

Case Report

Adrenal Myelolipoma: A report of three cases and review of literature

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Abstract

Adrenal myelolipoma is a rare and benign tumour composed of mature adipose tissue and haematopoietic elements that resemble bone marrow. It is mostly discovered incidentally on imaging of abdomen done for non adrenal related reasons or at autopsy. Usually asymptomatic, but has been reported to present with symptoms such as flank pain resulting from tumour bulk, necrosis or spontaneous retroperitoneal haemorrhage. Symptomatic tumours, growing tumours or tumours larger than 10 cm should be excised surgically. We report cases of 3 male patients presenting with flank pain and upper pole renal masses. All three were treated surgically with adrenalectomy. Myelolipoma was confirmed in all three on histology.

Introduction

Most myelolipomas are asymptomatic and hormonally non functional. They are discovered incidentally as a result of widespread noninvasive adrenal imaging. There are reported cases of endocrine dysfunction with myelolipomas, including conditions such as Cushing syndrome, Conn syndrome and congenital adrenal hyperplasia. In 85% of these patients, the abnormality involves the pituitary axis and in these patients stimulation with cortisol or adrenocortical hormone has been suggested to be involved in the pathogenesis of myelolipomas.¹ There is only one published case report of a surgically removed adrenal myelolipoma from Pakistan.² To the best of our knowledge this is the second report of symptomatic adrenal myelolipomas treated surgically from this country.

Case Reports

Case 1:

A 28 year old man presented to SIUT with complaints of mild right lumbar pain of 10 months duration. There was a past history of right pyelolithotomy 8 years back. Physical examination was unremarkable except for the presence of scar mark of the previous pyelolithotomy procedure. Blood pressure was within normal range. Abdominal ultrasonography showed a well circumscribed echogenic mass, arising from the upper pole of right kidney, measuring

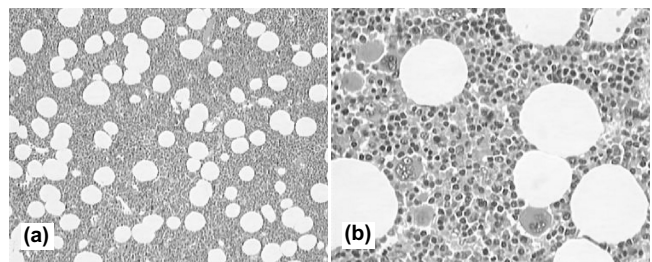


Fig-1: Histological features of Myelolipoma.(a) Low Power micrograph showing an intimate admixture of adipose tissue and bone marrow elements (H&E stain x 50). (b) High power micrograph showing fat cells and all three lineages of hematopoietic marrow (H&E stain x 400).

11.3x9.2 cms and CT scan showed a well defined 8.5x8.4 cms size mass at the upper pole of right kidney, predominantly fatty. Few specks of calcification were noted with the margin. The lesion did not take up contrast. Differential diagnoses of adrenal adenoma, primary adrenal malignancy, adrenal myelolipoma, and renal angiomyolipoma (least likely) was rendered. A chest X-ray showed no metastasis. Serum cortisol, urinary 24 hours VMA, CBC, Urine DR and Renal function were normal. Due to pain and size of the lesion, surgical removal was contemplated.

Peroperative findings revealed a large rounded mass at upper pole of right kidney (most probably adrenal in origin), completely encapsulated with a smooth outer surface.

Gross examination showed a large, rounded encapsulated tumour mass with smooth external surface, measuring 10x8x6 cms, cut section a solid tumour with a variegated appearance with yellowish areas alternating with dark red and haemorrhagic regions. Microscopy revealed characteristic admixture of mature adipose tissue with normal haematopoietic tissue. A thin rim of adrenal cortical cells was identified at the periphery of the lesion. A diagnosis of adrenal myelolipoma was made.

Case 2:

A 65-year-old male presented with severe right sided flank pain of acute onset. Blood pressure 125/80, pulse 88/min. Abdomen was benign. Ultrasound showed a mass at upper pole of right kidney. Serum cortisol and VMA levels were normal. CT scan showed a non enhancing



Fig-2: (a) Computed tomography showed a well-encapsulated large mass of fat and fluid density measuring 5x7.5 cm. (b) CT scan showing 10x13x9 cm mass, abutting against inferior surface of liver displacing IVC, head of pancreas and right kidney inferiorly

heterogenous, low density mass without calcification, 10x13x9cm in size in the right adrenal gland abutting against the inferior surface of right lobe of liver and displacing the Inferior Vena Cava.

Right adrenalectomy was done. Surgical pathology revealed a 9x12x9cm mass. Microscopy showed mature adipose tissue along with haematopoietic elements containing myeloid, erythroid, megakaryocytic and lymphoplasma cells in different stages of maturation with foci of necrosis and haemorrhage. A rim of normal adrenocortical tissue was present.

