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Case Series

Pathology Section

Adrenal Myelolipomas: An Interesting Case Series of the Silent Tumours with a Diagnostic Dilemma

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ABSTRACT

Adrenal myelolipomas are a variety of rare and endocrinologically non functional benign neoplasms of the adrenal gland. They are usually composed of haematopoietic elements with interspersed areas of fatty tissue, thus, resembling bone marrow tissue. These tumours are usually detected as an incidentaloma during routine scans on suspicion of other diagnoses and are mostly silent clinically. They have always been a subject of diagnostic dilemmas for the clinicians, as they are usually asymptomatic and benign, but are often operated upon in situations where a malignancy cannot be excluded. Differential diagnosis for these tumours include adrenal adenoma, adrenal carcinoma, retroperitoneal liposarcoma and renal angiomyolipomas. A careful histopathological examination usually settles the diagnosis. Usually, the tumours are unilateral with right sided preponderance and are small in size. Here, the authors report three interesting cases, including two females and one male, of adrenal myelolipomas with varied presentations with two cases presenting with large size tumours and in one case the adrenal tumour was located on left side.

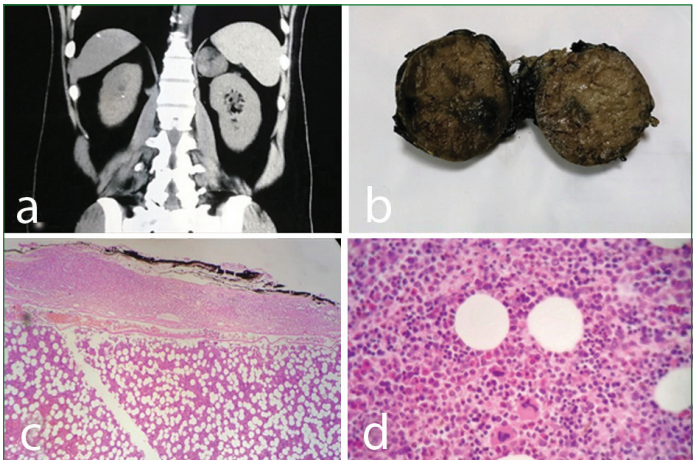
Keywords: Adrenal tumour, Incidentaloma, Non functional tumour

CASE SERIES

Case 1

A 48-year-old male patient presented in the Emergency Department with complaint of severe pain in the right upper abdomen for the past three months. Abdominal imaging revealed gall stone disease as a cause of the patient's symptoms. However, the Computed Tomography (CT) scan of abdomen also revealed an incidental finding of a 4×4 cm mass near the upper pole of left kidney seen arising from the left adrenal gland [Table/Fig-1a]. Due to adrenal origin of tumour test to measure the urinary catecholamines and metanephrines levels were done which were normal and hence the mass was judged to be a non functional tumour. Thereafter, the excisional biopsy of the lesion was done and the mass was subjected to histopathological examination. On gross appearance, the mass measuring 4 cm in diameter was globular and capsulated and had yellow and red areas on cut surface [Table/Fig-1b].

Microscopic examination showed a well-circumscribed lesion attached to compressed adrenal gland at one pole, comprising of predominantly mature cellular haematopoietic tissue admixed with fat cells [Table/Fig-1c,d]. The haematopoietic tissue comprised of all three lineages (erythroid, myeloid, megakaryocytic) and lymphoid aggregates. The lesion was partly surrounded by fibrous pseudo capsule. Focally, haemorrhage was also seen. These features were in consonance with the histologic features of adrenal myelolipoma and hence a diagnosis of left adrenal myelolipoma was made. The patient was asymptomatic two years postsurgery.



[Table/Fig-1]: Case 1: a) CT appearance of myelolipoma at upper pole of left kidney; b) Gross picture with yellow and red areas; c) Histopathologic picture showing haematopoietic tissue admixed with fat along with compressed peripheral adrenal tissue. (4x H&E); d) Histopathologic picture showing haematopoietic tissue with megakaryocytic precursors (10x H&E).

