

Giant adrenalmyelolipoma: A rare benign neoplasm

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Abstract

Adrenal myelolipoma is a relatively rare benign tumor composed of mature adipose tissues and a variable amount of hematopoietic elements. The incidence of adrenal myelolipoma is 0.08%-0.4%. The male to female ratio is 1:1. We are here reporting a surgically treated case of giant adrenal myelolipoma. A 52 years old man presented in our surgical outpatient clinic with retroperitoneal tumor. Right adrenalectomy was done. The origin of the tumor was suggested to be the right adrenal gland. The weight of the excised tumor was 700gm & size is 14×9.5×7 cm. The histopathological diagnosis was adrenal myelolipoma. The patient had uneventful recovery and was discharged from the hospital. He has been followed up in our outpatient clinic.

Keywords: adrenal myelolipoma, giant, benign neoplasm

Introduction

Adrenal myelolipoma is a relatively rare benign tumor composed of mature adipose tissues and a variable amount of hematopoietic elements. The male to female ratio is 1:1. The incidence of adrenal myelolipoma is reported to be 0.08%-0.4% including autopsy [1]. Although the diameter of Adrenal myelolipoma ranges from less than 1cm to more than 30cm, they are usually less than 5cm in diameter [13, 14]. Tumors of size more than 8 cm are referred to as giant myelolipoma [1, 4]. The largest adrenal myelolipoma (size-31×24.1×11.5cm, weight 6000gm) in a patient without endocrine disorder was described by Akamatsu *et al.*⁶, and the largest adrenal myelolipoma in a patient with congenital adrenal hyperplasia (size-34×24×10.5, weight 5900gm) was described by Boudreaux *et al.* [7].

Adrenal myelolipoma are nonfunctional tumors that are usually asymptomatic, however, they have been known to coexist with other endocrine disorders, such as Cushing's syndrome, congenital adrenal hyperplasia (CAH), Conn's syndrome and pheochromocytoma [2, 4]. Recently, adrenal myelolipoma have been reported in patient with congenital adrenal hyperplasia with increasing frequency. One study indicated that myelolipoma was detected in 4% of patients with congenital adrenal hyperplasia (CAH) [5].

We are reporting a relatively rare case of a giant adrenal myelolipoma measures 14×9.5×7 cm in size and 700gm in weight in a patient without endocrine disorder.

Case report

A 52 years old male patient presented with a right abdominal mass. He was referred to our surgical outpatient clinic to undergo a detailed examination and treatment for the right abdominal mass. A clinical examination revealed a soft, smooth-surfaced, painless moderately large tumor with poor mobility, which was located in the right upper

abdomen. Abdominal computed tomography (CT) & MRI of the abdomen was done and which revealed a retroperitoneal tumor. The retroperitoneal tumor was resected and sent to the pathology department for histological examination. The resected tumor was 14×9.5×7 cm in size and weight 700 gm. On examination of the cut surface of the tumor revealed a multilobular yellow, solid, soft mass with fibro fatty and hemorrhagic areas. Multiple sections studied from the tumor for histopathological examination with hematoxylin and eosin stain, which revealed that the tumor was composed of lobules of mature and variable sized adipocytes admixed with aggregates of hematopoietic elements, associated with compressed adrenal gland tissue in the periphery. There are also fibro collagenous tissues and focal areas of calcification.

These findings were compatible with adrenal myelolipoma. The patient had an uneventful recovery and was discharged from the hospital. He has been followed up in our outpatient clinic.



