ORIGINAL RESEARCH PAPER

General Surgery

KEY WORDS:

A RARE CASE OF SPORADIC ADRENAL ANGIOMYOLIPOMA

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INTRODUCTION

Angiomyolipoma is a rare mesenchymal tumour derived from perivascular epithelial cells. They are most commonly seen in kidneys and other sites like liver, colon amd skin. It is very rare in adrenals. There is association with tuberous sclerosis and it can also occur as sporadic adrenal angiomyolipoma. They are almost always benign but a few can have malignant potential and can recur locally. Here we are a presenting a large ADRENAL ANGIOMYOLIPOMA probably the second largest ever reported in literature.

Case Report

A 60 year old male was admitted with complaints of dull aching abdomen pain more in the right hypochondrium for 7days. History of vomiting 2-3 episodes per day was present. History of similar episode was noted 3months ago. He is a known diabetic and hypertensive for the past 5years on regular medications. On examination, he was obese and his vitals were stable. Abdominal examination revealed a vague mass in roght hypochondrium, margins illdefined, not able to insinuate finger below the right costal margin and not independently ballotable.

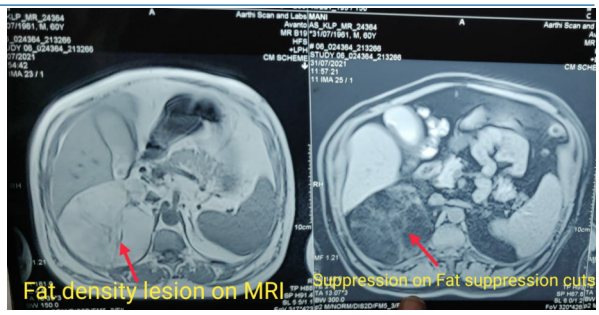
Mild right hypochondrial tenderness was present. CECT abdomen revealed a non enhancing well defined fat density lesion of size 10.8*10.8*9.8cm in right suprarenal region. A calculi of size 1.1cm in neck of gallbladder with mild pericholecystic inflammatory changes. MRI abdomen revealed well defined large predominantly fat containing lesion in right adrenal gland with T1 hyperintensity and T2 hypointensity. Marked suppression was noted in fat suppression images. His hormonal workup was within normal limits.

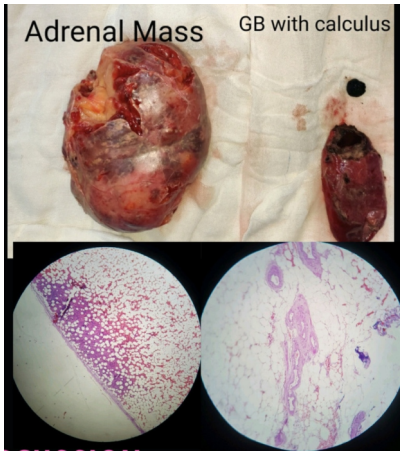
Patient was planned for right adrenalectomy with cholecystectomy.

The following were the major surgical steps:

- Right subcostal incision
- Cattell Braasch maneuver
- Kocherisation of duodenum
- Subtotal cholecystectomy (due to cholecystitis)
- Defining the right adrenal mass and it's margins
- Dissection of tumor from IVC medially amd liver bed superiorly
- Ligation of adrenal vein
- Adrenalectomy

Post operative period was uneventful and the patient was discharged on POD-10.





DISCUSSION

Angiomyolipoma is a rare mesenchymal tumour derived from perivascular epithelial cells. They are most commonly seen in kidneys and other sites like liver, colon amd skin.

It is very rarely seen in adrenal gland with less than 20 cases in literature.

There is association with tuberous sclerosis and it can also occur as sporadic adrenal angiomyolipoma.

Presence of abnormal blood vessels, smooth muscle cells and adipocytes will confirm the diagnosis of adrenal angiomyolipoma.

They are almost always benign but a few can have malignant potential and can recur locally.

Age: Tuberous sclerosis- third and fourth decade of life

Sporadic cases– older patients

Gender : Surgical series: 4:1 female to male ratio

Imaging detection- no apparent sex predilection

Most surgical series report 4 times as many sporadic angiomyolipomas as angiomyolipomas associated with tuberous sclerosis

Site: Cortex or medulla of kidney, liver, retroperitoneal soft tissue with or without renal attachment.

Clinical features- tuberous sclerosis- routine screening detected

sporadic– larger, flank pain, hematuria, palpable mass, retroperitoneal hemorrhage

CT– preoperative diagnosis- high fat content- distinctive pattern

Macroscopic Appearance: Well demarcated, not encapsulated may be locally infiltrative

- Size- 1cm to 20cm diameter. Symptoms above 4 cm
- Golden Yellow to pink tan depending on the relative proportions of the various tissue components
- Most solitary but multiple tumors may be present
- Very rarely prominent cystic or pseudocystic change
- Smooth muscle predominant - leiomyoma.

Immunophenotype:
- melanocytic markers- HMB45, melan A and microphthalmia transcription factor
- Smooth muscle markers – smooth muscle actin, calponin
- CD68, S100 protein, estrogen and progesterone receptors an desmin- positive
- Epithelial markers are always negative

Genetic Profile:
- TSC 1 gene – 9q34 – hamartin
- TSC 2 gene – 16p13 – tuberin – GTPase-activating protein for RAS related protein 1 and Rab protein 5

Angiomyolipoma shows LOH of variable portions of the TSC2 gene locus in both sporadic and tuberous sclerosis associated tumors. LOH of the TSC1 gene is occasionally seen

Differential diagnosis:
- Predominant lipomatous component:
- Lipoma – extensive sampling
- Liposarcoma
- Predominant smooth muscle component:
- Leiomyoma
- Predominant vascular component:
- Vascular malformation
- Epithelioid angiomyolipoma

Large adrenal angiomyolipoma should be removed even if asymptomatic to avoid the risks of spontaneous rupture due to presence of abnormal elastin and poor vascularity.