

Myelolipoma: A Rare Adrenal Incidentaloma

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Abstract

Background: Myelolipoma is a rarely encountered, adrenal incidentaloma diagnosed on the basis of its radiological features.

Aim: To describe a rare presentation with dual pathology.

Case presentation: Our patient, a 40-year-old lady presented with menorrhagia along with a large palpable uterine fibroid. Abdominal CECT detected a large, eight centimeter, left adrenal myelolipoma. After other possibilities were ruled out she underwent left adrenalectomy. Histopathology of the specimen revealed features of myelolipoma. She had an uneventful recovery and doing well now at six months follow up.

Conclusion: We present herewith a case of this uncommon tumour with dual pathology and discuss the clinical radiological and pathological features of adrenal myelolipoma.

Keywords: Incidentaloma; Adrenal; Myelolipoma

Introduction

Myelolipomas are rare, benign tumors composed of mature adipose tissue and hematopoietic elements (myeloid and erythroid cells) [1]. We describe a case of dual pathology where adrenal incidentaloma was successfully treated.

Case Report

A 40-year-old lady was referred to surgical outpatient clinic with an ultrasonologically detected adrenal mass, while being investigated for menorrhagia with a large pelvic mass. She did not have any symptoms related to the adrenal mass and was normotensive. Further biochemical investigations directed to adrenal pathology revealed a normal urinary VMA level, serum electrolytes and cortisol levels. The initial ultrasound scan suggested a well-defined, hyperechoic SOL in the left suprarenal region (94 x 80 mm) suggestive of a lipomatous tumour, along with a large intrauterine fibroid (119 x 96 mm). Subsequently, CECT of the abdomen revealed a large, well defined, mildly & heterogeneously enhancing mass lesion showing attenuation value of fat, involving left adrenal gland, suggestive of myolipoma (Figures 1 and 2). The right suprarenal was normal. In view of the large size of the uterine tumour and the fact that menorrhagia was under control, she underwent left adrenalectomy only in the first sitting, through a modified chevron incision. Macroscopical cut section of the specimen showed homogenous yellow surface with reddish streaking. Histopathology of the specimen revealed features of myelolipoma. She had an uneventful recovery and doing well now at six months follow up.

Discussion and Conclusion

Edgar von Gierke first described this lesion in the adrenal in 1905 but it was named, "myelolipoma", by Charles Oberling [2,3]. The adrenal gland is the most common site, but myelolipomas also (rarely) occur in extra-adrenal sites (14% of myelolipomas are extra-adrenal [4]) including the pelvis, mediastinum, retroperitoneum, and paravertebral region, as an isolated soft tissue mass [5].

It is usually hormonally inactive, and found in 0.08 to 0.2% of autopsy series [6,7] but comprise up to 15% of adrenal incidentalomas with the increasing use of noninvasive imaging [8] and account for 2.6% of all primary adrenal tumours [9]. Myelolipomas, affect both

sexes equally and usually occur during fifth and seventh decades of life [10]. Adrenal myelolipomas are in the majority of cases unilateral. However, they can also be bilateral [11]. Adrenal myelolipomas may be found coincidentally with other lesions in the adrenal glands, such as adenomas and less commonly with pheochromocytoma or metastases. These cases are described as "collision tumours" [12].

Adrenal myelolipomas vary in size, from several millimetres to more than 30 cm, and usually in the range of 2-10 cm in diameter [1,13]. The term giant myelolipoma is preferred when the size exceeds 8 cm [14], as seen in our case.



Figure 1: CT Coronal Image.

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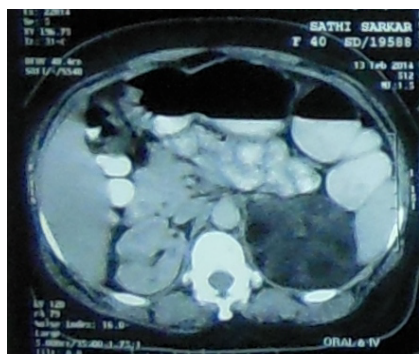


Figure 2: CT Axial Image.



Figure 3: Intraoperative Aspect.

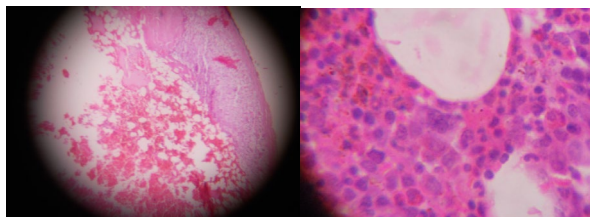


Figure 4: Histopathology (HE).

