ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 03 | March - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

ADRENAL ANGIOMYOLIPOMA: A SURGICAL RARITY

General Surgery

Dr. Mikail Merchant* (MBBS): General Surgery House Medical Officer. Dept. of General and Minimal Access Surgery, Sir H.N. Reliance Foundation Hospital and Research Centre, Mumbai, India. *Corresponding Author

Dr. Sushrut Tendulkar (MBBS): General Surgery DNB Resident, Dept. of General and Minimal Access Surgery, Sir H.N. Reliance Foundation Hospital and Research Centre, Mumbai, India

Dr. Abhijit Thakur (MBBS, MS): Consultant General and Laparoscopic Surgeon, Dept. of General and Minimal Access Surgery, Sir H.N. Reliance Foundation Hospital and Research Centre, Mumbai, India.

Dr. Seelam Lakshmi Mounika (MBBS): General Surgery DNB Resident, Dept. of General and Minimal Access Surgery, Sir H.N. Reliance Foundation Hospital and Research Centre, Mumbai, India

ABSTRACT

Adrenal Angiomyolipoma's are extremely rare forms of lipomatous tumours of adrenal gland. They may go undetected for years without symptoms. The patient may become symptomatic, once the tumour has gained a significant size, mainly back pain. We are reporting our case which is a one of a kind benign Adrenal tumour. Adrenal Angiomylipomas are found as incidentalomas in patients, generally presenting with mild to no symptoms, leading to a long delay before they are diagnosed. However, early diagnosis is key, not only to alleviate symptoms but to also prevent a future surgical disaster due to rupture causing cardiovascular shock if large enough. Our patient presented with dull aching flank pain without any systemic symptoms. The diagnosis requires a multifocal approach to be confirmed including hormonal assay and radiological testing whereas following a "clinical only" approach maybe insufficient and often mislead a clinician from establishing an accurate diagnosis. These patients could be offered laparoscopic resection at specialized centres with minimal risk of complication and early uneventful recovery.

KEYWORDS

Angiomyolipoma, Myelolipoma, Lipomatous tumours, Adrenalectomy

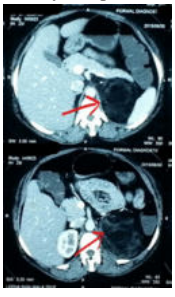
INTRODUCTION:

Angiolipomas are Benign Lipomatous tumours found most commonly in the kidneys, however in extremely rare cases may be found in the Adrenal glands. These tumours are derived from mesenchymal origin, mainly from perivascular epithelioid cells and generally contain mature fat, bone-marrow-like hematopoietic cells, along with blood vessels and muscle cells.[1]

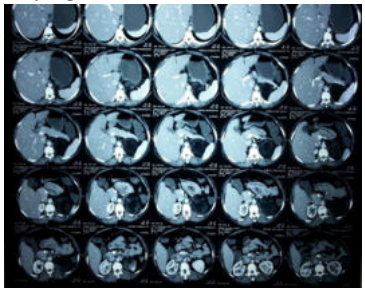
These tumours are generally asymptomatic[2] and are found incidentally on radiological imaging but sometimes may be associated with endocrine pathologies such as Cushing's syndrome, 21 -hydroxylase deficiency or Conn Syndrome[3]. The Incidence rate of these tumours is as low as 0.08 to 0.4% and there have been very few cases documented in medical literature till date[4]. Below we present a symptomatic case with flank pain and no other co-morbidities.

Case Report:

A 26 year old female patient presented with intermittent left side flank pain for last 6 months which was dull aching and non-radiating. She had no other systemic symptoms and no past history of any co-morbidities. Clinical examination was within normal limits and revealed no abnormality. She underwent ultrasound examination of the abdomen and pelvis, which revealed a 8 cm x 6 cm mass lesion in left adrenal region while rest of sonography was normal. Computed tomography (CT) of abdomen revealed a left adrenal mass of around 8 cm x 6 cm x 6 cm with heterogeneous internal composition including macroscopic fat with few interspersed solid appearing elements. No calcification was noted. A CT differential diagnosis was established as adrenal myelolipoma or angiomyolipoma.



(Fig 1: Axial CT focused)

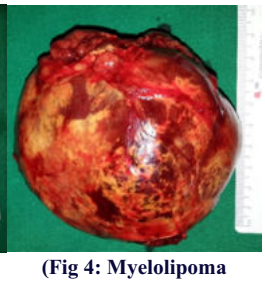


(Fig 2: Axial CT Film)

She was investigated for secreting adrenal mass by cortisol suppression test, serum aldosterone levels, plasma renin activity and plasma metanephrine and nor-metanephrine levels, all of which were normal.

In view of increased frequency of left flank pain, a decision was made to go ahead with resection of the tumour. She then underwent laparoscopic left adrenalectomy.

(Fig 3: Myelolipoma Resected Specimen)



(Fig 4: Myelolipoma Specimen focused)

The surgery was uneventful and histopathology reports confirmed diagnosis of Angiomyolipoma. Patient was discharged on the 3rd post-operative day. After following all post-discharge advise and undergoing complete post-operative recovery, the patient remains symptom free till date.

