Adrenal myelolipoma with osseous metaplasia and hypercortisolism

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INTRODUCTION

Adrenal myelolipomas are rare adrenal tumors generally diagnosed incidentally. A 42-year-old female reported to us with complaints of left flank pain attributable to her left ureteric calculi. On evaluation, a large adrenal mass was diagnosed along with hypercortisolism. After adrenalectomy, the histopathology revealed adrenal myelolipoma along with osseous metaplasia not reported in English literature, to the best of our knowledge till date.

CASE REPORT

A 42-year-old female reported to our outpatient department 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 dyspnea. Plain X-ray of the kidney-ureter-bladder region showed a small 6 mm left upper ureteric calculi, and the ultrasonography showed an incidental finding of right suprarenal mass. The intravenous urography showed left upper ureteric calculi along with displacement of the right kidney (due to the suprarenal mass) [Figure 1]. Both the kidneys showed normal function on the intravenous pyelogram. The computed tomography (CT) scan showed large well-defined lesion of size 126 mm × 146 mm × 180 mm in the right suprarenal region with fat density showing areas of calcification, necrosis producing mass effect over adjacent abdominal viscera [Figure 2]. Lesion was abutting right posterolateral abdominal wall, also producing compression...
over inferior vena cava, pancreatic head, and aorta shifted toward the 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 center, 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 but the patient had no overt external clinical features of Cushing syndrome. The patient was started on antihypertensives and insulin. Left upper ureteric calculi were managed conservatively because of its small size and no back pressure 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 24 cm × 17 cm × 15 cm in size [Figure 3]. 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 [Figures 4 and 5].

**DISCUSSION**

Adrenal myelolipomas are benign masses comprising 2%–4% of all adrenal tumors. It is commonly found in the fifth to seventh decades of life. This tumor was initially described by Giercke in 1905, and the term myelolipoma was given by Oberling in 1929.[3] These tumors can range from few millimeters to 30 cm and generally do not have any sex predilection. Largest reported adrenal myelolipoma was measuring 31 × 24.5 × 11.5 and weighing 6 kg.[4] Origin of these tumors is still speculative but among the various theories, the theory of metaplasia of reticuloendothelial cells of blood capillaries in the adrenal glands, in response to stress, infection, and necrosis, is the most accepted one.[5] Other theories

**Figure 1:** Intravenous urography showing displaced right kidney

**Figure 2:** Computed tomography scan showing large adrenal mass

**Figure 3:** Cut section of the excised specimen

**Figure 4:** Histopathological slide showing myelolipoma with osseous metaplasia
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proposed are bone marrow cells embolism, hyperplasia of heterotopic reticulum cell, etc.

Myelolipomas are generally discovered incidentally as most of the patients are asymptomatic. Among the various imaging modalities, CT scan is the 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. Rapid increase in size of the mass can rarely cause abdominal pain and discomfort due to hemorrhage, necrosis, and rupture. Acute hemorrhage is the most significant complication among these presenting with nausea, vomiting, and anemia. In general, these tumors are hormonally naive but about 10% can be associated with Cushing’s syndrome, Conn’s syndrome, congenital adrenal hyperplasia, pheochromocytoma, etc.

In general, these masses are well circumscribed with surrounding pseudocapsule. Biopsy of a myelolipoma reveals adipocytes with interspersed hematopoietic elements, comprising myeloid and erythroid precursors, along with megakaryocytes.

In our case, the patient had hypertension along with diabetes mellitus and also Cushing syndrome on evaluation. After proper workup, open adrenalectomy was done, and the specimen was sent for histopathological examination. Grossly, the specimen consisted of fibrofatty soft-tissue mass measuring 24 cm × 17 cm × 15 cm. Outer surface was congested and capsulated (pseudocapsule). On serial sectioning, the cut surface was fibrofatty with few hemorrhagic, cystic, and calcified areas. Histopathology revealed that almost whole of the adrenal gland was replaced by mature adipose tissue and trilinear hematopoietic element composed of myeloid cells with few clusters of erythroid cells and megakaryocytes. Areas of osseous metaplasia were seen.

Cushing syndrome patients require steroid replacement after surgery, until the contralateral gland recovers its function. Plasma cortisol monitoring helps in determining when the steroid is to be tapered and stopped. The differential diagnosis of adrenal myelolipoma is retroperitoneal liposarcoma, lipoma, and renal angiomylipoma. However, proper imaging and histopathology after excision help in diagnosing these lesions.

Gagner et al., started laparoscopic adrenalectomy for the treatment adrenal tumors. 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 the presence of diabetes mellitus, hypertension, and hypercortisolism. Prognostic significance of osseous metaplasia needs to be determined.

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