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Original Research Paper

Surgery

ADRENAL TUMORS – UNUSUAL PRESENTATION

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ABSTRACT

Background: Adrenal tumors are quite uncommon, but of late detected more due to better screening and imaging. We intend to study the rarest of these rare adrenal tumors over a period of 5 years and analyze them of their characteristics and follow up after surgical management.

METHODS: The number of adrenal tumors operated during this period was 54 from January 2008 to April 2017. Of these, we considered 5 tumors to be unusual.

RESULTS: The tumors were located 3 on the right side, 1 on the left & 1 bilateral. The histopathology of these tumors were bilateral non functioning paraganglioma, left hilar schwannoma, right malignant peripheral nerve sheath tumour (MPNST) with IVC invasion, right adrenal abscess and right adrenal ganglioneuroma.

CONCLUSION: Histological surprise in an adrenal tumor must be anticipated for better surgical care and need life-long follow-up.

KEYWORDS : Adrenal tumours

INTRODUCTION:

Historically, the adrenal tumor that was discovered incidentally, usually during an imaging procedure such as computed tomography (CT), magnetic resonance imaging (MRI), or Ultrasonography for symptoms unrelated to adrenal disease,(e.g., back pain) was called an *incidentaloma:'* As more physicians (and patients on their own) ordered these easily available imaging studies for common diseases an increasingly number of unsuspected adrenal tumors are found.

Here we discuss about 5 unusual adrenal tumors which we came across in our dept over a period of 9 yrs. The number of adrenal tumors operated during this period was 54 from January 2008 to April 2017. Of these, we considered 5 tumors to be unusual and retrieved the medical records and followed them. Mean age of these patients was 46.4 yrs (range 30yrs to 55yrs). All these tumors were non functional both by functionality and symptomatology. The varied presentations were non-specific including vague abdominal pain in majority (4 patients) and by incidental detection in 1 patient. Of these 5 patients, 4 were normotensive and 1 hypertensive. Table 1 & 2 shows the pre-op & postop diagnosis of all the patients who underwent Adrenalectomy in our hospital.

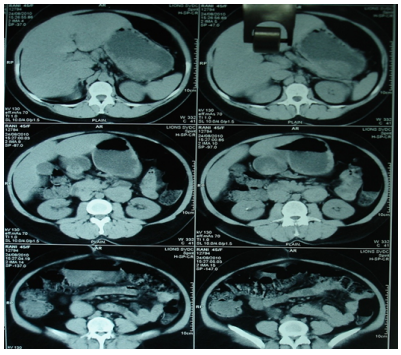
(Table 1)

Preoperative diagnosis	total	male	female
pheochromocytoma	18	9	7
Non functioning tumours	15	4	8
Adrenal cyst	3	0	3
Cushing`s Syndrome	2	0	2
Adreno cortical cancer	7	4	3
Conn`s Syndrome	4	2	2
Recurrent Pheochromocytoma	2	0	2
Recurrent adrenal adenoma	1	0	1
Retroperitoneal mass	2	0	2

(Table 2)

Post operative diagnosis	total	male	female
pheochromocytoma	17	9	6
Non functioning tumours	12	4	5
Adrenal cyst	3	0	3
Cushing`s Syndrome	2	0	2
Adreno cortical cancer	6	4	2
Conn`s Syndrome	4	2	2
Recurrent Pheochromocytoma	2	0	2
Recurrent adrenal adenoma	1	0	1
Retroperitoneal mass	1	0	1
B/L Non functioning Paraganglioma	1	0	1
Lipoma	1	0	1
Schwanoma	1	0	1
Malignant Peripheral Nerve Sheath Tumour	1	0	1
Adrenal Abscess	1	1	0
Ganglioneuroma	1	0	1

B/L Paraganglioma: 45 yr old female presented with vague abdominal pain. Her Bp was normal. On evaluation CT abdomen revealed a 6x3x4 cm mass right adrenal region and a 7x3x3 cm mass left adrenal region (fig 2). She was biochemically negative. By laparoscopy a bilateral para-aortic tumor of size Rt-8x4x4, Lt-8x4x4cm was removed. HPE revealed B/L paraganglioma.