DISCUSSION:

Angiomyolipomas in its first description was explained by Edgar von Gierke[5] in 1905 with the term "Myelolipoma" first being used by French pathologist Charles Obenling[6] in 1929. These are Lipomatous tumours of mesenchymal origin and are commonly seen in the kidney but can also be seen in other organs as extra-renal manifestations such as liver, spleen, lungs, retroperitoneum, bone, and ovaries[7]. Adrenal Angiomyolipomas are the most rarest form of Myelolipomas with its Incidence ranging from 0.08% to 0.4%, and less than 300 cases were reported in the literature before 2000[8]. The largest reported case of Adrenal Angiomyolipoma was 31 × 24.5 × 11.5 cm and weighing in at 6 kg. These tumours are generally non-secreting tumours, however a single case of secreting adrenal Angiomyolipoma has been reported in literature[9].

It is widely thought that Myelolipomas occur in response to stressful stimuli such as necrosis, inflammation or infection, theorizing it to be primarily caused by adrenocortical cell metaplasia[10]. While most of these tumours are asymptomatic and often detected as Incidentalomas, they may sometimes cause symptoms, especially pain. Diagnosis requires a multifocal evaluation with radiological imaging, CT scan being the gold standard mode of diagnosis showing the highest sensitivity[11], exposing Myelolipomas as adrenal lesions with well circumscribed fat containing components. Occasionally in cases, they may show punctate calcifications[12] and sometimes even haemorrhagic tissue in larger lesions[13]. Other modalities that may be used for imaging are ultrasonography and MRI.

Treatment of Adrenal Angiomyolipoma's depends on symptom development and probability of the tumour to cause future complications such as cardiovascular shock due to rupture and haemorrhage of large Myelolipoma[14]. Surgical resection is key for larger tumours which leads to highly improved future prognosis, ultimately causing regression of all symptoms and risk for future complications. In our case, a laparoscopic left adrenal resection was performed with complete Myelolipoma resection. Despite being majorly a non-secreting tumour, the patient should always be worked up to look for a secreting tumour with hormonal panels consisting off Cortisol Suppression test, Serum Aldosterone levels, plasma Renin activity and plasma Metanephrine and Nor-Metanephrine levels due to existence of a documented case of secreting Myelolipoma tumour in medical literature[9].

CONCLUSION:

Adrenal Angiomyolipoma's are rare benign lipomatous tumours which may be found as incidental findings or may produce mild symptoms, however may be more sinister than they appear if large enough due to their ability to cause dire surgical emergencies by itself in case of rupture causing cardiovascular shock. Imaging and hormonal assay could clinch the diagnosis. Smaller lesions less than 5 cm and asymptomatic could be observed but larger lesion more than 5 cm can be offered laparoscopic resection. Laparoscopic resection of these tumours is safe, simple and holds minimal risks and complications if performed well at equipped specialized centres.

REFERENCES:

1. Flum A.S., Hamoui N., Said M.A. An update on the diagnosis and management of renal angiomyolipoma. *J Urol*. 2015;111(1):21–28.
2. Gierke E. Uber knochenmarksgewebe in den nebenniene. Beitn Path Anat 1905; 7:311-325.
3. Oliva A, Duarte B, Hammadeh R, Ghosh L, Baker RJ. Myelolipoma and endocrine dysfunction. Surgery 1 988; 103:711-715
4. J. Settakorn, C. Sirivanichai, S. Rangdaeng, and B. Chaiwun, "Fine-needle aspiration cytology of adrenal myelolipoma: case report and review of the literature," *Diagnostic Cytopathology*, vol. 21, pp. 409–412, 1999.
5. Gierke E. Uber knochenmarksgewebe in den nebenniene. Beitn Path Anat 1905; 7:311-325.
6. Oberling C. Les formations myelolipomatouses. Bull Cancer 1929; 18:234-246.
7. D'Antonio A., Caleo A., Caleo O. Monotypic epithelioid angiomyolipoma of the adrenal gland: An unusual site for a rare extrarenal tumor. *Ann Diagn Pathol*. 2009;13(5):347–350.
8. J. Settakorn, C. Sirivanichai, S. Rangdaeng, and B. Chaiwun, "Fine-needle aspiration cytology of adrenal myelolipoma: case report and review of the literature," *Diagnostic Cytopathology*, vol. 21, pp. 409–412, 1999.
9. A. Brogna, G. Scalisi, R. Ferrara, and A. M. Bucceri, "Giant secreting adrenal myelolipoma in a man: a case report," *Journal of Medical Case Reports*, vol. 5, article 298, 2011.
10. A. Meyer and M. Behrend, "Presentation and therapy of myelolipoma," *International Journal of Urology*, vol. 12, no. 3, pp. 239–243, 2005.
11. P. J. Kenney, B. J. Wagner, P. Rao, and C. S. Heffess, "Myelolipoma: CT and pathologic features," *Radiology*, vol. 208, no. 1, pp. 87–95, 1998.
12. Craig WD, Fanburg-smith JC, Henry LR et-al. Fat-containing lesions of the retroperitoneum: radiologic-pathologic correlation. Radiographics. 29 (1): 261-90
13. Routhier JR, Woodfield CA, Mayo-smith WW. AJR teaching file: fat-containing retroperitoneal mass presenting with acute flank pain. AJR Am J Roentgenol. 2009;192 (6): S122-4.
14. C. Russell, B. W. Goodacre, E. Van Sonnenberg, and E. Orihuela, "Spontaneous rupture of adrenal myelolipoma: spiral CT appearance," *Abdominal Imaging*, vol. 25, no. 4, pp. 431–434, 2000.

Footnotes:

- Funding: Not required
- Conflict of interest: None to declare
- Ethical approval: Pre-obtained from Institutional ethics and research committee. Informed Consent was taken prior to carrying out this study.