Findings were consistent with myelolipoma.

Case 3:

A 60-year-old man with known hypertension presented with two year history of mild discomfort in the right flank. Pulse was 68/min, BP 160/110, rest of physical examination was benign. Routine blood tests and cortisol and VMA levels were normal. Ultrasound showed a right suprarenal mass. CT scan findings showed a 5x7.5cm right adrenal mass with foci of macroscopic fat and with no uptake of contrast or calcification.

Macroscopic and microscopic findings on the adrenalectomy specimen were consistent with Myelolipoma.

Discussion

Adrenal 'Incidentalomas' are clinically inapparent adrenal masses discovered during diagnostic testing or treatment for clinical conditions, not related to the adrenal gland. Adrenal myelolipoma is one of the adrenal incidentalomas. It is a female dominant disease although in our series all the patients were male. Most myelolipomas are asymptomatic; however symptoms that lead to imaging commonly are abdominal pain or haematuria. Pain is associated with mechanical compression from a large tumour, retroperitoneal haemorrhage or tumour necrosis.³

The most dramatic presentation of myelolipoma is that of an acute haemorrhage; however it is a rare entity. CT findings of the lesions with haemorrhage are almost similar to those of lesions that did not bleed.⁴ The most important is the difference in size with most bleeding lesions being greater than 10cm. Sizes have been variable ranging from microscopic lesions to giant masses as big as 31 cm.⁵

Adrenal myelolipomas with diameter of >5cm can be easily detected on ultrasound. CT scan is the most sensitive test for diagnosing myelolipoma.⁵ The lesion is typically seen as a well encapsulated heterogeneous suprarenal mass of low density with negative attenuation values, interspersed by dense myeloid tissue and with or without specks of calcification.⁶ Differentials include adrenal adenoma 41%, adrenal metastasis 19%, primary adrenal malignancy 10%, myelolipoma 9% and very rare are retroperitoneal lipoma and liposarcoma.³ MRI is also useful for diagnosis.⁷

Adrenal myelolipomas can co-exist with other adrenal tumours like adrenocortical adenomas⁸ and adenomatoid tumours of the adrenal gland.⁹

Endocrine evaluation is not helpful in diagnosing a myelolipomatous mass as it is defined as being mostly hormonally inactive. However if there are doubts regarding the radiographic diagnosis and there are features suggesting

a functional adrenal mass then the appropriate endocrine evaluation should be done along with a fine needle aspiration biopsy. The tumour size rule does not apply on myelolipoma as they are benign non-functioning tumours and most are >6cms in size.^{3,4}

Management of myelolipoma should be done on a case to case basis. Patients with lesions <10 cm defined as myelolipoma on imaging procedures, 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, however the follow-up will be life long because interval growth has been reported previously.^{2,3} If patient starts exhibiting symptoms or 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 haemorrhage. There are case reports of contralateral myelolipoma after resection of the primary lesion. Follow-up after adrenalectomy for unilateral myelolipoma is recommended.³

Regular follow up is thus essential for both groups of patients.

References

1. Hisamatsu H, Sakai H, Tsuda S, Shigematsu K. Combined adrenal adenoma and myelolipoma in a patient with Cushings syndrome: case report and review of literature. *Int J Urol* 2004; 11:416-8.
2. Bhurgri A, Bhurgri Y, Khwaja IA. Adrenal Myelolipoma - A case report. *J Pak Med Assoc* 2001; 51:85-6.
3. Han M, Burnett AL, Fishman EK, Marshall FF. The natural history and treatment of adrenal myelolipoma. *J Urol* 1997; 157:1213-6.
4. Kenney PJ, Wagner BJ, Rao P, Heffess CS. Myelolipoma: CT and pathologic features. *Radiology* 1998; 208:87-95.
5. Akamatsu H, Koseki M, Nakaba H, Sunada S. Giant adrenal myelolipoma; report of a case. *Surg Today* 2004; 34:283-5.
6. Haque F, Harish S, Ahmad I, Qamar A, Pandey H. Adrenal myelolipoma A case report. *Indian J Radiol Imag* 2004; 14: 301-2.
7. Guo YK, Yang ZG, Li Y, Deng YP, Ma ES, Min PQ, et al. Uncommon adrenal masses: CT and MRI features with histopathologic correlation. *Eur J Radiol* 2007; 62: 359-70.
8. Al-Brahim N, Asa A. Myelolipoma with adrenocortical adenoma: An unusual combination that can resemble carcinoma. *Endocr Pathol* 2007; 18:103-5.
9. Elizabeth R, Timonera, Paiva ME, Manuel LJ, Catarina E. Composite Adenomatoid Tumour and Myelolipoma of adrenal gland: Report of 2 cases. *Arch Path Lab* 2008; 132: 265-7.