Abstract:
Introduction: Schwannomas are rare tumors of schwann sheaths of peripheral, sympathetic and cranial nerves. They are of Neural crest origin. Usually head and neck, upper and lower extremity are commonly involved. GIT, retroperitoneal schwannomas are rare. Juxta adrenal schwannomas are very rare. Case presentation: A 50 year old female presented with dull aching loin pain on the left side and on imaging was found to have an adrenal mass lesion which was non functioning. The patient was taken for adrenalectomy considering the size of the mass and symptoms. The mass turned out to be a juxta adrenal schwannoma to our surprise. Conclusion: Schwannomas are very rare tumors in juxta adrenal location. They are usually asymptomatic in this location and grow to very large sizes. Rarely they can also be malignant. Imaging is inconclusive and often confirmation is made post-operatively after Histopathology and Immunohistochemistry reports.

These rare tumours should be borne in mind during differential diagnosis of an adrenal incidentaloma.

Keyword: juxta adrenal, schwannoma, non functioning

CASE PRESENTATION:
A 50 year old female presented with left loin pain for the past 2 months. It was dull aching, non radiating. Patient had no other urological symptoms. General condition was fair and abdomen examination. Per rectal and per vaginal examination were not contributory. On investigations, routine haemogram was normal. Urine pus cells 3-4 per hpf and no growth on culture of urine. Renal function and Liver function tests were normal. Chest radiograph was normal. Ultrasonography examination of abdomen – right Kidney measured 9.8 x 4.2 cms in size and corticomedullary differentiation was maintained and there was no dilatation of the pelvicalyceal system. There was a 6 x 5 cm left suprarenal mass, which was uniformly hypoechoic. The left kidney size and cortico
medullary differentiation were normal. Contrast enhanced CT scan well defined homogeneously enhancing mass in the region of left adrenal gland with smooth margins with no evidence of necrosis / heterodensity.

Biochemical evaluation was done. Plasma free metanephrines and 24 hour urinary cortisol were within normal limits. Pt was preoperatively prepared with phenoxybenzamine 10 mg bd for 2 weeks and taken up for surgery 11th rib bed incision. Massive splenomegaly (25 x 20 cm) was noted and splenic vein was stretched and tortuous. Major part of tumour was beneath the splenic hilum. Splenic vessels were meticulously dissected away from the mass. Then left adrenalectomy was completed, with minimal handling of tumour tissue.
OPERATIVE PICTURE

POST OP SPECIMEN

HPE The HPE report was encapsulated neoplasm from outside compressing adrenal gland consisting of intersecting bundles of elongated spindle cells and elongated nuclei in palisading arrangement. IHC was done for confirmation and was positive for S 100 confirming neural origin of the tumour suggestive of a schwannoma (fig 8,9)

DISCUSSION:
Schwannomas are rare tumors of schwann sheaths of peripheral, sympathetic and cranial nerves. They are of Neural crest origin. Usual sites are head and neck, upper and lower extremity. GIT, retroperitoneal locations are rare. Juxta adrenal location is very rare. They are benign, slow-growing, encapsulated tumors, but rarely may be malignant. Malignant varieties are seen in association with neurofibromatosis type 2. Juxta adrenal schwannoma’s cell of origin is from Schwann cells of the nerve fibers in retroperitoneum in close proximity to adrenal gland. They are usually asymptomatic and can grow to a very large size before producing symptoms. In cross sectional imaging (MRI) they appear as a well-circumscribed, homogeneous, round or oval mass, with slight enhancement. However based on the extent of cellularity they can also be very hyperdense mimicking a pheochromocytoma, as in this case. Definitive diagnosis is possible only after histological examination (IHC) of the operative specimen. HPE usually shows fascicular proliferation of spindle cells and nuclear palisading pattern. There may be alternating areas of high and low cellularity called as Antony A and Antony B areas respectively. These highly cellular areas give rise to high signal attenuation values in T2 weighted images. Definitive confirmation is made with Immunohistochemistry which is positive for S 100 and Vimentin.
CONCLUSIONS:
Schwanomas in retroperitoneal location are extremely rare tumours with only few cases reported in literature. They are usually slow growing tumours which can reach considerable size before diagnosis. The imaging characteristics are not specific and can be confused with a pheochromocytoma. These rare tumours should be borne in mind when evaluating an adrenal incidentaloma.

REFERENCES:


2. Juxta-adrenal malignant schwannoma with lymph node metastases.
