

Adrenal Myelolipoma- A Rare Case Report

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Abstract: Adrenal myelolipoma (AML) is a rare benign tumour composed of mature adipose tissue and hematopoietic tissue. Very few cases have been reported. Most of these patients are asymptomatic. We present a rare case of Adrenal Myelolipoma where the patient presented with hypertension and a clinical suspicion of Pheochromocytoma, which turned out to be an Adrenal myelolipoma. Adrenal myelolipoma is a rare entity, not encountered frequently and can occur as an incidental finding. Awareness regarding this entity is very much essential to exclude surgical exploration or extensive surgery.

Key Words: Myelolipoma, adrenal, incidentaloma

Introduction

Adrenal myelolipoma (AML) is a rare benign tumour composed of mature adipose tissue and hematopoietic tissue. They are found incidentally at autopsy or through CT scan done for other reasons[1]. Frequency of myelolipomas in adrenal incidentalomas varies between 7-15% [2]. Extra adrenal sites for myelolipomas include retroperitoneum, thorax and pelvis[3]. Until 2001, only 158 surgically documented cases of adrenal myelolipoma have been reported in English literature, following which very few cases are reported. They are hormonally inactive though they may sometimes coexist with primary aldosteronism, congenital adrenal hyperplasia (CAH), pheochromocytoma, adenoma and Cushing's syndrome [1,4]. Most of these patients are asymptomatic, occasionally may present with abdominal pain due to either being large or from spontaneous hemorrhage, more likely when predominantly composed of myeloid tissue. Awareness regarding this rare entity is essential to avoid extensive surgery.

Case History

A 33 years female presented with pain in the right hypochondrium and lumbar region since 2 months. On clinical examination she was found to be a hypertensive (BP: 150/100mmHg.) There was no other significant finding clinically.

Investigations: Routine hematological parameters like hemoglobin, complete blood count, peripheral smear were within normal limits. Blood glucose, urea, creatinine, sodium, potassium and bicarbonate levels were normal. Ultrasonography revealed a cavernous hemangioma in the right lobe of liver and a hyper echoic area in the suprarenal region. Computerized tomography scan showed a well outlined mass measuring 3.5x3cms, superior to the upper pole of right kidney in the suprarenal region. Right and left kidneys were normal. Urinary vanillyl mandelic acid (24hours) was 13 mg/day plasma adrenaline-128pg/ml, plasma noradrenaline-100pg/ml, 24hours urinary 17-ketosteroids -9.4mg/day. Ophthalmoscopy revealed Grade II hypertensive retinopathy. HBsAg was negative and HIV 1 & 2 was non reactive.

Pheochromocytoma was suspected clinically. Right adrenalectomy was performed under general anesthesia. Post operative period was uneventful.



Fig.1 Cut surface of the mass

Pathology: A globular well encapsulated soft tissue mass measuring 3.5x3.2x2cms was received. External surface was smooth, greyish brown in colour. Cut surface was dark brown with yellowish areas. (Fig. 1)

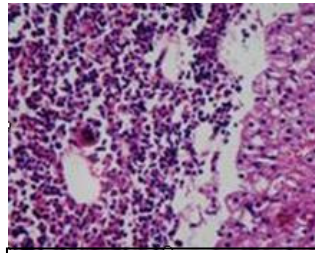


Fig.2:Microphotograph of adrenal myelolipoma showing strip of adrenal tissue with foci of fatty and hematopoietic tissue. (H & E; X100)

Microscopy revealed adrenal tissue at the periphery beneath which was seen a mass composed of large areas of hematopoietic tissue containing erythroid, lymphoid, granulocytic, megakaryocytic elements and scattered areas of mature adipose tissue. (Fig. 2 & 3) There was no evidence of any other associated elements like pheochromocytoma, adrenocortical hyperplasia or adenoma on microscopic examination. A histological diagnosis of myelolipoma was offered.

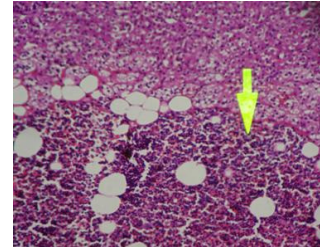


Fig.3 Microphotograph showing myeloid, adipose tissue and megakaryocytes. (H & E; X200).

Discussion

Adrenal myelolipoma has been reported in 5th to 7th decade of life, without any sex predilection. Right adrenal is more commonly involved than left. They are generally unilateral, smaller and measure less than 5mm in diameter[4]. Occasional giant Adrenal myelolipoma has been reported. The largest reported tumor is 6kg in weight [1]. Most of the patients are asymptomatic. Occasionally they may present with abdominal pain due to either being large or traumatic rupture leading to hemoperitoneum or from spontaneous hemorrhage. Frequent association with obesity and type 2 diabetes mellitus and hypertension is noted, possibly coincidental [5]. Adrenal myelolipoma has been reported to coexist in association with CAH due to 21 α hydroxylase or 17 α hydroxylase deficiency. It is believed that excess ACTH secretion over a long period may stimulate myelolipomatous alterations in adrenal gland [4]. The myelolipomatous tissue can replace either the tumorous or hyperplastic adrenocortical cells or may simply represent secondary degenerative changes. Differential proliferation of the undifferentiated mesenchymal stem cells of adrenal cortex into myeloid and adipose tissue in response to infection, stress and necrosis has been the projected view for aetiopathogenesis [6]. Demonstration of hypodensity within an adrenal mass is virtually diagnostic of myelolipoma by CT scan. Desai et al [7] felt that biochemically a nonfunctioning radiolucent solid adrenal mass by CT scan with no neovascularity on angiography is most likely a myelolipoma. MRI is

required to demonstrate the origin of the tumor, to define tissue planes when tumor is large and heterogenous and to distinguish benign from malignant lesions by comparing signal intensity ratios of adrenal to liver [4].

Management: It is felt that masses less than 4cms diameter and asymptomatic, diagnosed on imaging and or by cytological studies should be subjected to watchful monitoring. However, when malignancy is suspected or symptomatic tumors more than 4cms should be extirpated because of risk of spontaneous rupture with retroperitoneal bleeding [8].

Conclusion:

Adrenal Myelolipoma is a rare entity, not encountered frequently. Awareness regarding this entity is very much essential to exclude surgical exploration or extensive surgery.

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