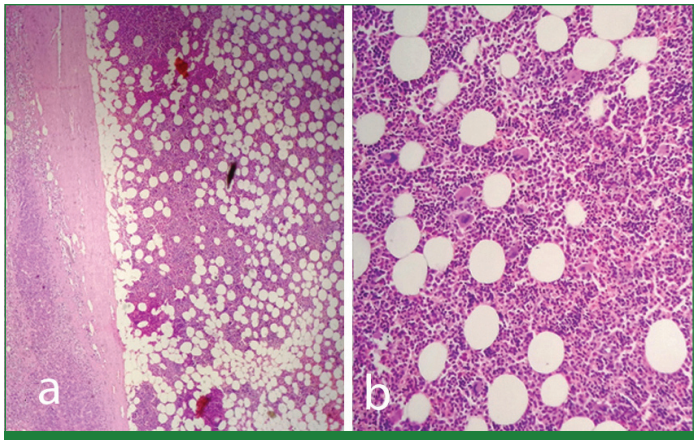
Case 2

This was a case of 55-year-old normotensive housewife, who had presented with a chronic pain in right flank of abdomen for the past eight months. Pain was intermittent and colicky in nature with no relationship to meals. The patient had received treatment for pain abdomen from multiple facilities but had persistence of symptoms. She was subjected to abdominal imaging including CT scan on suspicion of gall stone disease. However, CT was suggestive of a well-defined round lesion in right suprarenal location indicating a right adrenal mass. Urinary metanephrines and catecholamines were normal. The patient was then subjected to excisional biopsy of the lesion. Grossly, the tumour was 13.5×10.5×6 cm, was partly encapsulated externally and on cut surface showed solid bright yellow areas with foci of brown haemorrhagic areas.

Microscopically [Table/Fig-2a,b], a well-circumscribed encapsulated lesion comprising predominantly of mature adipose tissue with interspersed islands of haematopoietic cells showing erythroid, myeloid and megakaryocytic cells accompanied by lymphoplasmacytic cells were seen. Lymphoid follicles were also seen. Areas of haemorrhage were noted. Compressed adrenal gland tissue was seen attached to the lesions at the periphery. Finally, a diagnosis of adrenal myelolipoma was made. The patient was doing well during three months follow-up postsurgery.

Case 3

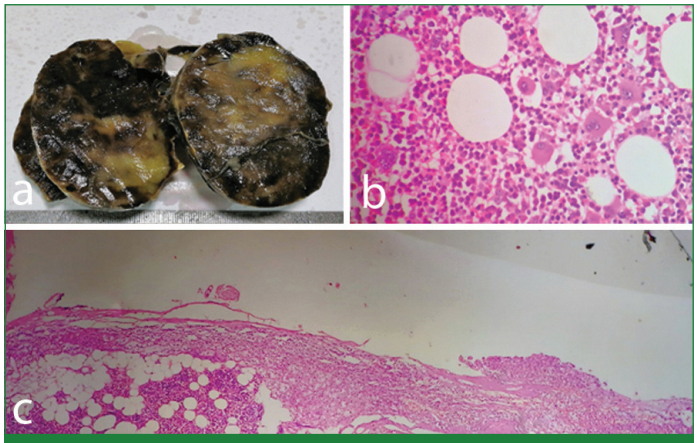
A 42-year-old normotensive female patient came with a history of chronic right abdominal pain which was dull and dragging in character and constant in nature along with history of feverishness and lethargy for the last one year. For the past six months, the patient was also complaining of irregular menses. The ultrasound abdomen revealed right adnexal mass with hydrosalpinx. The CT abdomen was performed to rule out genitourinary tuberculosis.



[Table/Fig-2]: Case 2: a) Histopathologic picture showing haematopoietic tissue admixed with fat along with compressed peripheral adrenal tissue (4x H&E); b) Histopathologic picture showing haematopoietic tissue with megakaryocytic precursors (10x H&E).

However, the scan revealed right adnexal cyst with hydrosalpinx and another mass in the right suprarenal region of size 8×5×5 cm. This suprarenal mass showed hypodense areas with CT attenuation values suggestive of fat. A diagnosis of renal angiomyolipoma was made. Thereafter, the patient underwent excisional biopsy of both the masses. On gross examination, during histopathological evaluation, the mass was received as an encapsulated globular tissue piece of size 8×6.5×5.5 cm [Table/Fig-3a]. Outer surface was smooth with a focal area of ruptured capsule. On cut surface, the mass had a variegated appearance containing soft fat like areas with interspersed areas of haemorrhage.

The microscopic examination [Table/Fig-3b] revealed adrenal gland with mature adipose tissue and islands of haematopoietic cells. Trilineage hematopoiesis was noted along with megakaryocytes [Table/Fig-3c]. Areas of hyalinisation were noted in between the adipocytes along with cystic changes. Hence, a final diagnosis of adrenal myelolipoma was reached. One month postsurgery, the patient was ambulatory and doing well.



[Table/Fig-3]: Case 3: a) Gross picture with yellow and haemorrhagic areas; b) and c) Histopathologic picture showing hematopoietic tissue with megakaryocytic precursors (40x H&E).

DISCUSSION

Adrenal myelolipomas are a variety of rare and endocrinologically non functional benign neoplasms of the adrenal gland. They have always been a subject of diagnostic dilemmas for the clinicians as they are usually asymptomatic and benign but are often operated upon in situations where a malignancy cannot be excluded. The first reported cases of this tumour, date back to early 20th century, when Gierke E described this tumour in 1905 [1] and Oberling C coined the term “myelolipoma” in 1929 [2].

