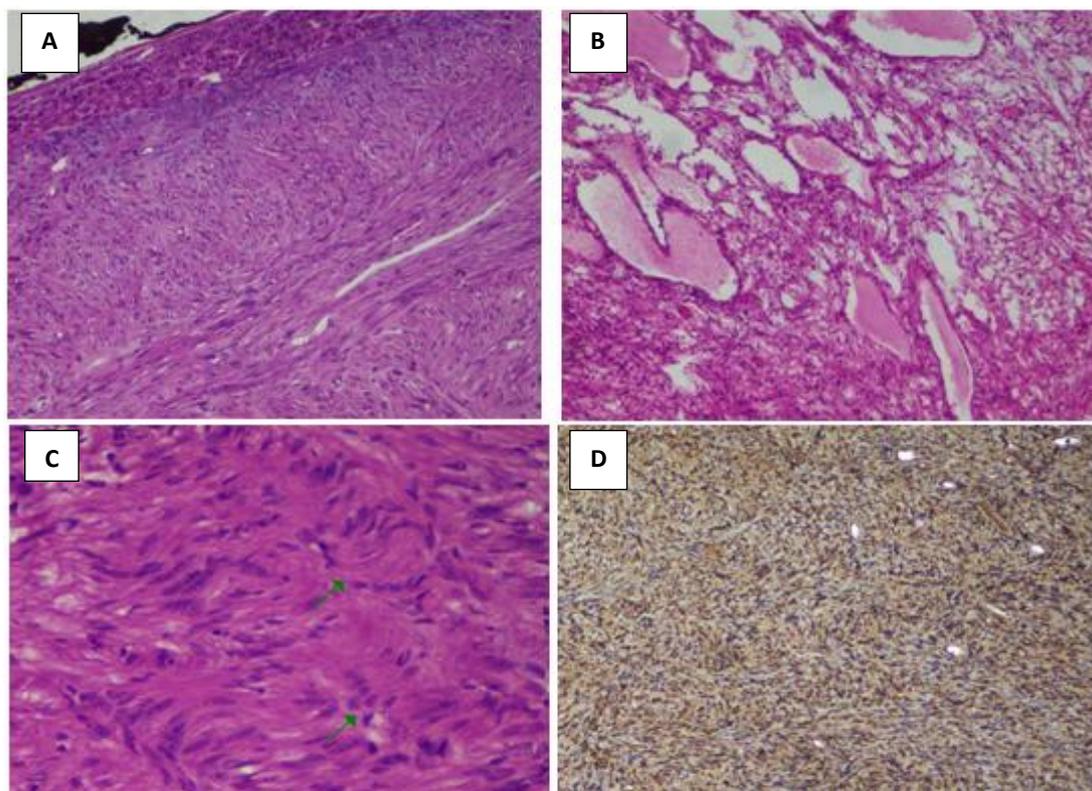
### Case Report

A 43 year old female presented to our hospital with the chief complaints of right flank pain. During examination, a right adrenal mass was found on ultrasonography. Her medical and family history were unremarkable. On physical examination, blood pressure was 128/80 mm Hg, pulse rate was 78 beats/min, blood glucose was within normal range, abdomen was soft with no organomegaly and there were no cushingoid features. Blood and biochemistry analysis were within normal limits. Endocrinology evaluations including 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 tumor.

CT scan reveals a heterogeneously enhancing soft tissue mass in the right adrenal gland measuring 9\*5 cm suggestive of malignancy. No additional mesentric or retroperitoneal lymphadenopathy was identified on abdominal CT. Thus, open

adrenalectomy was done and sent for histopathological examination.

Gross examination revealed a well circumscribed, lobulated and partially encapsulated adrenal mass measuring 9\*7.5\*5 cm. Cut section of which shows focal cystic and mucoid areas. Histologically, tumor was composed of cellular and hypocellular areas comprising of interlacing short fascicles and bundles of spindle cells with indistinct cell boundaries, mild pleomorphism without any areas of necrosis and mitosis. Focal microcystic areas, myxoid change and degenerative nuclear atypia with area of hyalinization were seen. Normal adrenal tissue was compressed at the periphery. Diagnosis was confirmed by immunohistochemical analysis with strong nuclear and cytoplasmic positivity for S-100. A final histopathological diagnosis of adrenal schwannoma with degenerative changes was made. The patient was discharged uneventfully on the 7<sup>th</sup> day after surgery.



