

Giant adrenal myelolipoma with contralateral staghorn calculus

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Abstract: Myelolipoma as the name suggests it comprises of hemopoietic elements (erythrocytes, leukocytes megakaryocytes) and adipose tissue. Most commonly it arises from adrenal gland. Mostly these masses are non-functional and benign in nature. Due to their large sizes or spontaneous haemorrhage which produces symptoms or they found incidentally (incidentaloma) during routine ultrasonographic examination of the abdomen. In this case report of a large myelolipoma in a 53-year-old male patient is discussed which is associated with large staghorn calculus with hydronephrosis of opposite kidney. Differential diagnosis of the adrenal tumour includes hormonal assessment and more sophisticated imaging techniques such as CT or MRI in order to investigate the morphology of the tumour.

Keywords: Myelo lipoma, ultrasonographic, hemorrhage.

INTRODUCTION:

Myelolipoma is a rare tumor. This disease was first describe by Gierke in 1905[1], and the term myelolipoma was coined by Oberling in 1929[2]. In a 1973 autopsy series, it had been estimated that the prevalence at autopsy was about 0.08%–0.4% [3,4]. 15% of all total Myelo lipomas are extra-adrenal, half of which arising from the presacral area. Thoracic, retroperitoneal, hepatic, pelvic, renal and gastric lesions have also been reported [5]. They may sometimes coexist with primary aldosterone's, congenital adrenal hyperplasia (CAH), pheochromocytoma, adenoma and Cushing's syndrome [6,7]. Having knowledge of such lesion is very important as this lesion can be managed conservatively.

CASE REPORT:

45 years old man presented to us with complain of right flank pain since 1 year with no history of fever, urinary complains. On examination Patient had a large intra-abdominal firm to hard fixed nontender lump palpated in left part of the abdomen. On USG right kidney had a stag horn calculus along with moderate hydronephrosis, in left side a large hyper echoic mass was seen in adrenal region having dimension of 19.5×13.5 cm. lesion pushed the kidney downwards. Further evaluation with CECT KUB revealed that right kidney had pelvic stag horn calculus of 30×17mm with multiple calyceal calculi with delayed excretion of contrast. On left side moderately large lesion was found having fat attenuation and

heterogeneous density, size of the lesion was 15.7cm×10.6cm in maximum dimension (Fig.-1).

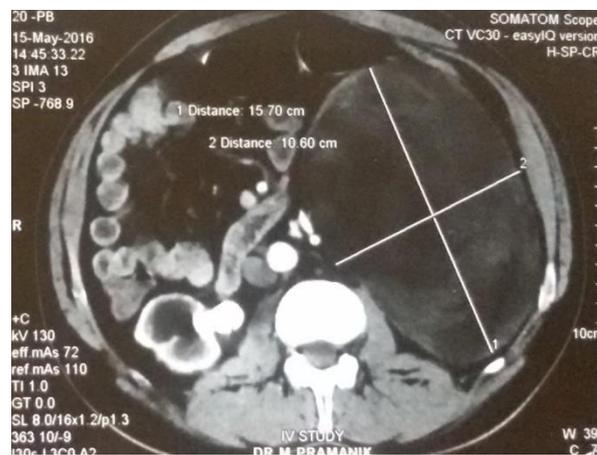


Fig 1: Contrast CT image of mass on left side along with right renal calculi with hydronephrosis

Urinary metanephrine was 165mcg/24 hours and urinary normetanephrine was 190mcg/24 hour which were found to be within normal limits. Platelet counts were $170 \times 10^3/\mu\text{l}$. After a preoperative workup, right DJ stenting along with left adrenalectomy were performed through subcostal incision by extraperitoneal approach. En bloc mass dissection was done sparing the left kidney. Gross examination of the specimen revealed a large, ovoid, and encapsulated mass with smooth external surface (Fig.-2).



Fig 2: Myelo lipoma after resection

On histopathology characteristic mixture of mature adipose tissue with hematopoietic elements (Fig. -3a, b) were found there were no signs of cell atypia, confirming our clinical diagnosis of adrenal myelolipoma.

