Case Report

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Adrenal Myelolipoma with Osseous Metaplasia and Hypercortisolism

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Abstract

A 42yrs female presented with a large suprarenal mass with hypertension and diabetes mellitus. Dexamethasone suppression test was positive suggestive of Hypercortisolism/Cushing's syndrome. Right adrenalectomy was done. Histopathology revealed Adrenal myelolipoma with osseous metaplasia. It is a rare presentation reported for the first time with hypercortisolism.

Keywords: Adrenal Myelolipoma; Osseous Metaplasia; Hypercortisolism.

Introduction

Adrenal myelolipoma is a rare variety of adrenal tumor with prevalence of 0.08 to 0.4% on autopsy[1]. An incidence of 2 to 4% of all adrenal tumors and 10-15% of all incidental adrenal masses [10]. They are composed of mature adipocytes along with normal haematopoietic tissue. Adrenal myelolipoma present as a site of extramedullary haematopoiesis. Majority of incidentally discovered lesions are small and asymptomatic, however there are reports of large symptomatic lesions. Generally, these lesions are non functional but hypercortisolemia have been reported occasionally. We report a case of Adrenal myelolipoma with osseous metaplasia and hypercortisolism, not reported in English literature to the best of our knowledge.

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Case Report

A 42 year old female reported to our OPD with complaints of left flank pain for 10 days. There was no documented history of hypertension , diabetes mellitus or any other significant illness in the past.

She had no complaints of weight loss or weight gain, nausea, vomiting or dyspnoea. Plain X- ray KUB showed a small 6mm left upper ureteric calculi and the Ultrasonography showed an incidental finding of right supra renal mass. The IVP showed left upper ureteric calculi along with displacement of the right kidney (due to the supra renal mass). Both the kidneys showed normal function on the IVP. The CT Scan showed large well defined lesion of size 126x146x180 mm in right suprarenal region with fat density showing areas of calcification , necrosis producing mass effect over adjacent abdominal viscera (Figure 1). Lesion was abutting right posterolateral abdominal wall, also producing compression over IVC, pancreatic head and aorta, shifted towards left side. The perilesional fat planes were found clear and there was no lymphadenopathy. General examination of the patient showed hypertension. After admitting the patient at our centre the hormonal and urinary parameters were tested along with other routine investigations.

Although all the other parameters were normal the dexamethasone suppression test was found positive and also the patient was diagnosed with Diabetes mellitus. The patient was started on anti hypertensives and insulin. Left upper ureteric calculi

was managed conservatively because of its small size and no backpressure changes.

After controlling the blood sugar and blood pressure of the patient the patient underwent an open right adrenalectomy.Perioperative period was uneventful. The gross specimen consisted of a capsulated fibrofatty soft tissue mass weighing 1.758 Kg and measuring 24x17x15 cm in size (Figure 2). Histopathological examination of the specimen revealed almost whole of the adrenal gland to be replaced by mature adipose tissue and trilinear hematopoietic elements composed of myeloid cells with few clusters of erythroid cells and megakaryocytes. Areas of osseous metaplasia were also seen (Figure 3).

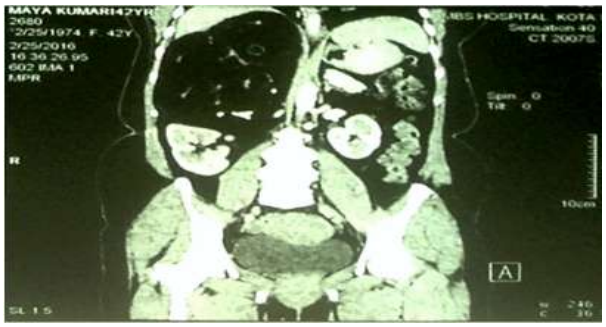

Fig. 1:

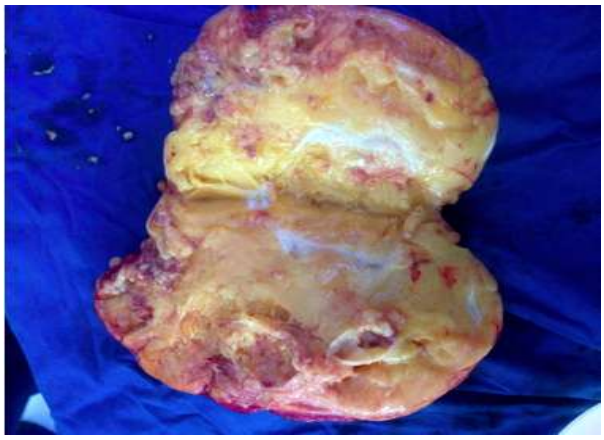

Fig. 2:

