Case Report

Asymptomatic Giant Adrenal Myelolipoma

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INTRODUCTION

Adrenal myelolipomas are nonfunctioning benign tumors composed of adipose tissue and hemopoietic elements resembling bone marrow. Before the availability of modern imaging, they were detected only at autopsy with an incidence of 0.08% to 0.2%.[(1)] These tumors are now being picked up more frequently with the availability of computed tomography and ultrasonography, but they are usually small and isolated.[(2)] The first excision of a myelolipoma was carried out in 1922. Since then, more than 100 cases of surgically resected adrenal myelolipomas have been documented. The management of incidentally found tumors and their pre-operative diagnosis is discussed. We describe the largest asymptomatic adrenal myelolipoma weighing 5.5 kg.

CASE REPORT

A 56-year-old man with type 2 diabetes mellitus presented to the diabetic clinic with an ulcer in his left foot due to ill-fitting shoes. He had an amputation of his left hallux for diabetic complications 6 years previously. On direct questioning, he denied any significant loss of appetite, abdominal bloating, urinary symptoms, or change of bowel habit. He had lost approximately 25 kg of weight over a 2-year period. On examination, he was thin with a distended abdomen. There was evidence of gynecomastia, but no jaundice or lymphadenopathy. He had a large right upper quadrant mass extending down to his pelvis with a recurrent umbilical hernia. His left leg was affected by cellulitis.

He was admitted to hospital for further management of his foot ulcer, control of his diabetes mellitus, and investigation of his abdominal mass. Ultrasonography revealed a large predominantly hyperechoic mass within the right retroperitoneum. Computed tomography of the abdomen confirmed a massive soft tissue mass containing fatty and soft tissue elements. It was well circumscribed and extended to the anterior, posterior, and right lateral abdominal wall without invasion. It displaced the liver anteriorly and to the left, and the right kidney inferiorly. The mass extended as far as the right pelvic brim. There was no ascites or lymphadenopathy (Figures 1 and 2). It suggested the diagnosis of an incidental retroperitoneal tumor, probably a liposarcoma.

It was decided that the best option for this patient would be a resection. His pre-operative workup, including complete blood count, biochemistry, and tumor markers, were all within normal limits. He was explored through

Figure 1. Liver hilum stretched by the huge retroperitoneal tumor.

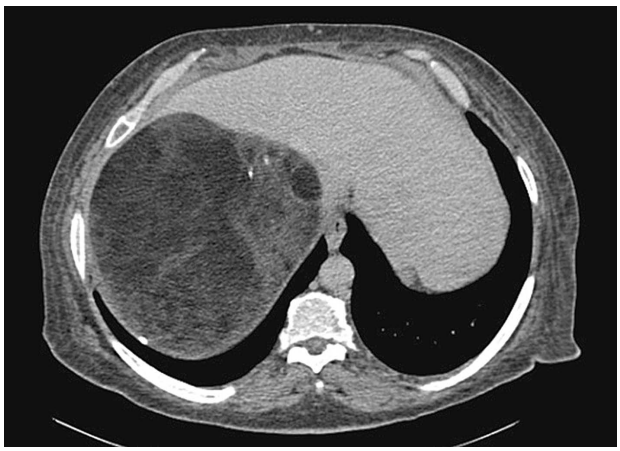

Figure 2. Huge retroperitoneal tumor displacing the liver.

a reverse L incision. A huge heterogeneous retroperitoneal tumor was found in the right upper quadrant of his abdomen. The liver was pushed anteriorly with the gallbladder and hilum stretched over the mass. The inferior vena cava and the right renal vein were grossly stretched and compressed by the mass. There was no bowel involvement. The right kidney was freed from the tumor down to the renal capsule. The tumor was then excised preserving all the other structures. A small nodule was seen superficially in the right lobe of the liver and a nonanatomical wedge resection using the TissueLink DS3. 5C radiofrequency dissector was performed. The umbilical hernial defect was repaired with PROCEED intraperitoneal mesh, stapled in situ. The procedure took approximately 4 hours and was associated with no transfusion requirement. Postoperatively, the patient was extubated immediately and made an uneventful recovery. He was discharged on the 10th postoperative day.

