CASE REPORT

Adrenal Myelolipoma: A Rare Case Report

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Abstract:

Adrenal myelolipoma is a rare benign tumour of the adrenal gland composed of mature adipose tissue and haematopoietic elements that resemble bone marrow. Largely discovered incidentally on imaging of abdomen was done for non adrenal related reasons or during autopsy. Usually asymptomatic, it has been reported to present with symptoms such as flank pain resulting from tumour bulk, necrosis or spontaneous retroperitoneal haemorrhage. Adrenal myelolipoma is usually unilateral and asymptomatic, though known to be associated with obesity, hypertension, endocrinological disorders and some malignancies. We are reporting a case of a patient who presented with abdominal mass with radiological findings revealing a right suprarenal mass measuring 8.5×5×4 cm. Histopathological findings confirmed an adrenal myelolipoma.

Keywords: Adrenal, Autopsy, Haemorrhage, Myelolipoma

Introduction

Adrenal myelolipoma is a rare benign urological lesion composed of mature adipose tissue and hematopoietic elements resembling a bone marrow. Initially was described by Gierke in 1905 and Oberling in 1929 termed the entity as *formations myelolipomatoses* [1]. It is usually detected at autopsy or in conditions of massive growth or an altered hormonal production which leads to its clinical presentation. However, due to radiological enhancements such as Ultrasonography (US), Computed Tomography (CT) and Magnetic Resonance Imaging (MRI), incidental detection has become more common [2]. The tumor affects males and females equally and seen most commonly in the fifth to seventh decade of life [2]. Its Incidence is 3–5% of all primary tumors of the adrenals, with increased incidence seen in the later age group [3]. These tumours are unilateral, small, and asymptomatic although some bilateral myelolipomas have been reported [2]. These are often smaller than 4 cm in diameter; the largest reported being 31×24.5×11.5 cm, which weighed 6 kg. Recurrence of these tumours is rare in nature [3].

Case Report:

A 37 year old male reported to the Surgery Out Patient Department with right sided abdominal mass and complaints of right flank pain, vomiting and malaise since one month. Radiological investigation revealed a well defined right suprarenal mass measuring 70×80 mm, cystic in consistency well separated from right suprarenal gland. A surgical resection of the mass was performed and the mass was received in histopathology. The gross features revealed a globular encapsulated grey tan mass measuring 8.5×5×4 cm. Cut section revealed the variegated dark brown haemorrhagic and yellowish fatty areas (Fig.1a and Fig.1b). Microscopy revealed compressed adrenal cortical tissue towards one side with 50% of the tissue comprised of the

underlying mature adipose tissue admixed with immature myeloid elements comprised of megakaryocytes, thus confirming the diagnosis of adrenal myelolipoma. CD 56 positivity confirmed compressed adrenal tissue and CK19 was negative (Fig.2a,b,c,d). One year follow up of the patient was unremarkable.

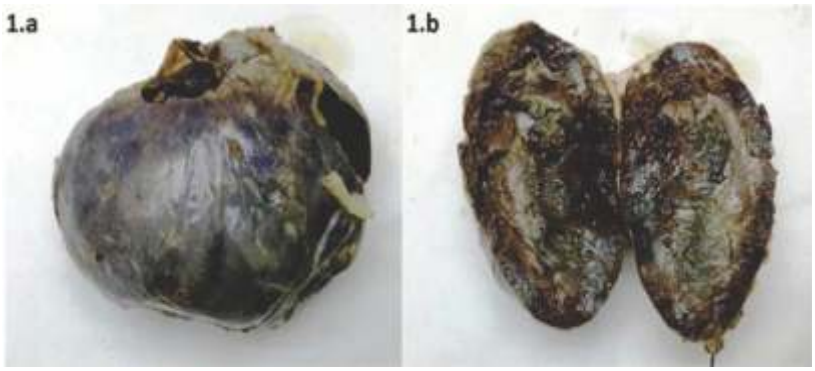

Fig. 1a: Gross: Globular Encapsulated Grey Tan Mass
Fig. 1b: Cut Section: Variegated Dark Brown Haemorrhagic and Yellowish Fatty Areas