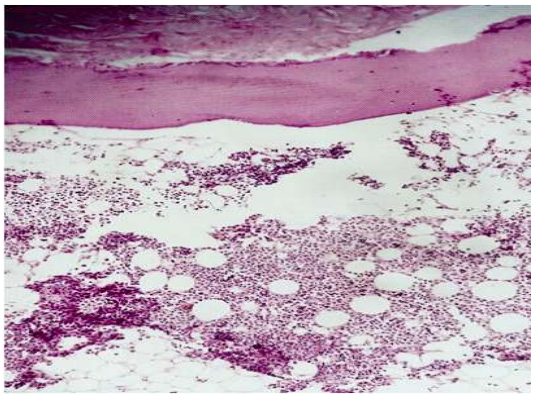

Fig. 3:

Discussion

Adrenal myelolipomas are benign masses comprising 2 to 4% of all adrenal tumors. It is commonly found in Fifth to seventh decades of life [10]. This tumor was initially described by Giercke in 1905 and the term myelolipoma was given by Oberling in 1929 [2].These tumors can range from few millimetres to 30 centimetres [3] and generally does not have any sex prediliction.Largest reported adrenal myelolipomawas measuring 31x24.5x11.5 and weighing 6 kgs [9]. Origin of these tumors is still speculative but among the various theories the theory of of metaplasia of reticuloendothelial cells of blood capillaries in the adrenal glands, in response to stress, infection and necrosis is the most accepted one [4]. Other theories proposed are bone marrow cells embolism, hyperplasia of heterotopic reticulum cell , etc.

Myelipomas are generally discovered incidentally as most of the patients are asymptomatic.Among the various imaging modalities CT Scan isthe most sensitive in detecting fat and hence in diagnosing these tumors. In a minority of the patients , i.e. in about 10% the tumors,due to larger size they can cause vague symptoms such as pain or heaviness [5]. Rapid increase in size of the mass can rarely cause abdominal pain and discomfort due to haemorrhage, necrosis and rupture. Acute haemorrhage is the most significant complication among these presenting with nausea, vomiting and anaemia. In general these tumors are hormonally naive but about 10% can be associated with Cushing's syndrome, Conn's syndrome, Congenital adrenal hyperplasia, pheochromocytomaetc [6].

Generally these masses are well circumscribed with surrounding pseudocapsule [7]. Biopsy of a myelolipoma reveals adipocytes with interspersed haematopoietic elements, comprising of myeloid and erthyroid precursors, along with megakaryocytes [8].

In our case the patient had hypertension along with diabetes mellitus and also Cushing syndrome on evaluation. After proper work up open adrenalectomy was done and the specimen was sent for histopathological examination. Grossly the specimen consisted of fibrofatty soft tissue mass measuring 24x17x15 cm. Outer surface was congested and capsulated (pseudocapsule). On serial sectioning the cut surface was fibrofatty with few haemorrhagic, cystic and calcified areas. Histopathology revealed that almost whole of the adrenal gland was replaced bymature adipose tissue and trilinear hematopoietic element composed of myeloid cells with few clusters

of erythroid cells and megakaryocytes. Areas of osseous metaplasia were seen.

The differential diagnosis of adrenal myelolipoma are retroperitoneal liposarcoma, lipoma and renal angiomyolipoma however proper imaging and histopahology after excision help in diagnosing these lesions.

Gagner et al., started laparoscopic adrenalectomy for the treatment adrenal tumours . Smaller lesions which are asymptomatic are monitored whereas larger symptomatic masses are excised.

Conclusion

Adrenal myelolipomas are rare tumors and adrenal myelolipoma with osseous metaplasia has not been reported till date. Other unique features were presence of Diabetes mellitus, hypertension and hypercortisolism. Prognostic significance of osseousmetaplasia needs to be determined.

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