**Fig. A to D:** Microscopic view of adrenal schwannoma: (A) Well encapsulated schwannoma with normal adrenal tissue (B) Areas with cystic change (C) Predominantly Antoni A area with small Antoni B area (D) Cells showing diffuse positivity for S 100.

## Discussion

Schwannoma is a tumor arising from the myelin sheath of the peripheral nerves.<sup>6</sup> Origin of the main component of schwannoma is from the neural crest cells.<sup>7</sup> Therefore, schwannomas can arise in every organ or nerve except cranial nerves 1 and 2.<sup>5</sup> A schwannoma is a well encapsulated, benign slow growing tumor. It was first described by Verocay in 1908.<sup>8</sup> Antoni in 1920 subclassified these tumor into two distinct categories on the basis of hyalinization and interstitial fibrosis.<sup>8</sup> They usually have predilection for the head, neck region and the upper and lower extremities.<sup>9</sup> However, there have been sporadic cases of these tumor arising in the trunk, gastrointestinal tract, liver, pancreas, kidney and retroperitoneum.<sup>9</sup>

Adrenal schwannoma has slight female preponderance and is mostly seen in the second to fifth decade of life as in the case presented.<sup>2</sup> Majority of the patients are asymptomatic, only few of them presents with non specific symptoms of abdominal or lumbar fullness, pain and discomfort secondary to adjacent organ compression as our case presented with right flank pain.<sup>3</sup> Adrenal schwannoma are mostly solitary and benign.<sup>7</sup> Adrenal adenoma, pheochromocytoma, myelolipomas, neuroblastoma, ganglioneuromas, cyst, malignancy and metastasis are the differential diagnosis of adrenal schwannoma.<sup>2</sup> Schwannoma is non functional adrenocortical tumor, due to its non secretory and asymptomatic nature they are difficult to diagnose preoperatively.<sup>6</sup>

Advances in the imaging modalities have led to an increased incidental detection of adrenal tumors.<sup>4</sup> On CT these lesions appear as well circumscribed, homogeneously enhancing solid white masses, while few are cystic with areas of hemorrhage and calcification giving it a heterogeneous appearance.<sup>2</sup> In the case presented, CT scan showed heterogeneously enhancing mass which correlates with degenerative changes in the lesion. Histopathological and immunohistochemical evaluation determine the final diagnosis.

The characteristic microscopic appearance of conventional schwannoma is of alternating

hypercellular (Antoni A) areas that palisades Verocay bodies and a myxoid hypocellular (Antoni B) areas.<sup>4</sup> Long standing tumors also show degenerative changes including interstitial fibrosis, cyst formation, calcification, hemosiderin deposition and hyalinization.<sup>9</sup> Malignant transformation can also be seen in schwannoma mostly in association with Von-Recklinghausen's disease & neurofibromatosis.<sup>6</sup> Confirmation of histopathological diagnosis is made by immunohistochemistry, lesion is immunoreactive for S-100, Vimentin, laminin and negative for desmin, SMA (smooth muscle actin), CD117. Schwannoma also stain positive for calretinin which helps it to differentiate it from neurofibroma.<sup>6</sup>

Treatment of choice for adrenal incidentaloma is surgical excision, for all lesion greater than 6 cm.<sup>9</sup> The factors determining the resectibility of the tumor are tumor size, imaging characteristics and the rate of growth of tumor. As in our case CT imaging were suggestive of large suspicious lesion warranting surgical excision. Recurrence rate is low for adrenal schwannoma and is generally associated with an incomplete surgical margins. Surgical excision is both diagnostic as well as therapeutic for adrenal schwannomas as they are not responsive to either chemotherapy or radiotherapy.<sup>3</sup>

## Conclusion

We report a rare case of an adrenal schwannoma in a 43 year old female patient. Pre-operative evaluation of these schwannomas were inconclusive. Thus prompt surgical excision followed by histopathological and immunohistochemical analysis is essential for the diagnosis. Adrenal schwannomas are rare tumor that are mostly found incidentally and its suspicion should be kept in any adrenal mass.

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**Conflicts of Interest -** Nil

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