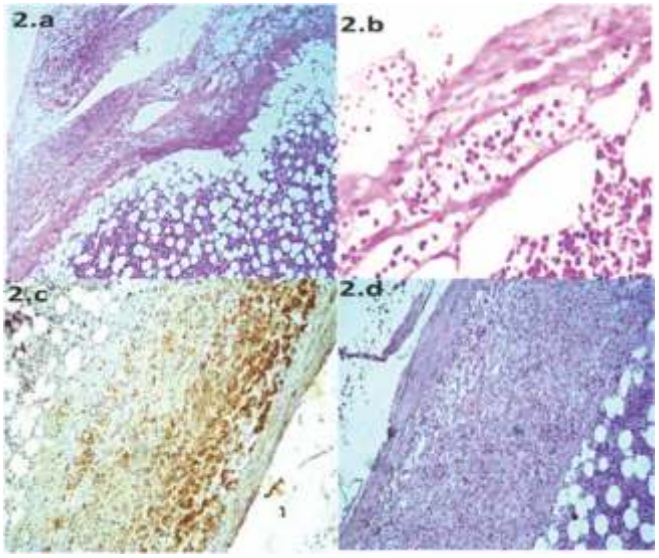

Fig. 2a: Mature Adipose Tissue Admixed with Immature Myeloid Elements (40X)
Fig. 2b: Normal Adrenal Cortical Tissue (100X)
Fig. 2c: CD 56 Positivity Noted in Compressed Adrenal Tissue (40X)
Fig. 2d: CK-19 was Negative (40X)

Discussion:

Adrenal myelolipoma constitutes a rare entity in urological practice composed of variable proportions of mature adipose tissue and active hematopoietic elements. The incidence ranges from 0.08% to 0.4%, and less than 300 cases were reported in the literature before 2000 [4]. However, their prevalence is increasing up to 10%, due to enhanced imaging modalities [5].

Its etiology has many theories; however, the most acceptable theory is adrenocortical cell metaplasia of reticuloendothelial cells of blood capillaries in the adrenal gland responding to necrosis, inflammation, infection, or stress etc. [6]. This chronic stimulation to the adrenal gland, which leads to increased incidence of the lesion in the elderly age group [7], may lead to development of benign as well as malignant tumours. Conditions associated with adrenal myelolipomas include Cushing's disease, obesity, hypertension, Conn's syndrome, congenital adrenal hyperplasia and diabetes which are major stimulating factors as well [8]. Ultrasonography, computed tomography, and MRI are all effective in diagnosing more than 90% of adrenal myelolipoma on the basis of identification of fat, with CT scan being the most sensitive [2, 9]. These are nonfunctional tumours, hence endocrinological characteristics and evaluations are not benificial, although there is a report of a hypertension associated secreting myelolipoma [3]. Among the differential diagnosis renal angiomyolipoma, which is triphasic with myoid spindle cells, islands of mature adipose tissue and dysmorphic thick walled blood vessels without elastic lamina and is HMB-45 positive should be included along with retroperitoneal lipoma which is comprised of only mature adipocytes, and liposarcoma which comprises of mature and variable sized adipocytes [10].

Adrenal myelolipoma should be managed on individual basis. Smaller lesions, which measure less than 5 cm and are asymptomatic, should be checked over a period of 1-2 years with imaging controls [11]. However, symptomatic tumours or masses larger than 7 cm should be surgically excised [2]. Possible threatening complications are spontaneous rupture and hemorrhage of the mass which presents with life threatening cardiovascular shock [2]. The prognosis of Myelolipoma depends on its size and location, with the mechanical compression by larger tumours, tumour necrosis and retroperitoneal hemorrhage cause extremely painful sensations [12]. Hormone levels should be obtained however, because co-existing functional cortical adenoma and myelolipoma have been reported. There are few case reports documenting presentations with virilisation, hypertension, hyperaldosteronism and hypercortisolism indicating hormone secretion leading to as well as pheochromocytoma [13].

Conclusion:

Adrenal myelolipomas are rare tumours, mostly of benign nature, and clinically silent. However, these “incidental” cases warrant systematic diagnostic study to plan proper management. Imaging modalities such as ultrasonography and computed tomography helps to yield the diagnosis, as seen in our case. With many cases of myelolipomas reporting with endocrine abnormalities, thus necessitating and encouraging the use of thorough workup before the surgery which also includes biochemical findings.

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