Prior to the turn of 21st century, these tumours were very rare and less than 300 cases in all, had been reported [3]. However, with the advent of the modern imaging techniques having greater resolution, the detection rate of these tumours picked up and now these tumours have an overall incidence of 0.08-0.2% and constitute about 10-15% of all incidental adrenal masses [4].

Grossly, these tumours are yellow to red in colour with areas of haemorrhage depending upon the proportions of fat and haematopoietic tissue. Microscopically, these tumours are composed of myeloid elements including haematopoietic tissue and adipose tissue in varying proportions, but are devoid of the reticular sinusoids and bony spicules seen in bone marrow [5]. The larger tumours may show haemorrhage, calcification and necrosis. All of our three cases showed haemorrhage. The aetiopathogenesis of these tumours is unclear. However, they are thought to arise from the metaplasia of reticular endothelial cells of blood capillaries in adrenal gland in response to stress, infection or necrosis [6].

The comparative features of adrenal myelolipomas reported in some important studies are summarised in [Table/Fig-4] [3,4,7-9]. As evident from [Table/Fig-4], the tumour has no sex predilection and occurs in middle to older age group. Also, most of the patients are asymptomatic but symptoms, when present, include abdominal pain and discomfort. Occasionally, in large tumours, symptoms due to compression may be seen.

Adrenal myelolipomas are usually an incidental finding and are mostly diagnosed at the time of work-up for other suspected diseases. In the present case series also, in the 1st and 3rd case the diagnosis was reached while the detailed investigation was done for gall stone disease and adnexal mass with hydrosalpinx, respectively. However, in the 2nd case there was no alternate explanation for the patients’ symptoms and hence, the symptoms could be attributed to the adrenal mass.

Medical literature reveals the predominance of tumour in the right adrenal gland as seen in [Table/Fig-4]. This was also the case with the 2nd and 3rd cases. However, the 1st case is an exception in this regard as it was located in left adrenal gland.

Another interesting observation from this case series is that, as mentioned in literature, these tumours are small in size [3]. However, in the present case series, in two cases, the tumours were larger and measured about 13 cm and 8 cm in the longest dimension. The probable explanation could be the long presenting history of symptoms. Although, a greater size poses a threat for malignant transformation, in the present study all the cases were benign.

S. No.	Author	Year	No. of cases	Sex	Age (years)	Side	Size in largest dimensions	Presentation	Management
1.	Bhansali A et al., [7]	2003	6	3 M 3 F	26-60	5 Right 1 Left	5-25 cm	2 Asymptomatic 4 Pain abdomen	Surgical resection
2.	Wani N et al., [4]	2010	2	2 M	48-52	2 Right	10-16 cm	1 Asymptomatic 1 Pain abdomen	Surgical resection in 1 patient
3.	Hsu SW et al., [8]	2012	6	3 M 3 F	44-51	5 Right 1 Left	6-16.5 cm	3 Asymptomatic 3 Pain abdomen	Surgical resection
4.	Nabi J et al., [3]	2013	1	M	63	Right	6.5 cm	Pain right abdomen	Surgical resection
5.	Sachan A et al., [9]	2018	1	M	40	Right	10 cm	Pain right abdomen	Surgical resection
6.	Present case series	2021	3	1 M 2 F	42-55	2 Right 1 Left	4-13 cm	1 Asymptomatic 2 Symptomatic	Surgical resection

[Table/Fig-4]: Comparative features of adrenal myelolipomas reported in some studies [3,4,7-9].

These tumours are usually non secretory in nature and hence, are non functional [3] but reports of increased adrenal gland secretion leading to hypertension are known. In the present study, all cases were non functional.

Differential diagnosis for these tumours include adrenal adenoma, adrenal carcinoma, retroperitoneal liposarcoma and renal angiomyolipomas [10]. A careful histopathological examination usually settles the diagnosis. Adrenal adenoma and adrenal carcinoma can be differentiated morphologically with classical distinguishing features, whereas tumours having adipose tissue component pose a diagnostic dilemma sometimes. Angiomyolipoma shows smooth muscles and characteristic vascular components along with adipose tissue, in contrast to predominant lipomatous component in liposarcoma. Myelolipoma however have haematopoietic tissue along with adipose tissue and extensive sampling is, sometimes, required to establish the presence of haematopoietic tissue.

Treatment options for adrenal myelolipomas include either conservative/expectant management for small lesions. Active/surgical treatment is usually reserved for cases with: (a) small-sized tumours that are increasing in size on subsequent radiological imaging; (b) symptomatic lesions that are 6 cm or larger; (c) tumours associated with haemorrhage and; (d) situations when malignancy cannot be excluded [11].

CONCLUSION(S)

Adrenal Myelolipomas are uncommon, benign adrenal tumours which are mostly non functional and asymptomatic and are usually detected incidentally on routine scans. However, such tumours pose a diagnostic dilemma to the clinicians and should, therefore, be subjected to careful diagnostic protocol to plan out the most appropriate treatment for each patient.

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