

## Myelolipoma - A Rare Benign Adrenal Tumour: A case report

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### Abstract

Myelolipoma is a rare benign hormonally inactive adrenal neoplasm containing mature adipose tissue and a variable amount of haematopoietic elements. Their mostly small size, unilateral site and clinical quiescence are accountable for predominantly incidental disclosure. A symptomatic case of myelolipoma was presented in our hospital with generalized body swelling, body rash and intermittent dull abdominal pain. He had H/O taking steroid for last 10 yrs to increase body weight and for better feeling. All laboratory investigations were found within normal limit and he was referred to our department for radiological investigations.

Plain X-ray, USG and CT scan of abdomen was done which revealed an well capsulated circumscribed highly echogenic, fat density soft tissue mass in right lumbar region which was suspected as suprarenal mass possibly myelolipoma and it was confirmed by FNAC as myelolipoma of right adrenal gland. : 34-36]

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### Introduction

Myelolipoma is a rare benign non- functioning tumour composed of mature adipose tissue and a variable amount of haematopoietic elements. Most lesions are small and asymptomatic, discovered incidentally during autopsy or on imaging studies performed for other reasons. So the tumours are also termed as “incidentaloma”<sup>1</sup>. Although they are commonly found in the adrenal glands, extra-adrenal myelolipoma in the retroperitoneum are rare but documented<sup>2</sup>. Myelolipoma is reported as being .08 to 4% at autopsy<sup>3</sup>. With the development and improvement of non-invasive imaging modalities its incidental detection has become more common, reaching upto 7% of the adrenal masses<sup>4</sup>. A study reported that male to female ratio is 2:3<sup>5</sup>. Adrenal myelolipoma presents as a site of extramedullary haematopoiesis. Although great numbers of

incidentally discovered lesions are small and asymptomatic, reports are not infrequent for cases of large symptomatic lesions of adrenal myelolipoma.<sup>6</sup> Though the tumors are hormonally inactive, management of the tumours are depended on the size of the tumours and presence of abdominal symptoms. Symptomatic lesions must be treated with the advent of minimal invasive surgery<sup>7</sup>.

We report one case of symptomatic myelolipoma where diagnosis was made on the basis of radiological features and image guided fine needle aspiration cytology.

### Case report

A 48 years old male, known case of diabetes mellitus was presented with generalized body rash for 1 year, generalized body swelling for 6 months and intermittent

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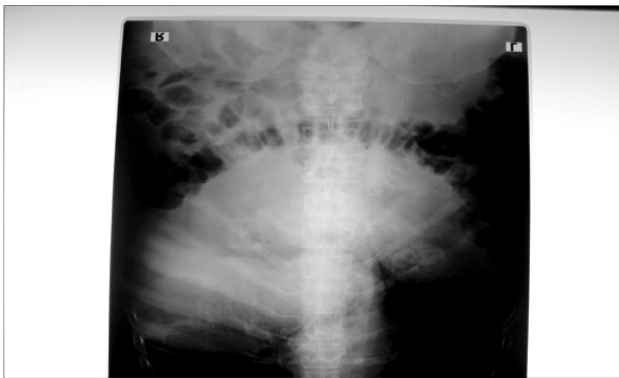
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**Contributions by authors:** All authors contributed in the management of case as well as involved in manuscript writing.

dull abdominal pain for 4 months. He had a H/O taking steroid for last 10 years which has no significant clinical cause but to increase body weight and for better feeling. Physical examination revealed generalized body swelling, skin rash, abdominal distension and mild tenderness on right hypochondriac region. He was clinically diagnosed as a case of iatrogenic cushing syndrome with contact dermatitis.

Routine blood and urine test including liver and kidney function test, blood sugar and lipid profile were within normal limit. Special laboratory investigations like S. electrolyte, serum ACTH and 24 hours urinary cortisol were investigated. Features were within normal limit. His chest X-ray and ECG findings were unremarkable.

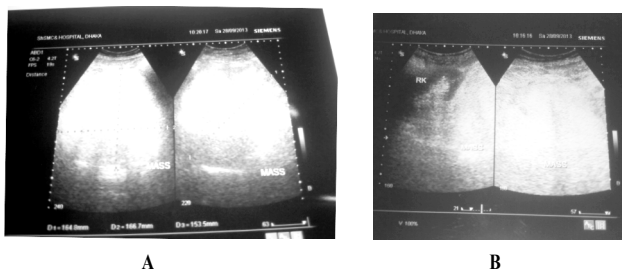
**Plain X-ray Abdomen:** showed bulging of flanks with soft tissue density shadow having internal lucencies in right lumbar region displacing the bowel shadows inferiorly.(figure-1)



**Fig-1:** Plain X-ray abdomen showing soft tissue density shadow having internal lucencies in right lumbar region displacing the bowel loops inferiorly.

