Giant myelolipoma of left adrenal gland simulating a retroperitoneal sarcoma

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ABSTRACT

Adrenal myelolipoma is a rare benign tumor composed of an admixture of adipose and hematopoietic tissue. Most tumors are asymptomatic, small in size, right-sided, and usually detected incidentally. Though they are often smaller than four cm in diameter, they can reach larger sizes. Giant adrenal myelolipomas are extremely rare tumors having only about 10 reported cases in the literature. Presently, a case is being reported in a 59-year-old lady who presented with abdominal distention and dragging sensation in the upper left abdomen. Computed tomography of the abdomen revealed a retroperitoneal tumor which was suspected to be a retroperitoneal sarcoma. Other fat containing retroperitoneal tumors such as a lipoma, myelolipoma and teratoma were also considered as possibilities. All hormonal studies related to adrenal gland were within normal limits. The patient underwent surgical resection of the tumor. The resected mass was 23 cm × 16 cm × 9 cm in size and weighed 3.3 kg. Histopathological examination revealed the tumor to be an adrenal myelolipoma. This present case of adrenal myelolipoma is unusual in view of its left-sided location and its large size which clinically and radiologically simulated a retroperitoneal sarcoma.

Key words: Adrenal gland, differential diagnoses, giant myelolipoma, large retroperitoneal mass

INTRODUCTION

Adrenal myelolipomas are benign, biochemically inactive neoplasms composed of an admixture of mature adipose and hematopoietic tissues in varying proportion. First described by Gierke in 1905, the term “myelolipoma” was coined by Oberling in 1929.[1] Most myelolipomas appear as unilateral adrenal masses; however, similar lesions may develop in extra-adrenal sites in the retroperitoneum.[2] Myelolipomas are often smaller than four cm in diameter, although they can reach larger sizes. Most of the adrenal myelolipomas arise from the right adrenal gland.[3] Radiologically, adrenal myelolipomas may mimic other fat containing retroperitoneal lesions, like liposarcoma, teratoma, and extramedullary hematopoiesis.[4] Recent cytogenetic studies have shown that adrenal myelolipoma is a clonal lesion.[5]

Giant adrenal myelolipomas are extremely rare tumors with only about 10 reported cases.[6] The largest adrenal myelolipoma reported in the literature weighed 6 kg and

Access this article online

Quick Response Code:
Website:
www.ijamhrjournal.org
DOI:
10.4103/2349-4220.172896

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How to cite this article: Saha M, Dasgupta S, Chakrabarti S, Chakraborty J. Giant myelolipoma of left adrenal gland simulating a retroperitoneal sarcoma. Int J Adv Med Health Res 2015;2:122-5.
measured 31 cm × 24.5 cm × 11.5 cm. Such large tumors may mimic other retroperitoneal soft-tissue tumors including sarcomas and can give rise to the substantial diagnostic dilemma.

**CASE REPORT**

A 59-year-old female patient presented with a history of gradually increasing abdominal distention and dragging sensation in the upper left side of the abdomen for the last 4 months. She was a known hypertensive. Clinically, the patient had good general health with stable vitals. There was a palpable swelling in the left hypochondrium, approximately 15 cm × 10 cm in size. The swelling was firm with a regular surface. It was nontender, and the overlying skin was not fixed to the swelling.

Routine blood tests, routine urine examination, chest X-ray and electrocardiography were done. The blood biochemical report identified the patient as diabetic. The other tests were within normal limits.

Computed tomography (CT) of the abdomen showed a large fat density lesion in the retroperitoneum (left the lumbar region) with areas of soft-tissue attenuation. The approximate dimensions of the lesion were 19.5 cm × 14.6 cm [Figure 1]. Radiologically, the tumor was suspected to be a retroperitoneal sarcoma. Other fat containing retroperitoneal tumors such as a lipoma, myelolipoma, and teratoma were also considered as possibilities. CT guided fine needle aspiration cytology (FNAC) was done. Smears showed mature fat cells, collagen fibers, fibroblasts and red blood cells. A cytodiagnosis of lipoma was given after correlating with clinicalradiological features.

An extensive hormone assay was done which included serum cortisol, serum catecholamines, serum dopamine, urinary vanillylmandelic acid, 17-ketosteroid, and testosterone levels. The results were found to be within normal levels.