The exact pathogenesis of myelolipoma is not clear at present. Most theories involve the development and differentiation of either ectopic adrenal rests or hematopoietic stem cell rests in response to a triggering stimulus, particularly endocrine dysfunction [15]. Other researchers found clonal cytogenetic abnormalities, which suggested it was of neoplastic pathogenesis [16]. Chromosomal translocations (3;21) (q25;p11) detected in myelolipomas and in benign lipomatous neoplasia seen in patients with acute myelogenous leukemia or myelodysplastic syndrome suggest the origin of this tumor to be bone marrow, and may indicate that myelolipoma is derived from erroneously transferred erythroid cells [17].

Adrenal myelolipomas, which are usually asymptomatic and non-functioning, are generally important purposes [19]. The most frequent symptoms of large myelolipomas are non-specific abdominal pain, hematuria, renovascular hypertension and other symptoms secondary to the mechanical compression of adjacent organs [18-22]. Although rare, surgical emergencies such as retroperitoneal hemorrhage may also be encountered [21].

Adrenal myelolipomas are generally hormonally inactive, although there are case reports of their association with overproduction of adrenal hormones. They have been associated with overproduction of dehydroepiandrosterone- sulphate (DHEAS), congenital adrenal hyperplasia caused by 21-hydroxylase deficiency, congenital adrenal

α -hydroxylase deficiency, Cushing disease, Conn's syndrome, adrenal insufficiency, and pheochromocytoma [23-28].

The extensive use of abdominal CT-scan and magnetic resonance imaging has led to a dramatic increase in incidentally discovered adrenal masses that have also been called adrenal incidentalomas [29]. Radiological imaging typically reveals a well-circumscribed mass with a heterogeneous appearance due to the varying proportions of fat within the mass. Adipose tissue is characterized by low attenuation on CT imaging (i.e., - 25 to - 100 Hounsfield units). On MRI, fat displays high signal intensity on T1-weighted images whereas the myeloid component of these tumors has a T2-weighted signal. Contrast enhancement with CT scan or MRI will vary depending on the composition of the mass. Soft tissue components enhance whereas adipose tissue does not [30]. Calcification is present in a minority of cases on CT. Because of their characteristic appearance on CT, adrenal myelolipomas if small can usually be diagnosed without intervention and followed radiographically. Extra-adrenal myelolipomas, however, are more difficult to diagnose preoperatively because they are easily confused with several malignancies. If a definite diagnosis is needed, a fine-needle biopsy is indicated either under US or CT guidance.

Grossly, myelolipoma is a solitary circumscribed mass ranging in size from a few centimeters to 27cm [31]. The tumor is usually spherical to ovoid, well circumscribed, sometimes surrounded by a pseudo-capsule. The cut surface typically has a variegated appearance, with areas of greasy-appearing soft yellow tissue alternating with irregular areas of dark red-brown friable tissue, as we found in our case. Microscopically, the tumor is composed of a variable admixture of mature adipose tissue with islands and nests of hematopoietic elements of different percentages. The cellularity of hematopoietic precursors is variable and the three hematopoietic cell lineages (granulopoietic, erythropoietic and megakaryocytic) are present. In some cases, areas of infarction, hemorrhage, and rarely foci of calcification are noted [32]. Immunohistochemical staining and molecular testing is of no clinical or histological benefit.

When the diagnosis of myelolipoma is considered, it should be differentiated from other fat containing retroperitoneal tumors including retroperitoneal lipomas, retroperitoneal liposarcoma, extra-renal angiomyolipoma, extramedullary hematopoietic 'tumors', retroperitoneal leiomyosarcoma, primary or metastatic adrenal malignancy and teratomas [9,33,34].

Once adrenal myelolipoma is diagnosed, regular follow-up with sonography or CT is recommended and surgery is reserved for symptomatic cases. Some studies suggest surgical intervention for symptomatic tumors, growing tumors, or tumors larger than 10 cm to reduce the risk of developing abdominal pain or life-threatening hemorrhage [35]. From the reviewed papers, 17% cases whose tumor size was greater than 6 cm experienced spontaneous rupture. Therefore, elective surgery can prevent more severe symptom presentation and life-threatening progression and can allow accurate diagnosis in patients with tumors larger than 6 cm [33]. Castillo et al. advocated laparoscopic adrenalectomies for myelolipoma. [36,37]

In our case the tumour was almost 10 cm and presence of another large tumour in the pelvis made the case unusual and required early intervention to prevent rupture or haemorrhage and to reduce the confusion of double pathology.

Conflict of interests

Authors have no conflict of interests to disclose.

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