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Original Research Article

Giant adrenal myelolipoma - clinical spectrum and management: a single centre experience

Santosh Kumar, Ankur Mittal*, Aditya Prakash Sharma, Abhinandan Mukhopadhyay

Department of Urology, PGIMER, Chandigarh, India

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***Correspondence:**
Dr. Ankur Mittal,
E-mail: drmittal.ankur@gmail.com

ABSTRACT

Background: Adrenal myelolipomas are rare benign tumor composed of mature adipocytes and normal haematopoetic cells. Giant adrenal myelolipomas are rare clinical entities. Most of them are symptomatic. We present 15 such cases, their clinical spectrum and management.
Methods: Retrospective analysis of giant adrenal myelolipomas from a tertiary level institute. Initial diagnosis was made by computed tomography.
Results: Mean age of patient was 45.6±11 years with slightly higher female preponderance. All patients were overweight with predominantly left sided adrenal myelolipoma. Majority of them (93%) were symptomatic and presented with abdominal pain, anemia or fever. Mean size of the mass on imaging was 14 cm±6 cm, with largest lesion measuring 26 cm.
Conclusions: The article highlights the varying clinical presentations including rare emergency presentations of giant adrenal myelolipomas. A brief literature review is also presented.

Keywords: Adrenal hemorrhage, Adrenal myelolipoma, Giant, Surgical management

INTRODUCTION

Adrenal myelolipomas are rare benign tumor composed of mature adipocytes and normal haematopoetic cells. It was first described by Gierke in 1905 and later termed as myelolipomatoses by Oberling in 1929.[1,2]

These are thought to arise from metaplasia of undifferentiated stromal cells in response to stress, infection, necrosis or excessive ACTH stimulation.[3] Although majority of giant myelolipomas are symptomatic but in this era of diagnostic imaging are usually diagnosed incidentally in asymptomatic patients.

They are rarely functional still they warrant detailed hormonal work up. In this context we share our experience of managing varying clinical presentations of giant adrenal myelolipoma (>8cm) from a tertiary care centre in north India in last 10 years.

METHODS

A retrospective analysis was performed by reviewing medical records and imaging studies. Data of the available cases of giant adrenal myelolipoma between 2004 and 2015 was retrieved. Total of fifteen patients were diagnosed and treated for giant adrenal myelolipoma. Initial diagnosis was made by computed tomography. All patients underwent surgery and definitive diagnosis was made by histopathology. Descriptive statistics was used for analysis.

RESULTS

Mean age of patient was 45.6±11 years with female (53%) preponderance. All patients were overweight with the mean BMI of 29.8±2.3 kg/m^2. Only one out of 15 patients had left sided adrenal myelolipoma.

Majority of them (93%) were symptomatic and presented with abdominal pain, anemia or fever (Table 1). Mean size of the mass on imaging was 14 cm±6 cm, with largest lesion measuring 26cm. All patients had pre-operative CT images suggestive of myelolipoma.

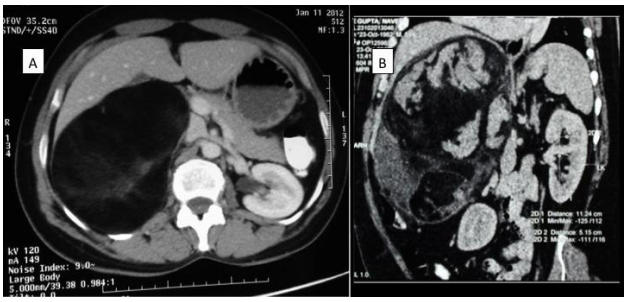

Figure 1: (A) CECT axial section showing giant adrenal myelolipomas; (B) CECT coronal section showing displacement of right kidney by adrenal myelolipoma.