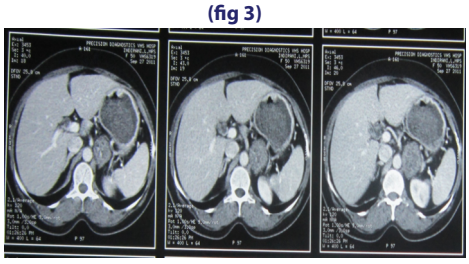


(fig2)

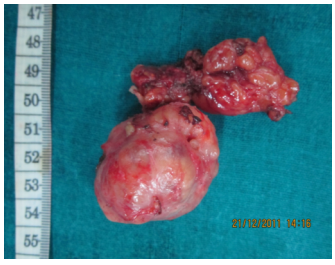
A paraganglioma is rare neuroendocrine neoplasm originating from paraganglia in chromaffin-negative glomus cells but are related to pheochromocytoma which are chromaffin-positive. 1–3% of tumours secretes hormones such as catecholamine and resembles pheochromocytoma. About 85% of paraganglioma develop in the abdomen, 12% in the chest and 3% in the head and neck region. The paraganglioma are solitary, polypoid vascular tumor with a firm to rubbery consistency. Nerve of origin does not penetrate substance of tumor but is in the periphery. Dumbbell tumor in posterior mediastinum originates from or extends into vertebral canal. Organ of Zuckerkandl paraganglioma may invade inferior vena cava. MRI is the most sensitive imaging modality.

Schwannoma: A 50 yr old female presented with urinary retention. On evaluation CT showed a 3x3 cm mass Rt adrenal (fig 3). Her Bp was normal and she was biochemically negative. On laparoscopy a 4x4 cm mass over left renal hilum excised (fig 4). HPE revealed a Left Schwannoma.

(fig 3)



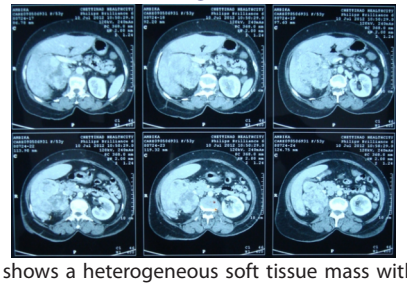
(fig 4)



Neural sheath tumors or schwannoma (neurilemmoma) generally are benign and occur as solitary tumors from the cranial and peripheral nerves of the head, neck, extremities, stomach, retroperitoneal space(1-5%), spinal nerves and the adrenal medulla constituting 0.2% of adrenal incidental tumors. The role of Schwann cells is to produce the myelin sheath that covers peripheral nerves. Malignant schwannoma are associated with neurofibromatosis types 1 and 2 and results in a high-grade sarcoma, local dissemination and distant metastasis

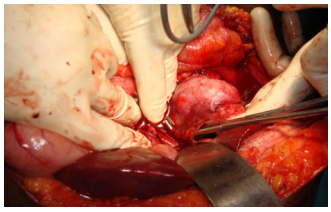
Malignant Peripheral Nerve Sheath Tumour: A 52 yr old female presented with abdomen pain right side for past 3 months. Her Bp was normal & she was biochemically negative. CT revealed a 20x16x12 cm mass right adrenal (fig 5). With a pre op diagnosis of right Adrenocortical carcinoma laparotomy revealed a 13x9x6 cm mass over right adrenal. The mass was infiltrating the IVC & adherent to liver (fig 6). IVC injured around 5cm linearly & repaired. The patient developed right pleural effusion & ileo-femoral thrombosis. HPE report was a malignant peripheral nerve sheath tumor (MPNST) (fig 7). The pt developed spinal metastases at 3 months follow-up & was referred for EBRT for pain reliefMPNST and expired 4 months later.

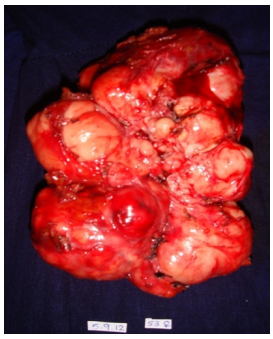
(fig 5)



CT image shows a heterogeneous soft tissue mass with multiple well-enhanced nodules at its periphery

(fig 6)



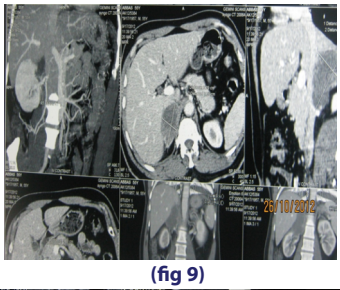


A solid grey brown /yellow nodular mass with a pseudocapsule of fibrous tissue- 13 x 9 x 6 cm (fig 7)

