Case Report

Adrenal myelolipoma masquerading as retroperitoneal sarcoma

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ABSTRACT

Adrenal myelolipomas are rare, benign, hormonally inactive tumors. Most patients are asymptomatic. We report a case of 47-year-old female who was evaluated for joint pain and pedal edema, detected to have a retroperitoneal mass in the right sub-hepatic region, clinically simulating a retroperitoneal sarcoma, and diagnosed as adrenal myelolipoma by histopathology.

Key words: Adrenal, myelolipoma, retroperitoneal sarcoma

INTRODUCTION

Adrenal myelolipoma is a rare benign tumor first described by Gierke in 1905. The name adrenal myelolipoma was given by Oberling in 1929. Myelolipoma is composed of mature adipose tissue and hemopoietic tissue. It is mostly discovered incidentally on imaging of the abdomen done for nonadrenal-related reasons or at autopsy.[1] Most adrenal myelolipomas are small and asymptomatic, but spontaneous rupture of larger tumors have also been reported.[2] We report a case of 47-year-old female with retroperitoneal mass in the right sub-hepatic region and diagnosed as adrenal myelolipoma by histopathology.

CASE REPORT

A 47-year-old female presented with joint pains in lower limb of 6 months duration. General examination was normal except for pedal edema. Computed tomography (CT) scan of the abdomen showed 16 cm × 15 cm predominant fat density lesion with septation in the right sub-hepatic region causing mass effects on retroperitoneal structures [Figure 1a]. CT scan findings were suggestive of retroperitoneal liposarcoma, adrenal myelolipoma, or angiomyolipoma.

Exploratory laparotomy was done with excision of retroperitoneal tumor and excision of the capsule over the inferior surface of the liver. Intraoperative findings showed large well-encapsulated tumor of size 16 cm × 12 cm in the suprarenal region. Kidney was free of tumor. Gross examination showed a well-encapsulated yellowish nodular mass measuring 18 cm × 15 cm × 9 cm and weighed 1.2 kg. Cut surface showed yellowish and circumscribed brownish areas [Figure 1b]. Microscopy revealed adrenal tissue at the periphery, beneath which was seen a mass composed of varying proportions of mature adipose tissue admixed with areas of hemopoietic tissue containing erythroid, myeloid, megakaryocytic, and lymphoid elements [Figure 1c and d]. No evidence of other associated elements like the pheochromocytoma, adrenocortical hyperplasia, or adenoma was present on microscopic examination. Thus, diagnosis of adrenal myelolipoma was made.

DISCUSSION

Adrenal myelolipomas account for 2.5% of primary adrenal tumors.[3] They are usually noted in mid to late adult life, and there is no sex predilection reported in most series. They are usually unilateral, and measure <5 mm in diameter.[1] There are few case reports of giant adrenal myelolipoma. The largest reported tumor weighed 6 kg.

Pathogenesis of adrenal myelolipomas is controversial. Differential proliferation of embryonic mesenchymal

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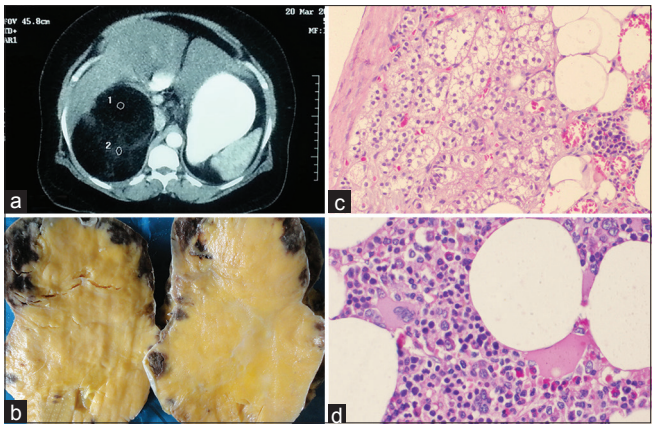

Figure 1: (a) Postcontrast axial computed tomography section of the abdomen at the suprarenal level. A relatively well-defined, predominantly fatty attenuation lesion at the right suprarenal level, showing few enhancing septae and peripheral solid areas within; the right adrenal gland is not visualized separately. (b) Gross examination showed a well-encapsulated nodular mass with yellowish and brownish areas. (c) Residual adrenal tissue at the periphery of the tumor (H and E, ×200). (d) Adipose tissue component admixed with hemopoietic elements (H and E, ×400)

rests in adrenal glands into myeloid and adipose tissue, hematogenous seedling of bone marrow emboli, and metaplasia of mesenchymal cells as a result of chronic stress are the proposed theories.[4] Large tumors may cause spontaneous hemorrhage or hemoperitoneum due to traumatic rupture of the adrenal gland. The characteristic CT and magnetic resonance findings of myelolipoma include demonstration of areas of the obvious fat within a well-defined adrenal mass.[5] The differential diagnosis following the CT analysis in our case included retroperitoneal liposarcoma, angiomyolipoma, and adrenal myelolipoma. Myelolipomas have indications for surgical removal when it is symptomatic and for larger, asymptomatic tumors more than 4 cm due to the risks of spontaneous haemorrhage.[6] Periodic radiographic imaging is to be done for small nonfunctional tumors. Functioning adrenal myelolipoma can be clinically mistaken for endocrine causes like pheochromocytoma.[7] A novel association of adrenal myelolipoma and keratoconus has been described.[8] There are case reports of occurrence of adrenal myelolipoma with adrenal cortical carcinomas and adrenocortical adenomas in the same adrenal gland.[9,10] To conclude, adrenal myelolipoma being a rare entity, awareness about this entity is very much needed to avoid radical surgical procedures.

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