Subsequently, the patient underwent laparotomy. A huge encapsulated space occupying lesion was found in the left renal region, pushing the left kidney down and the stomach, pancreas, intestine anteriorly. The mass was removed and sent for histopathological examination.

Gross examination showed that the mass was 23 cm × 16 cm × 9 cm in size and weighed 3.3 kg. The outer surface was brown to yellowish in color. Cut surface showed yellow, greasy areas along with large areas of hemorrhage [Figure 2].

Microscopic examination revealed a capsulated mass with compressed normal adrenal tissue in the periphery. The mass was predominantly composed of mature adipocytes interspersed with normal hematopoietic elements. Trilineage hematopoietic was noted [Figures 3 and 4]. A final diagnosis of giant adrenal myelolipoma was awarded.

The patient was on 1-year follow-up, which was uneventful.

**DISCUSSION**

Adrenal myelolipomas are rare tumors of the adrenal gland, which usually occur in the fifth to a seventh decade with no gender preference. The incidence of these tumors is very low-varying between 0.08% and 0.4%. The association of adrenal myelolipoma with obesity, hypertension, atherosclerosis, diabetes mellitus and malignancy has been described previously. In our case, the patient was in the sixth decade, and she was both hypertensive and diabetic. The tumor was found to be located in the left side, contrary to its common occurrence from the right adrenal gland. Udupa et al. [10]
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and Brogna et al.[11] are among few others who have reported similar rare cases of left-sided giant adrenal myelolipoma in the past.

Four distinct clinicopathological patterns of this tumor have been described — isolated adrenal myelolipoma, adrenal myelolipoma with hemorrhage, extra-adrenal myelolipoma and myelolipoma associated with other adrenal diseases like adrenal adenomas or endocrine disorders including Cushing’s syndrome, Conn’s syndrome and congenital adrenal hyperplasia.[8]

Adrenal myelolipomas generally get noticed incidentally during imaging studies of the abdomen or in the autopsy, hence referred to as “incidentalomas.” This name reflects their small size and asymptomatic nature in most cases. Myelolipomas are mostly smaller than four cm in diameter, although they can occasionally reach larger sizes.[3] The giant adrenal myelolipomas are extremely rare tumors.[6] Although most adrenal myelolipomas are usually asymptomatic, abdominal, or flank pain and an abdominal mass are the most common presenting symptoms. The compression of neighboring structures by large tumors (mass effect), infection, tumor necrosis and intratumoral hemorrhage are the main complications associated with it.[12] Despite the huge size of the tumor, our patient presented only with abdominal mass without complications commonly associated with tumors of such large dimensions.

On CT scan, detection of a fat component within the adrenal gland is suggestive of myelolipoma. The presence of fat renders a negative Hounsfield unit value to the lesion. The attenuation value of myelolipoma is slightly higher than that of retroperitoneal fat owing to the simultaneous presence of hematopoietic elements. However, these guidelines are not sufficient to conclusively differentiate between giant adrenal myelolipoma and well-differentiated liposarcoma.

Radiological analysis takes account of location, size, vascularity, and local invasion. Well-differentiated liposarcomas do not invade renal parenchyma, similar to adrenal myelolipomas, thereby making it nearly impossible to distinguish one from the other.[13]

In our patient, based on CT scan observation, differential diagnoses of retroperitoneal sarcoma, lipoma, myelolipoma, and teratoma were considered. Temizoz et al.[13] stated that the precise diagnosis of retroperitoneal masses is difficult on the basis of radiology alone. Differential diagnoses of adrenal myelolipomas may include lipoma, liposarcoma, myelolipoma, and angiomylipoma. Other disorders to be considered are extramedullary hemopoiesis, lymphoma, and amyloidosis. The authors, therefore, stressed on the need of histopathological analysis of retroperitoneal tumors.

Daneshmand and Quek[3] opined that fine needle aspiration should be considered if the diagnosis of adrenal myelolipoma cannot be conclusively made using noninvasive imaging techniques. The presence of mature adipocytes and hematopoietic elements in cytologic smears is diagnostic of myelolipoma. Though our patient underwent CT guided FNAC, the smears only demonstrated mature fat cells, collagen fibers, fibroblasts and red blood cells. Since the smears did not reveal any hematopoietic elements, a provisional diagnosis of lipoma was offered.

