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Adrenal myelolipoma in a patient presented with hypertension- A Case Report

Ali A.M. Attarbashii (1)

(1) Department of Urology, Al-Jomhory Teaching Hospital, Al-Shifa'a quarter, Mosul, Iraq

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ABSTRACT

Adrenal myelolipoma 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. I 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.

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*Corresponding author E mail : attarbashii73@yahoo.com

Introduction:

Adrenal myelolipoma 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]. Extra-adrenal sites for myelolipomas include retroperitoneum, thorax and pelvis[2]. Frequency of myelolipomas in adrenal incidentalomas varies between 7-15%[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 report: a 45 years old female presented with intermittent right loin pain with paroxysmal headache and exertional dyspnea for the last three months. On clinical examination she was found to be a hypertensive (BP:160/100mmHg.).She was obese (Body weight was 95 kg). There was no other significant finding. Pheochromocytoma was suspected clinically.

Ultrasonography revealed a highly echogenic well defined mass 6×4.5cm in the right suprarenal region. Computerized tomography scan showed a well outlined mass that has some fatty content measuring (6×5×3.5cm) superior to the upper pole of right kidney in the suprarenal region. Both kidneys were normal with only downward displacement of the right kidney (Fig.1).

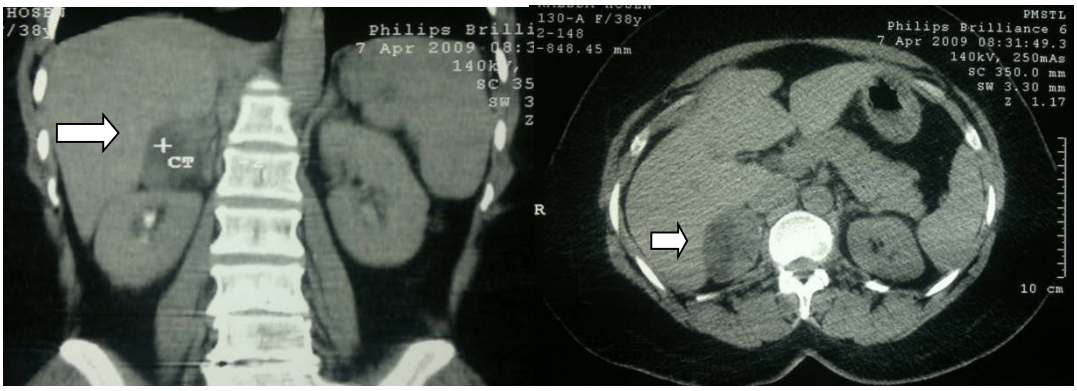
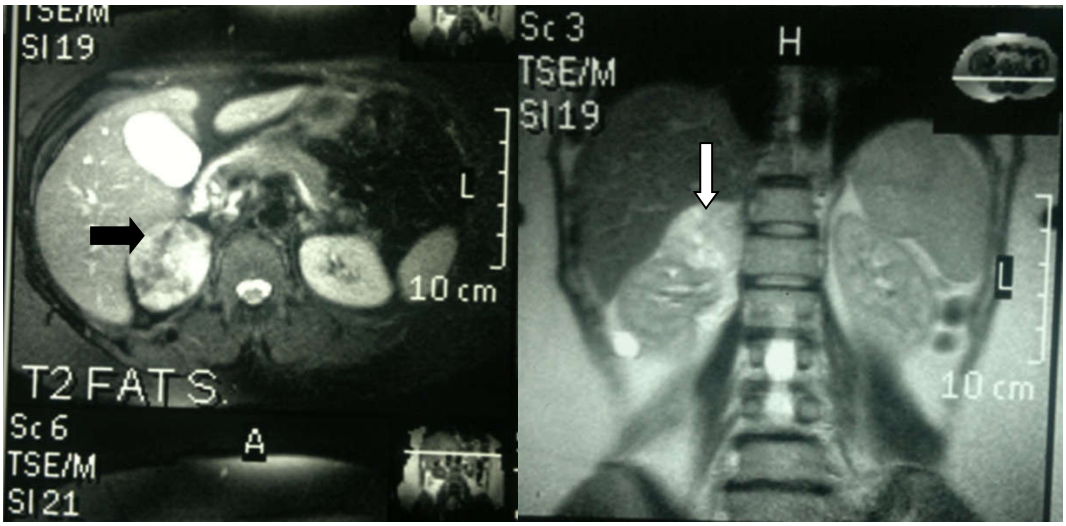


Figure (1): CT-scan films showing right adrenal mass.