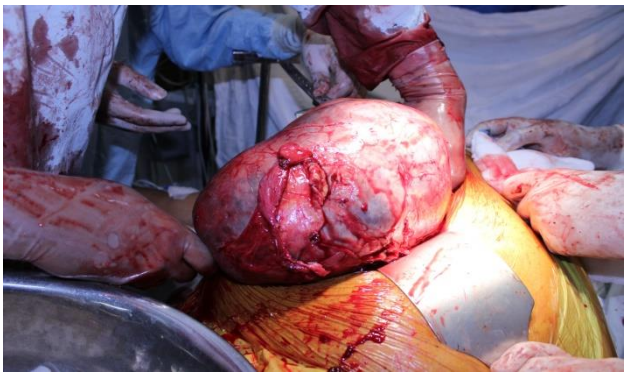

Figure 2: Intraoperative image of gaint adrenal myelolipoma.

Majority of patients had associated comorbidities which included hypertension (64.2%), idiopathic thrombo-cytopenic purpura, dyslipidaemia, diabetes, chronic anaemia, gall stones, horseshoe kidney, renal stone disease and hypothyroidism. Biochemical work up for cushing's syndrome, pheochromocytoma and aldosterone secreting adenoma was negative in 14 patients but one patient had raised serum normetanephrine levels.

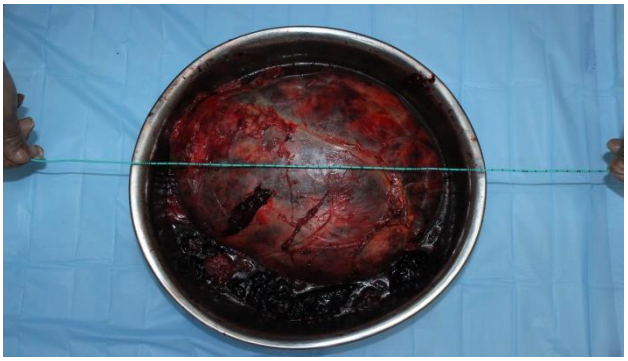

Figure 3: 26 cm tumor showing haemorrhage.

All patients underwent surgery. Laparoscopic adrenalectomy was done in 4 (27%) patients, laparoscopic adrenal sparing tumor excision was done in one (6%) patient and rest all underwent open adrenalectomy (67%). Laparoscopically the largest size tumor removed in our series had diameter of 13.7cm. Histopathology was confirmatory in all the patients. There was no recurrence in any of the patients with median follow up of 20 months (range from 1 to 120 months).

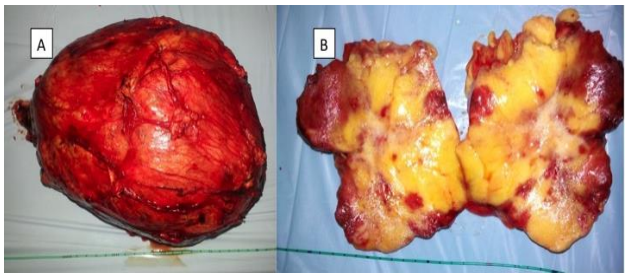

Figure 4: (A) Gross specimen of adrenal myelolipoma; (B) Cut section showing both myeloid and lipoid elements.

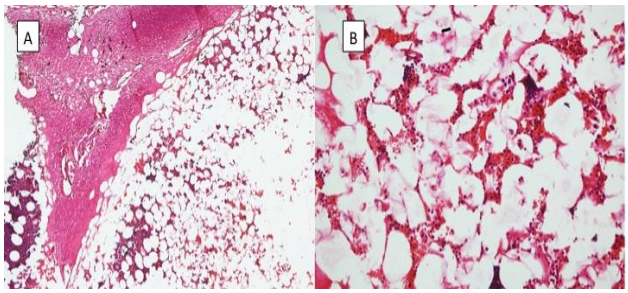

Figure 5: (A) Microscopic view of adrenal myelolipoma showing mature adipocytes; (B) Microscopic view of adrenal myelolipoma showing hematopoietic elements.