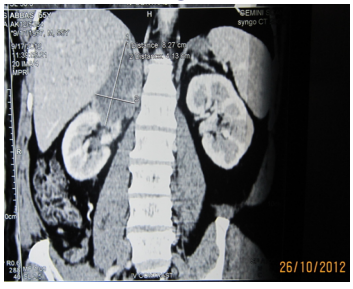
Malignant peripheral nerve sheath tumors (MPNSTs) are rare with an incidence of 0.1/100,000 accounting for 5-10% of all soft tissue tumors. On CT MPNST shows as a solid iso-dense in-homogenous expansile mass. On contrast the differentiating points are peripheral enhancement, peri-lesional edema like zone and intratumoral cystic lesion. Evidence of necrosis, encasement of Aorta, displacement of adjacent structures denotes malignancy.

Adrenal Abscess: A 55 yr old male presented with right abdominal pain. Apart from being a hypertensive he had no other complaints. CT revealed an 8.1x5.3x3.5 cm mass right adrenal (fig 8 & 9).

(fig 8)



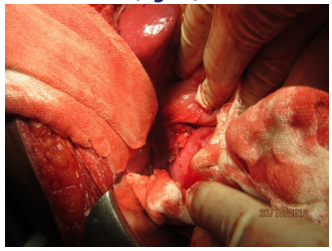
(fig 9)



CT shows a space occupying lesion with no calcification or fat in the mass & loss of fat plane between mass & liver

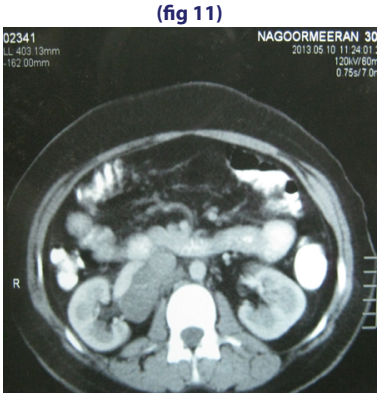
Laparotomy revealed a yellowish white fibrotic hard mass above right kidney plastered to IVC & surrounding structures (fig 10). Nil aspirate due to chronic inflammation. Due to adherence & distortion of normal anatomy & no space for dissection the abdomen was closed. On 3 months postop USG revealed a decrease in mass size.

(fig 10)

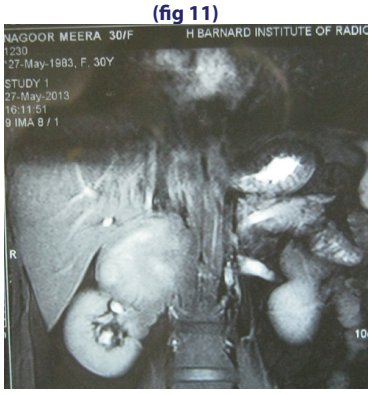


Ganglioneuroma: A 30 yr old female had an accidental fall & evaluation for abdominal pain revealed an incidentaloma right adrenal. CT showed a well-encapsulated 7.4x5.6cm hypo-echoic lesion right adrenal (fig 11 & 12).

(fig 11)



(fig 11)



On laparotomy an 8x8x6 cm mass over right supra renal & hilum. The vascular lesion was adherent to posterior portion of IVC extending up to Aorta & adherent to right kidney with right renal vein over the mass. Right renal vein injured & repaired. Due to adherence debulking of tumor done & nephrectomy deferred. HPE revealed mature ganglion cells and Schwann cells among a fibrous stroma-a Ganglioneuroma. Post op period was uneventful & patient is on follow-up.

Ganglioneuroma is a benign Neoplasm that arises from neural crest cells of sympathetic ganglia or adrenal medulla. It occurs more often in adults between the ages of 40-50 years with a slight female predominance (male/female ratio 2:3). The main locations are the posterior mediastinum, retroperitoneum, adrenal gland, and neck with 20% located in the adrenal medulla. On imaging, it appears as a homogeneous, encapsulated mass, with well-defined edges and without invading nearby structures. Calcification occurs in 40%-60%.

With a mean follow up of 14.6 months (range 3 to 29 months) the patient with MPNST developed spinal metastases at 3 months and referred for EBRT but died 6 months later. The remaining 4 patients had no complications/ recurrence and are on follow-up.

Conclusion: The presentation of adrenal tumors though varied, histological surprise is still a possibility. These unusual adrenal tumors may behave differently and still need life-long follow-up. These possibilities must be kept in mind intra-operatively and anticipated for better surgical care and outcome.