Fig 1: Gross morphology of resected specimen

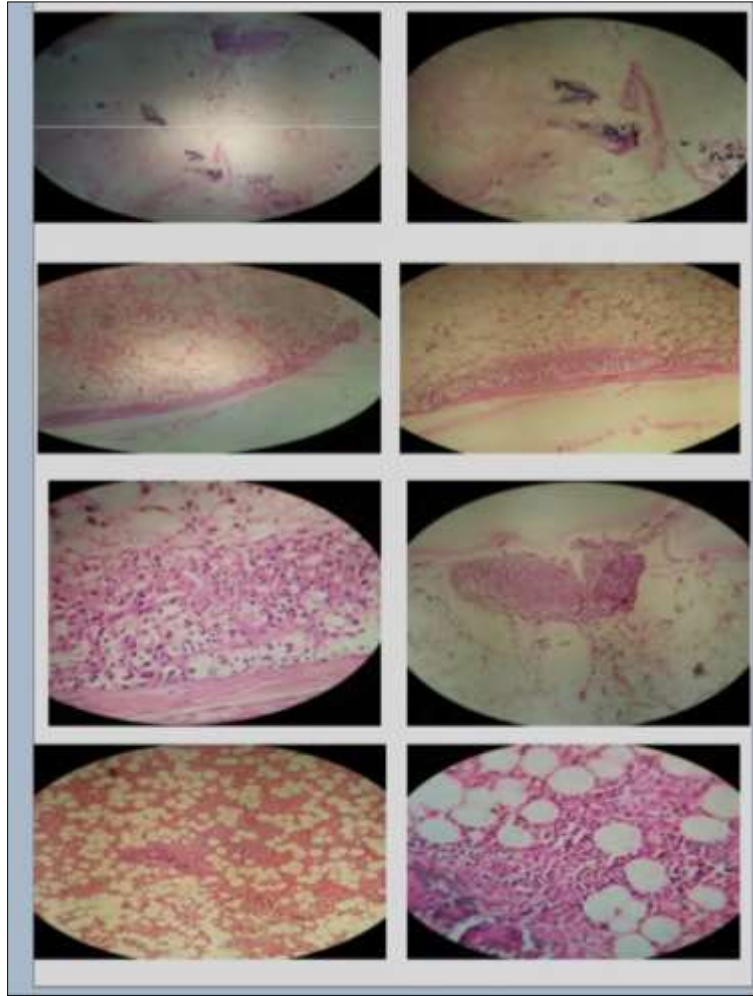


Fig 2: H & E stain (10x) showing microscopic features of myelolipoma

Discussion

The etiology of Adrenal myelolipoma remains unclear. Some of the hypothesized etiologies include extra medullary hematopoiesis due to autonomous proliferation of bone marrow cells transferred during embryogenesis, degeneration of epithelial tissue of the adrenal cortex and adreno-cortical cells, metaplasia of the reticulo endothelial cells of the blood capillaries in response to stimuli such as necrosis, infection or stress [1, 8, 10]. The most widely accepted theory is that myelolipoma arise due to metaplasia of the reticulo endothelial cells of the blood capillaries in the adrenal gland in response to stimuli such as chronic stress, infection, necrosis or inflammation [11, 12]. Although the diameter of adrenal myelolipoma ranges from less than 1cm to more than 30cm, they are usually less than 5cm in diameter [13, 14]. Tumors of size more than 8 cm are referred to as giant myelolipoma [1, 4]. Adrenal myelolipoma is often asymptomatic sometimes leading to very large adrenal masses (≥ 10 cm in diameter). These are often called “giant” adrenal myelolipoma [15]. Lawler *et al.* proposed a definition of the often-quoted term “giant” AML [16]. The largest adrenal myelolipoma (31 \times 24.1 \times 11.5cm, weight 6000gm) in a patient without endocrine disorder was described by Akamatsu *et al.* [6] and the largest adrenal myelolipoma in a patient with congenital adrenal hyperplasia (CAH) (size-34 \times 24 \times 10.5, in size and weight 5900 gm) was described by Boudreaux *et al.* [7].

We are reporting a relatively rare case of a giant adrenal myelolipoma of 700gm in weight and 14 \times 9.5 \times 7 cm in size

in a patient without endocrine disorder. Ultrasonography, CT and MRI are effective for diagnosis of adrenal myelolipoma in $\geq 90\%$ of cases [4, 7]. Recently with the widespread use of imaging studies such as ultrasonography, CT and MRI, the incidental detection of adrenal myelolipoma has been more common, and they now represent up to 10-15% of incidentally detected adrenal masses [17]. Ultrasonography shows myelolipoma as a well-defined tumor with varying degrees of hyper echoic (fatty tissue) and hypo echoic (myeloid tissue) components. CT shows myelolipoma as a well-delineated fat tissue with more dense areas of myeloid tissue.

Management of adrenal myelolipoma should be individualized. Small lesions, which are asymptomatic and measure less than 5cm, should be monitored over a period of 1-2 years with imaging controls [18]. On the other hand, surgery is indicated when the patient is symptomatic, when the lesion is more than 5cm in size due to rupture – which is a rare event or when malignancy is suspected [18]. The most recognized complication of adrenal myelolipoma is spontaneous retroperitoneal hemorrhage [14, 16]. Daneshmand *et al.* suggested that symptomatic tumors or myelolipoma of ≥ 7 cm in size should removed because they are associated with an increased risk of spontaneous rupture with retroperitoneal hemorrhage [4].

Conclusion

We reported a relatively rare case of a giant adrenal myelolipoma in a patient without an endocrine disorder. It is

very important to provide suitable management on an individual basis.

Conflict of Interests: There is no conflict of interests

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