DISCUSSION

The incidence of adrenal myelolipoma varies between 0.08 to 0.2% in previous autopsy series.[4] However, it is more commonly noted in recent years due to advances in imaging modalities. Exact etiology of this tumor is unknown but as postulated it may arise from undifferentiated stromal metaplasia. They account for 3-5% all adrenal tumors and usually present in 5th to 7th decade of life.[4-6] In present series the mean age was 45.6 years. There is no sex predilection. It is usually unilateral with available literature reporting equal predilection for both the glands. However, in our series 93% tumors were on the right side with slight female predilection. Though rare, bilateral myelolipomas have been described in literature.[7-13]

Myelolipomas are usually of small size often less than 4 cm diameter. Majority of the patients are asymptomatic but it can attain very large size and cause symptoms.[14]

The term giant adrenal myelolipoma is given to tumors larger than 8cm 15. Largest tumor was described by Akamatsu and colleagues (diameter 31 cm, weight 6 kg).[15] In present series the mean size of the tumor was 14 cm and largest described was 26 cm. Majority (93%) were symptomatic (Figure 1). Adrenal gland is the most common site of myelolipomas but extraadrenal occurrence in pelvis, mediastinum, retroperitoneum and paravertebral region, renal, intra peritoneal and as nasal polyp has also been described.[16-20]

Table 1: Demographic profile of patients.

Age (years)	Sex	Presenting feature	BMI	Imaging done	Side	Tumor size (CMS.)	Management	Follow-up (months)
69	M	Abdominal pain	29.9	CECT	R	9.6X7.1X6.4	Lap adrenalectomy	5
23	F	Anaemia	28.8	CECT+ MRI	R	8.5X5.9X6.1	Lap adrenalectomy	20
58	F	Abdominal pain	25.3	CECT	R	8X5X4	Lap adrenal sparing tumor excision	27
40	M	Abdominal pain	26.2	CECT	R	12X10.5X10	Open adrenalectomy	36
35	F	Abdominal pain	28.5	CECT	R	15X15X12	Open adrenalectomy	4
53	F	Anaemia	31.1	CECT	L	13.7X8.5X6	Lap adrenalectomy	5
42	F	Abdominal pain	31.2	CECT	R	25X15X8	Open adrenalectomy	120
50	M	Fever	32.8	CECT	R	22X18X23	Open adrenalectomy	21
51	M	Incidental	30.2	CECT	R	10X10X7.6	Open adrenalectomy	26
52	M	Anaemia and spontaneous haemorrhage	30.4	CECT	R	26X12X15	Open adrenalectomy	8
35	F	Abdominal pain	32.1	CECT	R	13x9x12.5	Open adrenalectomy	32
46	F	Abdominal pain	29.6	CECT	R	9x7x4	Open adrenalectomy	120
49	M	Abdominal pain	28.5	CECT	R	16.1x10x13.6	Open adrenalectomy	1
44	M	Abdominal pain	28.8	CECT	R	14.5X9.7X14	Open adrenalectomy	1
37	F	Abdominal pain	34.3	CECT	R	8x4x3	Lap adrenalectomy	1

Most common symptom is nonspecific flank or abdominal pain due to pressure effect, intra tumoral bleed or tumor necrosis. Anemia and fever are also unusual presentations as in our series. Three of our patients presented with chronic anemia due to intra tumoral bleed and one patient presented with fever due to intra tumoral pus collection (Figure 2 and 3).

In such cases renal preservation is of utmost importance together with prevention of injury to major vessels. It may also present as hypertension, retroperitoneal haemorrhage or spontaneous rupture.[21,22] Chance of rupture increases if size becomes more than 7cm. The association of myelolipoma with obesity, hypertension, chronic disease, and malignancies has been described. In our series all the patients were overweight with 64.2% patients having hypertension. Surprisingly two patients with hypertension had decreased requirement of antihypertensives post operatively.

