Bilateral Adrenal Myelolipoma: A Case Report and Review of Literature

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Abstract

Myelolipoma of the adrenal gland is a rare, benign and endocrinologically inactive tumor. The characteristic histological picture shows mature adipose tissue and normal hematopoietic elements that resemble bone marrow. These tumors are clinically quiescent and most of the cases are detected during imaging procedures obtained for other reasons. Very rarely these tumors give rise to symptoms, usually abdominal pain due to tumor enlargement. Most of them occur unilaterally but rarely bilateral occurrences have been reported in literature. We report a rare case of bilateral adrenal myelolipoma along with management strategies and review of literature.

Introduction

Adrenal myelolipoma is a rare benign adrenal neoplasm presenting in 0.08%-0.4% of population based on autopsy series. In recent years, the development of improved imaging techniques has increased the detection rate and thus adding these tumors in the heterogeneous group of ‘incidentalomas’ of the adrenal gland.

Case Report(s)

This is a 42 year old woman who presented with sudden onset of continuous, dull aching pain in the right upper abdomen of few weeks duration. There was no relation to food intake. There was no associated nausea, vomiting or jaundice. No urinary symptoms. Physical examination revealed height of 147 cm, weight of 54.0 kg, and blood pressure of 140/90 mmHg. Abdominal examination revealed bilateral loin mass which was smooth, moving with respiration and was bimanually palpable and ballotable.

CT abdomen revealed a 12.5cm x12cm x11cm well circumscribed, heterogeneous, non enhancing mass with hypo dense areas with attenuation values suggestive of fat [Figure-1] in the right supra-renal region causing downward displacement of the right kidney and 9.7cm x9.5cm x6cm left suprarenal mass with similar hypodense areas with features consistent with myelolipoma. The 24-hour test of urine catecholamines was normal, ruling out pheochromocytoma. Both the plasma and urinary cortisol tests were normal, ruling out Cushing’s syndrome. The plasma aldosterone was normal.

A provisional diagnosis of bilateral adrenal myelolipoma was made. Since patient was asymptomatic, surgery was planned. Right adrenalectomy was done after confirming with intra-operative frozen section. Biopsy was also taken from the left side tumor and sent for frozen section. After ruling out malignancy it was decided not to remove the left side tumor as it was asymptomatic and also to circumvent cortisol insufficiency. Post operative serum cortisol was within normal range and patient was discharged after one week. Patient was subjected to regular follow up. Histopathology was reported as adrenal myelolipoma [Figure-2].

Follow up period was clinically uneventful for 4 years after which, patient developed dull aching pain in the left hypochondrium. CT abdomen also showed tumor enlargement now measuring17cm x10cm x8cm with features consistent with myelolipoma. Elective left adrenalectomy done after confirming with frozen section and the patient was on supplemented with oral steroids. Post operative period was uneventful. Patient was advised for regular follow up.

Discussion

Adrenal Myelolipoma is a benign, usually hormonally non functioning tumor. Meaglia and Schmidt proposed that myelolipoma arises due to metaplasia of the reticulo-endothelial cells of blood capillaries in the adrenal gland in response to stimuli such as infection, stress or necrosis. Commonly these tumors are asymptomatic but rarely are symptomatic owing to large or increasing size and complications like intra-lesional hemorrhage or necrosis. Imaging features of myelolipoma depend on the proportions of fat, myeloid element, hemorrhage or calcification present. Ultrasound of the abdomen may be helpful in the diagnosis of Adrenal Myelolipoma due to its ability
to differentiate the echogenic, fatty supra-renal mass from the normal kidneys. Computerized tomography (CT) is regarded as the most sensitive imaging procedure and demonstration of fat density (Hounsfield units -100 to -200) within an adrenal mass by CT is virtually diagnostic of adrenal myelolipoma [2] which is the most common lipomatous tumor of adrenal gland. [1,3] However, if CT shows non-homogenous features or tumor more than 6 cm, the possibility of underlying malignancy cannot be ruled out. In this situation, an imaging guided needle biopsy or on-table frozen section must be performed to confirm the diagnosis. USG and CT have greatly enhanced the diagnostic accuracy of adrenal myelolipoma in recent years. [4,5,6]

The classical histological picture shows predominantly mature adipose tissue with hematopoietic tissue, consisting of immature and mature cells of granulopoiesis, erythropoiesis, megakaryocytes and lymphoid cells, interspersed in the adipose tissue. Following diagnosis patient can be clinically followed up for development of new symptoms or progression of existing symptoms. Routine radiological follow up is not mandatory as the tumor exhibits variable growth and moreover the size of tumor neither correlates with symptoms always nor suggests the imminence of life threatening shock associated with spontaneous hemorrhage. The recommended treatment of adrenal myelolipoma is conservative and surgery is mostly reserved for symptomatic patients. [7,8].

In our case, a provisional diagnosis of adrenal myelolipoma was made after CT. Since the patient was symptomatic we planned to proceed with surgery. Right adrenalectomy was done after confirming with frozen section. The left side tumor was not removed as it did not cause symptoms and to avoid adrenal insufficiency. Postoperative stay was uneventful and serum cortisol level was within normal range. On clinical follow-up, patient remained asymptomatic for 4 years after which she developed continuous dull aching pain in the left hypochondrium. CT abdomen was remarkable for increase in size of tumor without any other intraslesional complications as mentioned earlier. Hence left adrenalectomy was done and patient was started on steroid supplementation.

Thus we conclude that treatment of adrenal myelolipoma should be individualized and surgical treatment is favorable for bilateral larger tumors even if asymptomatic in contrary to the recommended conservative treatment for asymptomatic tumors.

Reference

Illustrations

Illustration 1

CT abdomen revealed a 12.5cm x12cm x11cm well circumscribed, heterogeneous, non enhancing mass with hypo dense areas with attenuation values suggestive of fat.

Illustration 2

Histological picture shows predominantly mature adipose tissue and hematopoietic tissue.
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