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Giant adrenal myelolipoma: A rare entity

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Abstract: Adrenal myelolipoma is a rare, benign and hormonally inert tumor that is consisted of hematopoietic tissue and mature adipose tissue. It is usually diagnosed incidentally and nowadays much more frequently because of predominant use of ultrasonography, computed tomography and magnetic resonance imaging. Adrenal myelolipoma is usually unilateral and asymptomatic. They are known to be associated with endocrinological disease, obesity, hypertension, and some cancers. We report hereby a case of right-sided giant adrenal myelolipoma presenting with abdominal pain for about 20 days. A Computed Tomography scan revealed a large mass on the right side of the abdomen reaching the midline and was suggestive of a retroperitoneal Liposarcoma. Mass was resected and histological examination revealed it as adrenal myelolipoma.
Keywords: Adrenal gland, Adrenal tumor, myelolipoma, Adrenalectomy

INTRODUCTION:

Adrenal myelolipoma is a rare, benign tumour consisting of irregular mixture of hematopoietic elements and mature fat that resembles bone marrow. They are usually small (<5cm), unilateral and asymptomatic, though bilateral lesions have also been reported. Larger myelolipomas, known as giant myelolipoma, are rare. Both men and women are equally affected, mostly in their fifth to seventh decades. Although benign, this tumor can cause a dilemma for the treating surgeon. They can be mistaken radiologically for a retroperitoneal liposarcoma, so histopathological examination is essential for its diagnosis. Here we describe a case of unilateral giant adrenal myelolipoma in an elderly lady.

CASE REPORT:

A 70-year-old lady was presented to our hospital, complaining of abdominal pain for about 20 days, the pain was tolerable. On Examination a firm, fixed mass was palpable in the right flank. Her routine blood count and liver and renal function tests were normal. A Computed Tomography scan revealed a 20 x 15 x 10 cms hypoechoic mass on the right side of the abdomen reaching the midline and was suggestive of a retroperitoneal Liposarcoma. The hormonal analysis of the patient was unremarkable.

Surgical excision was done by open laparotomy, and a large tumor mass was found in the right adrenals almost replacing the adrenal glands. Specimen was received for histopathological examination.Gross examination revealed a round encapsulated mass of 20 x 12 x 8 cms. External surface showed capsule with adherent fat and areas of congestion. On cut section, the tumour was greasy and yellowish in colour, few irregular reddish brown areas noted at the periphery with areas of haemorrhage.

Microscopic examination revealed predominantly mature adipose tissue interspersed with active bone marrow tissue (Figure.1). This hematopoietic tissue contained erythroid, granulocytic cell lines, few lymphoid cells and many megakaryocytes (Figure.2). A thin rim of normal adrenal tissue was seen in the periphery. The patient had a normal postoperative course and was discharged after 5 days of surgery.