The probable explanation for it may be release of pressure effect over kidney and vessels or relief of stress and pain post-surgery. Other disorders found in patients were dyslipidemia, diabetes, anaemia, hypothyroidism,

gall stones and renal stones. Two of our patients had idiopathic thrombocytopenic purpura and horse shoe kidney respectively.

Adrenal myelolipoma is hormonally inactive in general but it may be associated with overproduction of dehydroepiandrosterone-sulphate (DHEAS), congenital adrenal hyperplasia (CAH) caused by 21-hydroxylase deficiency, congenital adrenal 17 α-hydroxylase deficiency, Cushing disease, Conn's syndrome, adrenal insufficiency, and pheochromocytoma. Some of these tumors coexist with adrenal adenomas, which may in part account for the hormonal activity.[23]

Overall, more than 25 cases of endocrine dysfunction associated with myelolipoma have been reported in literature. Cushing and CAH appears to be most commonly described endocrine associations. In our series one patient with hypertension and increased serum normetanephrine was diagnosed as functional myelolipoma. To our knowledge it is the third case reported so far. Large tumors have been reported to be associated with functionality more often however in our series majority of the large tumors were nonfunctional.[24]

Diagnosis of myelolipoma can be suspected by ultrasound in which the lesion has mixed hyperechoic and hypoechoic areas due to presence of both fat and myeloid cells. By modern imaging like Computerised tomography [CT] and Magnetic resonance imaging (MRI) it can be diagnosed with certainty in more than 90% of cases. Myelolipoma often has a discrete capsule and appears as well-delineated heterogeneous masses with regions of less than -30 Hounsfield units that correspond to low-density mature fat in CT scan.

In MRI, fat tissue has high signal intensity in both T1 and T2 images, whereas myeloid tissue has low signal intensity in T1 and moderate signal intensity in T2 images. Sometimes loss of signal in out of phase images and suppression of signal on T2 fat saturation images might help. Other fat containing tumors like teratoma, lipoma, liposarcoma and rarely angiomyolipoma, mass-forming extramedullary hematopoiesis and adenoma should be kept in mind as differential diagnosis. If there is any suspicion of malignancy then FNA can diagnose myelolipoma with certainity.[25]

Management of symptomatic myelolipoma is surgical excision. Surgical treatment should be done in cases of large tumor because of risk of rupture, bleeding and associated malignancy.

In all our patients there was neither any symptom recurrence nor any tumor recurrence after a median follow up of 20 months (range from 1 to 120 months). Exact size after which surgery is advised in asymptomatic patients is still a matter of debate but some authors suggest size of more than 7 cm has been taken as cut off point.[15] Han and colleagues in their study reported follow up of 16 adrenal myelolipomas without surgical intervention for an average of 3.2 years [range 0.3 to 10.8 years]. Thirteen patients remained asymptomatic and 2 experienced persistent vague abdominal discomfort. A total of 13 tumors from 12 patients were serially imaged, with tumor size increasing in 6, decreasing in 2, and remaining unchanged in 5.[26]

One of our patients was kept on follow up. The lady had 4.5 cm tumor diagnosed in 2005 but increased in size to 8.5 cm in 2011.

Most patients underwent open transperitoneal adrenalectomy in our series. Laparoscopic transperitoneal adrenalectomy was done in 5 cases and laparoscopic adrenal sparing myelolipoma excision was done in one case. Laparoscopy is safe and feasible in tumor size upto 10 cm.[27] Largest size of tumor done laparoscopically was 14 cm in Castillo et al series, comparable to that operated by us (13.7 cm).[28]

Myelolipomas are usually encapsulated with defined yellow adipose tissue and red brown hematopoietic elements on cut section (Figure 4). Microscopic examination reveals mixture of mature adipocytes and hematopoietic elements (Figure 5). There is no recurrence on median follow up of 20 months (range 1 month to 120 months).

CONCLUSION

The article highlights the varying clinical presentations including rare emergency presentations of giant adrenal myelolipomas. A brief literature review is also presented.

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