The tumor was a large lobulated greyish yellow mass with a pseudocapsule. It weighed 5.5 kg and measured 28 × 26 × 17 cm. On sectioning, there were areas representing well-differentiated fat as well as areas of hemorrhage and infarction. Histological examination revealed that the lesion arose from the adrenal gland and consisted of lobules of mature adipocytes in the background of hemorrhage and some sclerosis. Extramedullary hematopoiesis was seen. Mitotic figures were not seen. These features suggested the diagnosis of a myelolipoma. Further immunohistochemistry was positive, confirming the presence of erythroblasts, and cytogenetic analysis was negative for MDM2-CDK4. The latter is present in atypical lipomatous tumors/well-differentiated liposarcoma. The liver lesion turned out to be a sclerosed hemangioma.

DISCUSSION

Adrenal myelolipomas are rare, benign, nonfunctioning tumors composed of mature adipose tissue and hemopoietic elements.[1] In 1979, Boudreaux and coworkers reported the first case ever of a giant myelolipoma.[3] The tumor, resected en bloc with the kidney and retroperitoneal soft tissues, weighed 5.9 kg. However, the weight of the actual tumor was not reported. The largest adrenal myelolipoma to be resected was reported by Akamatsu and colleagues from Japan.[4] The tumor measured 31 × 24.5 × 11.5 cm and weighed 6 kg. The only other reported myelolipoma weighing more than 5 kg was reported by Wilhelmus and colleagues who described a tumor weighing 5.5 kg.[5] Our case represents the 4th largest adrenal myelolipoma, weighing 5.5 kg. However, this represents the largest asymptomatic adrenal myelolipoma. All the previously described adrenal myelolipomas weighing more than 5 kg have been symptomatic at presentation.

Giant adrenal tumors are quite rare and mostly affect patients in their 4th to 6th decades. The association of myelolipoma with obesity, hypertension, chronic disease, and malignancies have been described.[6] Our patient's mass had

become visible with recent profound weight loss. His only comorbidity was diabetes mellitus. However, due to the frequent association of obesity and hypertension in the affected group of older patients, these are probably incidental associations.[7]

The pathogenesis of adrenal myelolipomas is still unclear. Various theories have been proposed including autonomous proliferation of bone marrow cells during embryogenesis, degeneration of the epithelial cells of the adrenal cortex, metaplasia of mesenchymal cells, and bone marrow emboli.[5] Adrenal myelolipomas are usually asymptomatic. Only when the tumor attains an enormous size does it cause symptoms. The most common symptom is abdominal pain, probably due to hemorrhage, tumor necrosis or compression of surrounding structures.[7] They are usually nonfunctioning, though occasional secretion of catecholamines or hormones have been reported.[8]

The classic myelolipoma is lucent on plain films, echogenic on ultrasonography, and avascular on angiography. Computed tomography is a very sensitive modality for diagnosis, because it can show an adrenal mass with tissue density similar to that of fat. It also has signal intensity similar to that of fat on T-weighted magnetic resonance imaging. However, distinguishing between myelolipoma and liposarcoma is impossible radiologically, as happened in our case.

Previously, most of the patients with myelolipoma underwent surgical resection for a suspected malignant neoplasm. Nowadays, with the frequent detection of these "incidentalomas," the treatment has been subjected to a lot of debate.[9] When the tumor is 4 cm or smaller and asymptomatic, follow-up with computed tomography is recommended. If symptoms occur, surgery should be carried out promptly, especially in large myelolipomas, as spontaneous rupture of the lesion with hemorrhage is possible.[10] Resection of larger lesions due to diagnostic doubt is probably inevitable.

CONFLICT OF INTEREST

None declared.

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