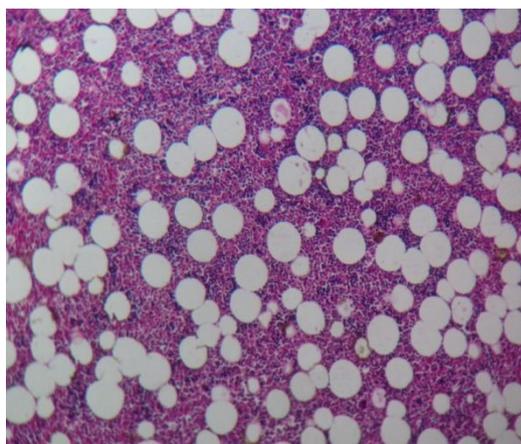


Fig-3a: 100 x magnification showing fat cells

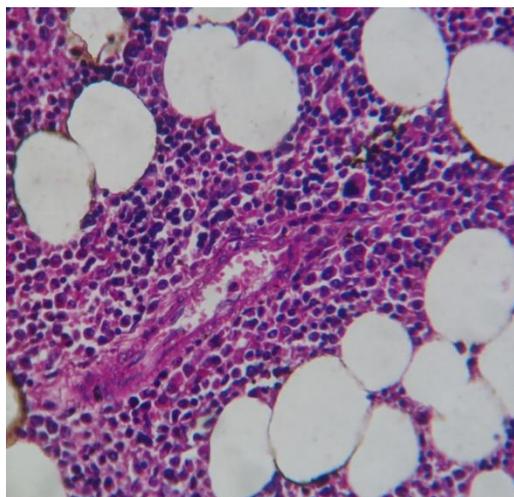


Fig.3b: 400x magnification showing myeloid component and fat cell.

Left partial staghorn calculi were latter on managed by right PCNL after 6 weeks of adrenal surgery.

DISCUSSION:

Adrenal Myelo lipoma is a rare finding in urological practice. As stated above it composed of variable proportions of mature adipose tissue along with hematopoietic elements in it. They are also termed as “incidentalomas” since their diagnosis is unrelated to adrenal diseases. There were less than 300 cases, reported in the literature till 2000 [8]. But now, their prevalence appears to be increasing up to 10%, due to the rise of non-invasive imaging techniques [9]. Myelo lipomas are often small and usually do not present with any hormonal secretion. The pathogenesis of Myelo lipoma is not understood at present. Multiple theories finely points out the development and differentiation of either ectopic adrenal rests or hematopoietic stem cell rests in response to a triggering stimulus, especially endocrine dysfunction [10]. Clonal cytogenetic abnormalities, is another mechanism proposed for its neoplastic pathogenesis [11]. Chromosomal translocations (3;21) (q25;p11) in benign lipomatous neoplasia and in Myelo lipomas ,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 [12].

On ultrasonography its appearances are different depending on individual tumour tissue component [13]. Usually be seen as a heterogeneous mass of mixed hyper- and hypoechoic components with the former primarily resulting from fatty portions. With CT appearance myelolipoma appears as an adrenal mass with fat-containing components. Fat amount in mass is variable. Small punctate calcifications may be seen in 25-30% of cases[14]. MRI can also delineate both macroscopic and microscopic fat by using explicit fat saturation technique and chemical shift imaging. Rarely CT guided percutaneous biopsy may be required to confirm the diagnosis. A multislice CT scan can differentiate adrenal Myelo lipoma from angiomyolipoma which arises from the upper pole of kidney [3].

The differential diagnosis for myelolipoma should include lipoma and liposarcoma. The small asymptomatic adrenal myelolipoma (<5cm) is usually managed conservatively follow-up with 6-12 months interval by ultrasound or CT. For tumor size is more 10cm, operation is recommended because of potential risk of hemorrhagic complication and malignant change [5, 15].

CONCLUSION:

After extensive search of the literature probably this is the first case of adrenal myelolipoma which has got asymptomatic incidentally diagnosed

with stag horn calculus in opposite kidney. Since in our case size of the mass was well above the 10 cm the nature of the mass was major concern for us we have surgically removed the mass first. Histopathologically it came to be a Myelo lipoma simultaneously the opposite kidney carefully managed with percutaneous nephrolithotomy.

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