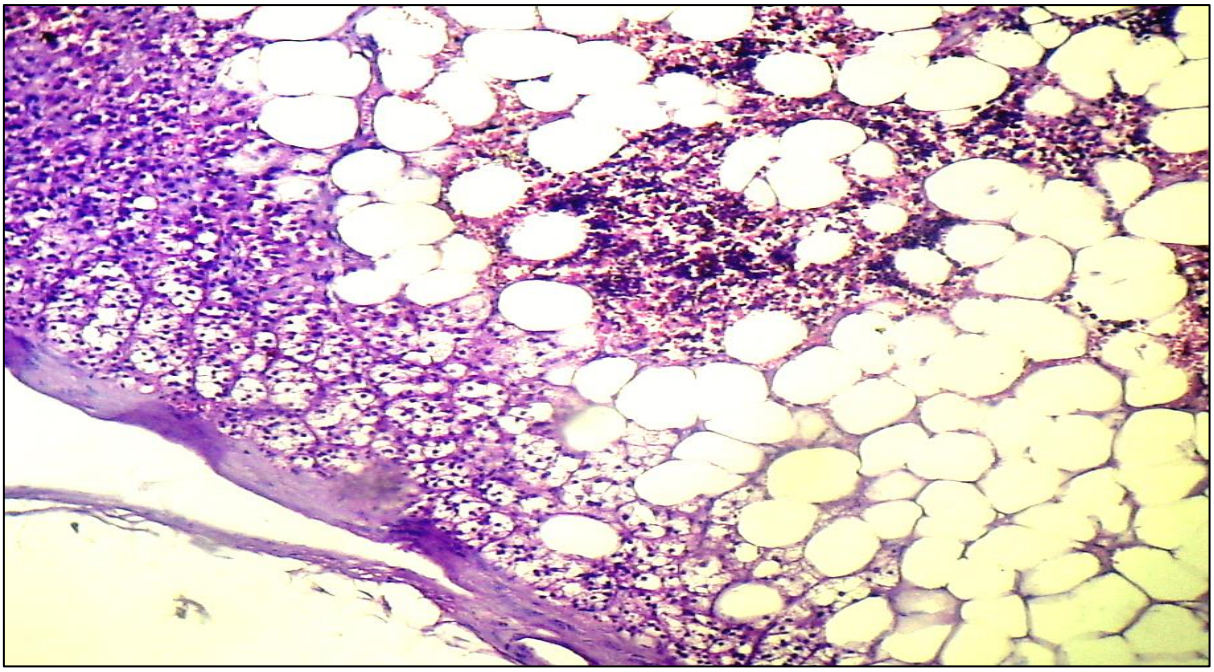

Fig 1: Histological picture showed adrenal myelolipoma composed of a mixture of mature adipocytes and hematopoietic elements with a rim of normal adrenal tissue (hematoxylin-eosin, × 400)

