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A Rare And Valuble Case Report On Adrenal Myelolipoma And Extramedullary Hematopoiesis

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Abstract: *Adrenal Myelolipoma is a rare nonfunctional adrenal tumor usually detected incidentally on imaging done for un-related cause or due to compressive symptoms or rarely due to adrenal hormone hyper secretion or malignant transformation. A case of 32yr female was complained with amenorrhea since 15yrs.The physical examination had no significant finding. Ultrasound and subsequently CT scan abdomen showed a large well-circumscribed mass m/s 4.2*3.2*5.2cm is seen in relation to the lateral limb of right adrenal. It shows multiple foci of fat attenuation.The absolute wash out is >60% suggestive of adrenal adenoma.*

Key Words*- Adrenal Myelolipoma, Extramedullary hematopoiesis*

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I Introduction

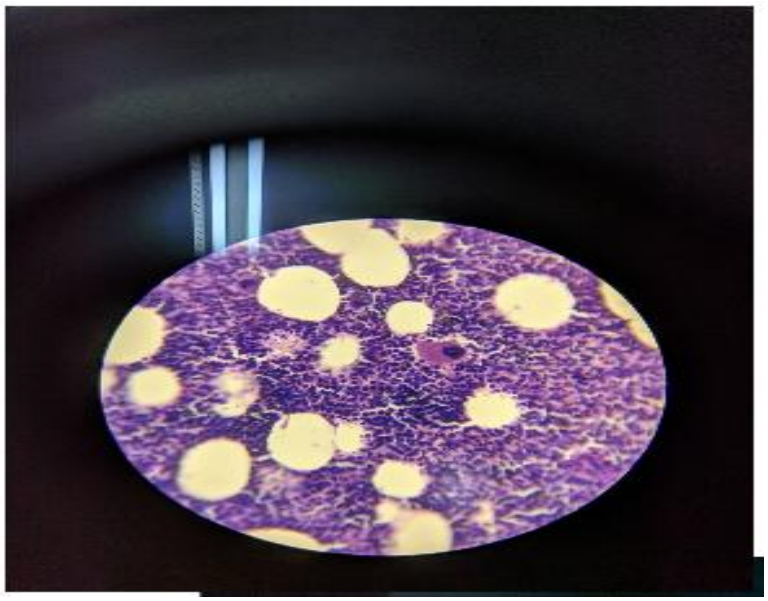
To the availbal literature, Myelolipomas account for approximately 3 to 5% of all primary adrenal tumors (1). Even though the incidence of these tumors are mysterious, it is belief to be between 0.08% and 0.4%, both gender appear to be equally affected by these tumor which are commonly found between the 50 to 70 age group (2).According to Brogna; the incidence in various series range from 0.01-0.2%.Adrenal myelolipoma is a rare tumor that is benign in nature, usually asymptomatic, unilateral, and nonsecreting. It is composed of variable mixture of mature adipose tissue and hematopoietic elements and develops within the adrenal gland. With the Adrenal myelolipoma is a rare tumor that is benign in nature, usually asymptomatic, unilateral, and nonsecreting. It is composed of variable mixture of mature adipose tissue and hematopoietic elements and develops within the adrenal gland.The tumor appears to affect men and women equally and most commonly found between the fifth and the seventh decade of life . Accounting for 3–5% of all primary tumors of the adrenals, the true incidence of these tumors is not known, although it is thought to be 0.08%–0.4%, with increased incidence noted in the later decades of life [3]. The majority of these tumors are unilateral, small, and asymptomatic although some bilateral myelolipomas have been described . They are generally nonsecreting in nature, and only one case of secreting is detected.