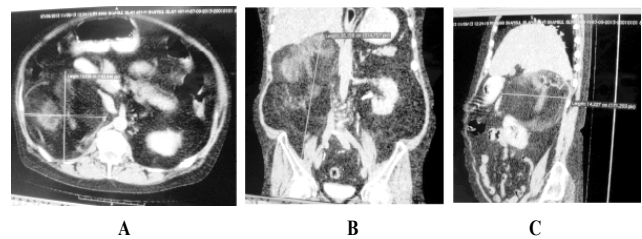
**USG findings revealed:**

A highly echogenic, heterogenous well circumscribed mass measuring about 14 cm x 13 cm x12 cm seen in right lumbar region at upper pole of right kidney which was separated from liver and right kidney was displaced inferiorly. (figure-2)



**Fig-2:** (a) & (b) USG of abdomen showing highly echogenic mass in right lumbar region separated from right kidney

Pre and post contrast CT scan of upper abdomen showed: A fairly large encapsulated heterogeneous density mass predominantly hypodense of fat density (-20 to -125 HU) without any calcification having fibrous strands within it measuring 14.9 cm x12.5 cm x12.6 cm was seen in right lumbar region at paravertebral location postero-superior to right kidney abutting against the inferior surface of liver. Vertically it extends from D11 to SI vertebrae displacing right kidney and bowel loops antero-inferiorly and compressing IVC & renal vessels. After IV contrast mild enhancement of relatively solid components were noted. Overall intra abdominal (mesenteric, omental, retroperitoneal) and subcutaneous fat content was more than normal (figure-3).



**Fig-3:** (a) axial (b) coronal & (c) Sagittal CT scan of abdomen showing an ncapsulated fat density mass having fibrous strands in right paravertebral location displacing right kidney & bowel loops.

Our radiological diagnosis was right suprarenal mass, possibly myelolipoma. D/D were – Renal angimyolipoma, retroperitoneal myolipoma. FNAC revealed moderately cellular smear containing mature adipose tissue fragments and immature haemopoietic cells comprising of megakaryocytes, granulocytic and erythroid cells and moderate number of lymphocytes resembling aspirated bone marrow. No malignant cell or cellular atypia was seen. Findings were consistent with adrenal myelolipoma.

**Discussion**

Myelolipoma have been somewhat of medical curiosity as they are uncommon, benign and usually asymptomatic. The term “myelolipoma” was used by Oblerling in 1929, though the lesion was first reported by Gierke in 19058. Since then more than 200 cases have been reported in the literature<sup>9</sup>. Most of them were measuring 1-2 cm in diameter, only 3 cases with tumours weighing more than 3 kg were published<sup>10</sup>. Usually unilateral, only 4 cases of bilateral tumours have been reported<sup>11</sup>. The usual age of diagnosis is the seventh decade of life<sup>12</sup>. These tumours are composed of mature fat and haemopoietic tissues including erythroid. Myeloid, megakaryocyte types and often lymphocytes. Unlike the bone marrow, myelolipoma lacks bony spicules and reticulocytes. The most accepted theory of etiopathogenesis of adrenal myelolipoma is metaplasia of reticuloendothelial cells of blood capillaries in adrenal gland in response to stimuli such as necrosis, infection, stress or long term ACTH stimulation<sup>13</sup>. Though usually asymptomatic symptoms may occur such as flank pain

resulting from mechanical compression of large tumour, retroperitoneal hemorrhage and tumor necrosis. There are reported cases of endocrine dysfunction with myelolipoma including conditions such as Cushing's syndrome, Conn's syndrome and congenital adrenal hyperplasia<sup>13</sup>.

**Grossly:** It is pseudo encapsulated due to compression of the adrenal tissue. It has smooth surface and it is yellow in colour with varying amounts of red-brown hematopoietic elements.

**Microscopically:** The tumor entails a mixed component of fat cells and bone marrow elements of myeloid, erythroid and megakaryocytic cells in varying degrees. No malignant features are present<sup>14</sup>.

The characteristic fatty component of the tumor can be detected with the aid of USG, CT Scan and MRI. Myelolipoma is highly suggested if the USG shows a highly echogenic mass in lumbar regions. CT findings are more confirmatory and shows low density mass with areas of interspersed higher attenuated myeloid tissue with or without specks of calcifications<sup>15</sup>. On Magnetic resonance imaging the predominantly fatty areas demonstrates increased signal intensity on T1-weighted images and moderate hyperintensity on T2-weighted images. T2-weighted image is more complex because of the admixture of marrow and soft tissue elements within the fat. The imaging appearance is altered by the presence of hemorrhage. So CT is the most accurate method for evaluation of myelolipoma and image guided FNAC can be utilized to confirm the diagnosis. Management of myelolipoma should be done on a case to case basis. Patient with lesion <10 cm should be observed closely for 1-2 years. If patient is asymptomatic and there is no tumour growth then the follow-up can be done at increasing time intervals. If patient starts exhibiting symptoms and there is tumour growth then surgery is recommended. Large asymptomatic tumours > 10 cm found incidentally should be excised due to risk of life threatening shock secondary to retroperitoneal hemorrhage. There are case reports of contraletaral myelolipoma after resection of primary lesion. Follow-up after adrenalectomy for unilateral myelolipoma is recommended<sup>16</sup>.

### Conclusion

We have reported a rare case of giant adrenal myelolipoma

with a rare association of features of Cushing's syndrome apparently suggesting hormonally active tumour but it was due to long term intake of steroid and not caused by the tumour itself. The tumour of our patient was found non-functioning and his general condition was improved with conservative treatment and he was discharged with advice.

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