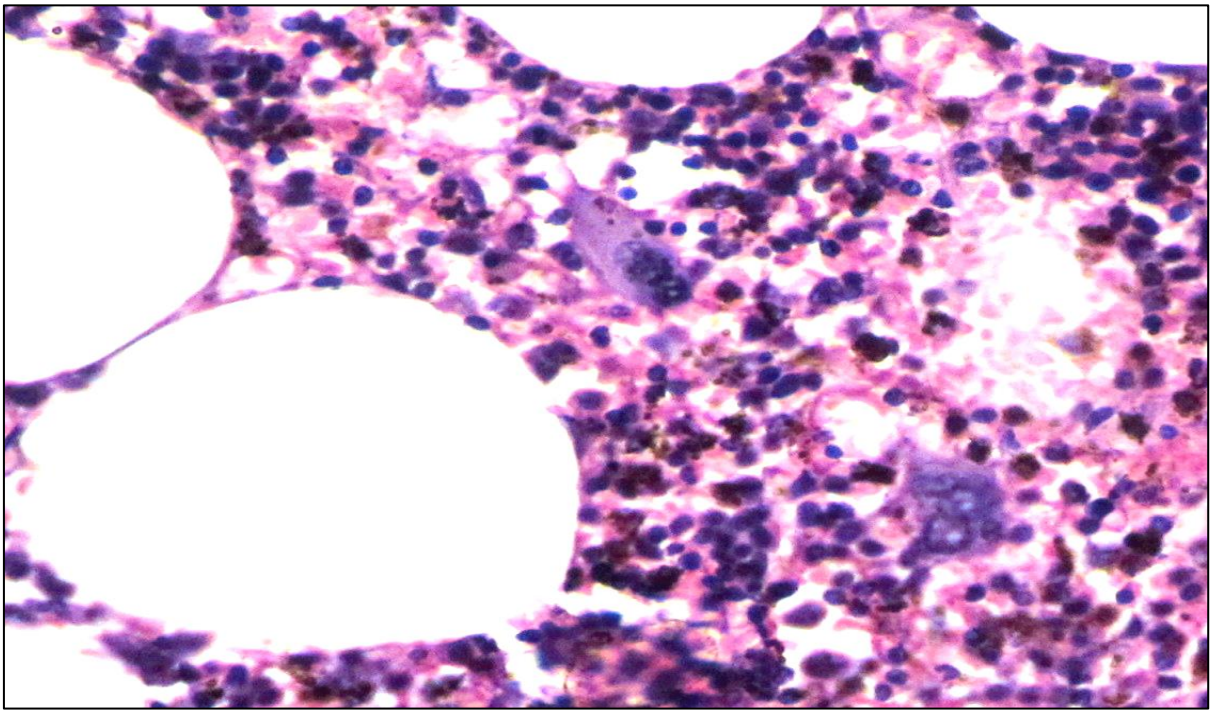

Fig 2: Hematopoietic tissue in adrenal myelolipoma contained erythroid, granulocytic cell lines, few lymphoid cells and many megakaryocytes (hematoxylin-eosin, × 1000)

DISCUSSION:

Adrenal myelolipoma is a benign tumor of adrenal gland composed of variable amounts of mature adipose tissue and all hematopoietic lineages of the bone marrow, i.e., myeloid, erythroid, lymphoid, and megakaryocytic lineage. Despite that it does not contribute as a hematopoietic source [1]. This tumour was first described by Gierke in 1905 [2] and the term "Myelolipoma" was given by Oberling in 1929 [3]. Myelolipoma is usually unilateral and most commonly found in adrenal gland but may rarely occur in extra-adrenal locations like the retroperitoneum, thorax and pelvis [4]. This neoplasm does not cause an endocrine disorder and no malignant potential has been demonstrated. Adrenal myelolipomas usually occur in late adult life and both males and females are affected equally [5]. The incidence is estimated at 0.08% to 0.25%. Myelolipomas account for approximately 3 to

5% of all primary adrenal tumors. These lesions rarely measure more than 5 cms. These tumors used to be discovered primarily during post-mortem; however, nowadays widespread use of newer radiological modalities has made diagnosis more frequent [6].

Multiple theories regarding the aetiology of adrenal myelolipoma have been studied. The most widely accepted amongst all of them is that it occurs due to metaplastic transformation in undifferentiated stromal cells or in the reticuloendothelial cells of blood capillaries in response to stimuli including necrosis, infection or stress [7, 8]. Another cause cited is prolonged stimulation with high levels of adrenocorticotropic hormone or adrenal androgen [9]. Bennett BD *et al.;* reported occurrence of myelolipoma with Cushing syndrome, congenital adrenal hyperplasia, and adrenal Ganglioneuroma [10].

Although, most of the lesions discovered incidentally are small and asymptomatic, reports are uncommon for cases of large symptomatic lesions. The term giant adrenal myelolipoma is given to tumors larger than 8cm [11]. Giant myelolipomas may be asymptomatic or present with feeling of an abdominal mass, dragging abdominal pain and may cause well-recognized complications like spontaneous retroperitoneal haemorrhage or abscess with compression of neighbouring structures [12]. Therefore, a symptomatic tumour should be treated surgically.

The differential diagnosis of the lesion on radiology comprises of renal angiomyolipoma, retroperitoneal lipoma, and liposarcoma. That's why histopathological study is mandatory, the microscopic features like mature adipose tissue with major blood forming elements like myeloid, erythroid and megakaryocytic series are confirmatory of adrenal myelolipoma.

CONCLUSION:

Our case report highlights the rare occurrence of giant, unilateral and nonfunctional adrenal myelolipoma. The lack of symptoms and non functional nature keeps them undiagnosed till they increase in size significantly. As far as our knowledge only few cases of Giant Adrenal Myelolipoma have been reported in literature. It is treated by simple Adrenalectomy and is completely curable. Awareness of this rare Adrenal entity and its correct diagnosis is important. However, even with the contemporary radiological techniques, accurate diagnosis is difficult. Thus, histopathological study is essential for confirmatory diagnosis of this rare adrenal neoplasm.

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