In case of myelolipomas of smaller dimensions, that show no progression, simple observation is recommended using imaging techniques. Surgical treatment is advised when the diameter of tumor exceeds six cm, the tumor shows the tendency of fast growth or causes clinical disorders.[14] In the present case, due to the large size and suspicion of retroperitoneal sarcoma, surgical excision of the mass was undertaken.
Differential diagnoses of myelolipoma, on histopathologic examination, include renal angiomyolipoma, retroperitoneal lipoma, and liposarcoma. Angiomyolipomas are composed of a variable mixture of mature fat, thick walled blood vessels and smooth muscles. Lipoma consists of only mature fat. Low-grade liposarcomas are infiltrative with cellular atypia and lipoblasts. In our case, histopathological sections revealed that the mass was predominantly composed of mature adipocytes interspersed with normal hematopoietic elements and a final diagnosis of giant adrenal myelolipoma was rendered.

It has long been a topic of debate whether myelolipomas are true neoplasms or a reactive process. Bishop et al. successfully demonstrated nonrandom X chromosome inactivation in the hematopoietic elements and fat in 8 of 11 myelolipomas from female patients suggesting a clonal origin of these tumors.

The case is being presented to highlight the rarity of giant myelolipoma, and its unusual occurrence in the left adrenal gland. Clinically, as it simulates a retroperitoneal sarcoma, awareness of this rare entity is critical for its accurate diagnosis and appropriate management.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Kumar S, Jayant K, Prasad S, Agrawal S, Parma KM, Roat R, et al. Rare adrenal gland emergencies: A case series of giant myelolipoma presenting with massive hemorrhage and abscess. Nephrourol Mon 2015;7:e22971.
2. De Leelis RA, Mangray S. The adrenal gland. In: Mills SE, Carter D, Greenerson JK, Reuter VE, Stoler MH, editors. Sternberg's Diagnostic Surgical Pathology. 5th ed. Philadelphia: Lipincott Williams and Wilkins; 2010. p. 545-86.
3. Daneshmand S, Quck ML. Adrenal myelolipoma: Diagnosis and management. Urol J 2006;3:71-4.
4. Baker KS, Lee D, Huang M, Gould ES. Presacral myelolipoma: A case report and review of imaging findings. J Radiol Case Rep 2012;6:1-9.
5. Bishop E, Eble JN, Cheng L, Wang M, Chase DR, Orazi A, et al. Adrenal myelolipomas show nonrandom X-chromosome inactivation in hematopoietic elements and fat: Support for a clonal origin of myelolipomas. Am J Surg Pathol 2006;30:838-43.
6. Chakrabarti I, Ghosh N, Das V. Giant adrenal myelolipoma with hemorrhage masquerading as retroperitoneal sarcoma. J Midlife Health 2012;3:42-4.
7. Mukherjee S, Pericleous S, Hutchins RR, Friedman PS. Asymptomatic giant adrenal myelolipoma. Urol J 2010;7:66-8.
8. Dodd S, Singhal T, Leake T, Sinha P. Management of an incidentally found large adrenal myelolipoma: A case report. Cases J 2009;2:8414.
9. Hofmockel G, Dämmrich J, Manzanilla Garcia H, Frohmüller H. Myelolipoma of the adrenal gland associated with contralateral renal cell carcinoma: Case report and review of the literature. J Urol 1995;153:129-32.
10. Udupa S, Usha M, Visveswara RN, Desai MG. Left-sided giant adrenal myelolipoma secreting catecholamine. Indian J Pathol Microbiol 2012;55:389-91.
11. Brogna A, Scalisi G, Ferrara R, Bucceri AM. Giant secreting adrenal myelolipoma in a man: A case report. J Med Case Rep 2011;5:298.
12. Bano S, Yadav SN, Chaudhary V, Garga UC. Symptomatic giant adrenal myelolipoma associated with cholelithiasis: Two case reports. Urol Ann 2012;4:55-60.
13. Temizoz O, Genchellac H, Demir MK, Unlu E, Ozturk H. Bilateral extra-adrenal perirenal myelolipomas: CT features. Br J Radiol 2010;83:e198-9.
14. Hsu SW, Shu K, Lee WC, Cheng YT, Chiang PH. Adrenal myelolipoma: A 10-year single-center experience and literature review. Kaohsiung J Med Sci 2012;28:377-82.
15. Talwalkar SS, Shaheen SP 2nd. Extra-adrenal myelolipoma in the renal hilum: A case report and review of the literature. Arch Pathol Lab Med 2006;130:1049-52.