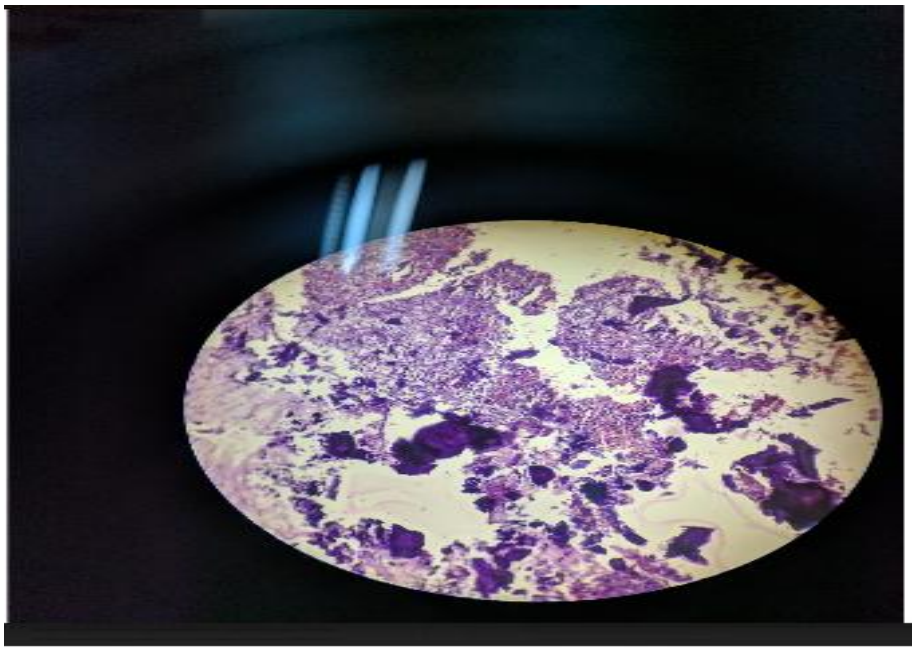
Case report -Nonencapsulated, bright yellow adipose-like tissue with hemorrhagic foci (contains marrow tissue) May be up to 5 kg

Gross picture-

The histopathology- showed an encapsulated tumor comprising predominantly of mature adipocytes with intervening areas of hematopoietic cells which are comprising of myeloid cells, erthyoid cells and occasional

megakaryocytes and a portion of normal adrenal at the periphery.Usually show normal trilineage hematopoiesis, but often with markedly increased megakaryocytes.Larger tumors may have hemorrhage, necrosis, calcification and cysts.





Extramedullary Hematopoeisis	
Myelolipoma	
Poorly circumscribed	
Circumscribed	
Overrun fat	
Fat integral to tumor	
No prominent lymphoid component	**Lymphocytes may be prominent**
Hematologic abnormalities associated	**No hematologic abnormalities**
Lipoblastoma with Extramedullary Hematopoiesis	
Myelolipoma	
Immature fat cells present	**No immature fat cells**
Areas with vascular myxoid stroma	**Lacks vascular myxoid stroma**
Usually under age 10, virtually always under 20	
Usually over age 40	

Usually over age 40 .Adrenal myelolipoma constitutes a rare entity in urological practice. They are composed of variable proportions of mature adipose tissue and active hematopoietic elements. They are also called "incidentalomas" since their diagnosis is based on autopsy or imaging modalities which are performed for reasons usually unrelated to adrenal diseases. Incidence ranges from 0.08% to 0.4%, and less than 300 cases were reported in the literature before 2000(3). However, their prevalence appears to be increasing up to 10%, due to the increased use of noninvasive and enhanced imaging technique (4).

There are several theories for the etiology and the natural history of adrenal myelolipoma(5) .However, the most widely accepted theory is adrenocortical cell metaplasia in response to stimuli, such as necrosis, inflammation, infection, or stress (6). This chronic stimulation to the adrenal gland, which is evidenced by the increased incidence of the lesion in the advanced age(7)could trigger the benign and malignant lesion.

Adrenal myelolipoma are infrequent, nonmalignant tumor composed of matured adipose tissue and hematopoietic elements (mixture of myeloid and erythroid cells). The most common site of myelolipoma is adrenal gland but it is infrequently present in the paravertebral regionas, retroperitoneum, pelvis, and mediastinum, as an isolated soft tissue mass .Adrenal myelolipoma equally affects both genders with similar rates and most cases are diagnosed between the 4th and 7th decades of life.