MRI revealed oval shape solid mass (6×5×3.5cm) located in the right suprarenal area, low intensity on T1, high T2 signal and showed heterogenous enhancement post-contrast injection (Fig.2).



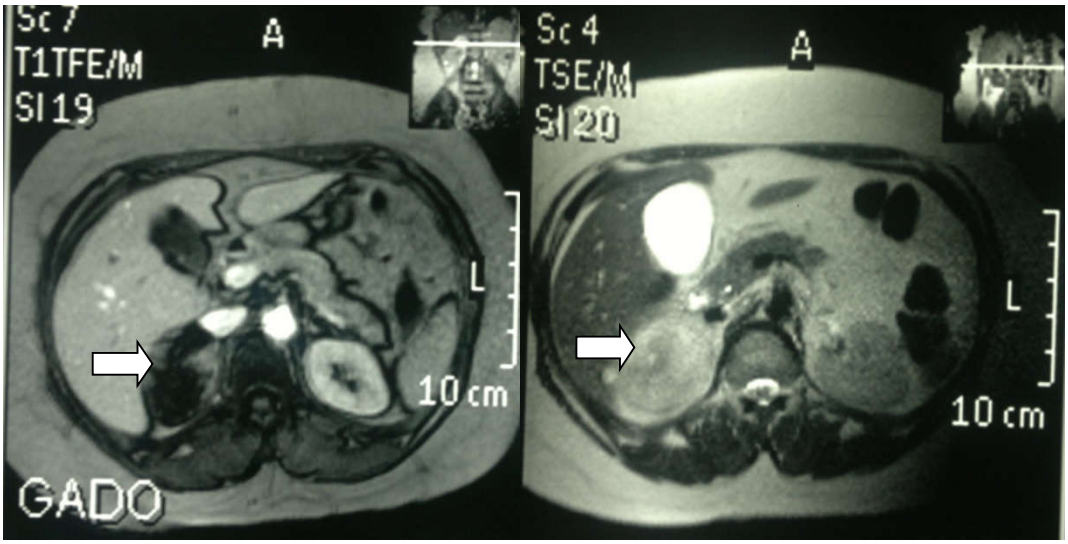
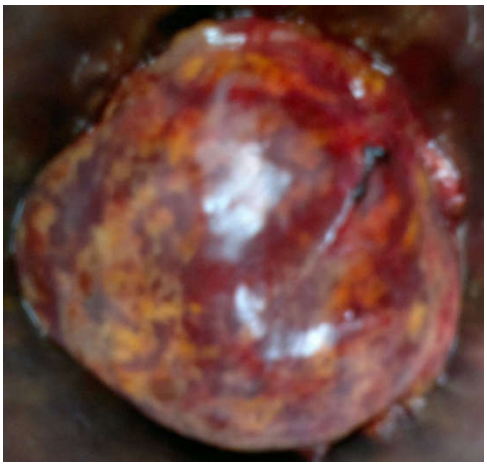


Figure (2): MRI films showing right adrenal mass.

Laboratory parameters like haemoglobin, complete blood count, ESR, peripheral smear were within normal limits. Blood glucose, urea, creatinine, sodium, potassium and cloride levels were normal. 24-hours urinary vanillyl mandelic acid was 27.6 mmol\l (N.R.<68.6 mmol\l). HBsAg was negative and HIV 1 & 2 was non reactive. ECG, chest x-ray and echocardiography were all normal. After a preoperative control of hypertention by using alpha-blockers (Prazosin 50mg\day) with beta-blockers (Atenolol 50mg\day), right adrenalectomy through a flank retroperitoneal approach was performed under general anesthesia. Post operative period was uneventful with normalization of the patient's blood pressure without antihypertensive medication. Pathologically A globular well encapsulated soft tissue mass measuring 6×5.5×3.5 cm weighing 60 grams **(**normal weight in females 8.3 grams).



Post-adrenalectomy gross appearance of the right adrenal tumour (Myelolipoma).

Sections manifested intradermal mature adipose tissue heavily infiltrated by trilineage haematopoietic cells (granulocytes, erythroid cells and megakaryocytes). The residual adrenal cortex tissue is unremarkable. There was no evidence of any other associated elements like pheochromocytoma, adrenocortical hyperplasia or adenoma on microscopic examination. A histological diagnosis of adrenal myelolipoma was offered.

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, usually 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 congenital adrenal hyperplasia (CAH) due to 21-hydroxylase or 17-hydoxylase 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 suggestive 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]. 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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