They are equally distributed in the right and left adrenal glands but in the majority of cases unilateral, although bilateral lesions are also reported in the literature

Adrenal myelolipomas are usually small (<4 cm) in diameter, however they can attain very large size. When the size exceeds 8cm, the term giant myelolipoma is preferred.

II Discussion

Adrenal myelolipoma constitutes a rare entity in urological practice. They are composed of variable proportions of mature adipose tissue and active hematopoietic elements.

They are also called "incidentalomas" since their diagnosis is based on autopsy or imaging modalities which are performed for reasons usually unrelated to adrenal diseases. Incidence ranges from 0.08% to 0.4%, and less than 300 cases were reported in the literature before 2000 (8). However, their prevalence appears to be increasing up to 10%, due to the increased use of noninvasive and enhanced imaging techniques (9).There are several theories for the etiology and the natural history of adrenal myelolipoma. However, the most widely accepted theory is adrenocortical cell metaplasia in response to stimuli, such as necrosis, inflammation, infection,or stress (10) .This chronic stimulation to the adrenal gland, which is evidenced by the increased incidence of the lesion in the advanced age [10], could trigger the development of benign as well as malignant tumors. The conditions often associated with adrenal myelolipomas include Cushing'sdisease, obesity, hypertension, and diabetes which can becharacterized as major adrenal stimuli (11).Other contem-porary authors have speculated about a stressful lifestyleand an unbalanced diet as factors that may be involved in the pathogenesis of this tumor. [Several case series have reported the predominance of the tumor in the right adrenal gland [12], which is yet to be explained.Ultrasonography, computed tomography, and MRI are all effective in diagnosing more than 90% of adrenal myelolipoma on the basis of identification of fat, with CTscan being the most sensitive(Since tumors are nonfunctional, endocrinological evaluations may not be useful, although there is a report of a secreting myelolipoma causing hypertension . The differential diagnosis should include renal angiomyolipoma, retroperitoneal lipoma, andliposarcoma (13).

In CT Scan has a discrete capsule and appears as well-delineated heterogeneous masses with regions of less than −30 Hounsfield units that correspond to low-density mature fat .The MRI evaluation have not much advantage over CT scan, however the myeloid tissue has low and moderate signal intensity in T1 and T2 images respectively whereas adipose tissue has high signal intensity in both T1 and T2 images. In case a diagnosis has not been confirmed, a fine needle aspiration can be helpful absolutely rule out malignancy .

Management - of adrenal myelolipoma should be considered on individual basis. Small lesions, which are asymptomatic and measure less than 5 cm, should be monitored over a period of 1-2 years with imaging controls. (11). It is suggested that symptomatic tumors or myelolipomas larger than 7 cm should be surgically excised , immediate surgery will be carried out, particularly for a large myelolipoma. If not, there may be spontaneous rupture of the tumor with hemorrhage . According to NIH guidelines, small, asymptomatic, non-functional myelolipoma may be managed conservatively, however giant adrenal myelolipoma should be excised as the high chances of malignancy and life threatening retroperitoneal or intra-tumoral bleeding is there . Trans-peritoneal open adrenalectomy is recommended, for larger tumors, however the laparoscopic adrenalectomy may be attempted, depending on the surgeon's expertise. The long term prognosis is excellent, if managed appropriately.

III Conclusion

The adrenal mass was detected incidentally when doing ultrasound of abdomen and followed by CT-Scan. Hyperechoic region was observed in USG due to the presence of adipose tissue in adrenal mass. CT-scan with intravenous contrast injection shows heterogeneous area with well-defined margin which clarify the presence of myelolipoma. Here in we describe a case of a young male with giant adrenal myelolipoma detected incidentally. Therefore, it is very significant to diagnose carefully for the planning of therapeutic management.

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