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CASE REPORT

Giant Adrenal Myelolipoma Associated with Beta-Thalassemia: A Case Report

Kamani F, Mohammadi SS, Hessami R, Rezvani N, Peirovi H, Tavassoli S

Department of Surgery, Taleghani Hospital, Shahid Beheshti University of Medical Sciences, Tehran, I.R.Iran.

Adrenal myelolipomas are rare benign tumors composed of mature adipose tissue and hematopoietic elements that resemble bone marrow. They most commonly occur in the adrenal gland, yet only comprise about 3% of all adrenal tumours. Although several hypotheses have been proposed as to the cause of myelolipoma, the causative process is still not clearly understood. The majority of myelolipomas are asymptomatic. It usually represents as accidental finding in imaging studies. There are some case reports about coincidence of adrenal giant myelolipoma with RBC disorders. In this case report, we present a β thalassemic 26 year old male patient who had giant adrenal myelolipoma.

Key Words: Adrenal Myelolipoma, Beta-thalassemia major, Abdominal CT scan, Hemoglobin electrophoresis

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Introduction

Adrenal myelolipomas are rare benign tumors composed of mature adipose tissue and hematopoietic elements that resemble bone marrow. They most commonly occur in on the adrenal gland, yet only comprise about 3% of all adrenal tumours. Although several hypotheses have been proposed as to the cause of myelolipoma, the etiology is still not clearly understood. The majority of myelolipomas are asymptomatic. It usually represents as accidental finding in imaging studies. There are some case reports on the coincidence of adrenal giant myelolipoma with RBC disorders. In this case report we present a β thalassemic 26 year-old male patient who presented with a giant adrenal myelolipoma.

Correspondence: SS Mohammadi. Department of surgery, Shahid Beheshti University of Medical, sciences, Taleghani Hospital, Tehran, Iran
*E-mail:*Drssm59@yahoo.com

Case presentation:

A 26 year-old male patient presented because of the accidental finding of an adrenal mass during ultrasonography. He was a known case of major beta thalassemia and had a history of periodical packed cell blood transfusions. The patient had had splenectomy when he was 5 years old. Abdominal CT scan revealed a well-defined and a non-calcified, heterogeneously enhanced mass lesion, with a large deposit of macroscopic fat, measuring 93 in 87 in 61mm, in diameter, located just superior to the right kidney and resulting in the somewhat downward displacement of the right kidney. No significant abdominal, pelvic abdominal lymphadenopathy or ascites was noted, nor was there any pleural effusion on either side. The liver and pancreas were of normal

diameter and showed homogeneous attenuation. The spleen was not observed, indicating the patient had undergone splenectomy. (Fig.1)

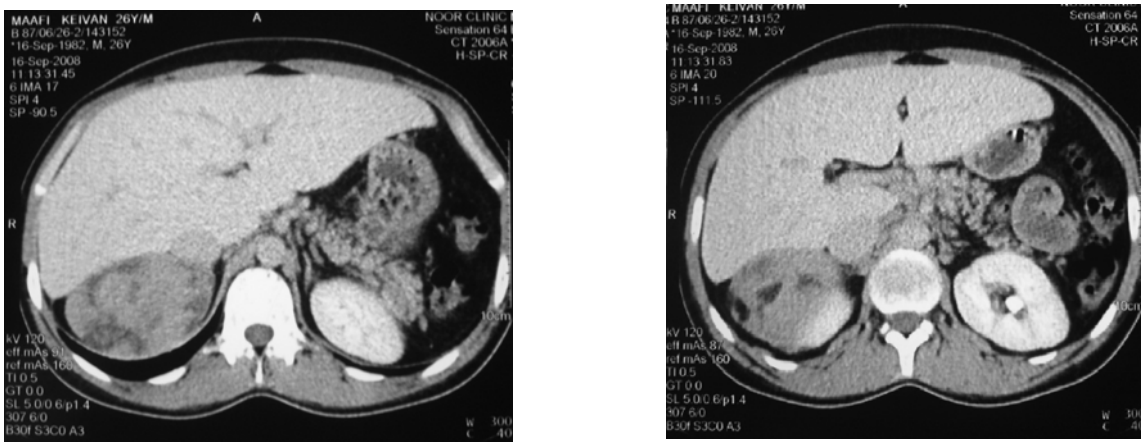

Fig. 1: Abdominal CT scan of the patient, spleen is not observed.

Laboratory tests showed Hb 9.2 g/dL, Hct 29%, MCV 82.6 fL, MCH 26.2 pg, MCHC 31.7 g/dL, RBC 3.5×10^6/µL, WBC 28400/µL, and platelet count 472×10^3/µL; biochemistry showed blood sugar 92 mg/dL, BUN 14 mg/dL, Cr 0.7 mg/dL, K 3.6 meq/L. Hemoglobin electrophoresis showed HbA: 65 HbF: 32.4 HbA2: 2.6

Endocrine studies showed normal adrenal function with urine free cortisol 68 ug/24hours, urine metanephrine 112 ug/24 hours, urine normetanephrine 294ug/24hours, and vanyl mandelic acid (VMA) 4.8mg/ 24hours.

The patient underwent exploratory laparotomy with right adrenalectomy. Findings were a 9x9x6cm tumor on the right adrenal, meckle diverticulum on 60Cm from ileosecal valve, normal liver on shape and size, and gallbladder with no stone.

Gross pathologic study of tumor revealed encapsulated adrenal mass measuring 9.5x7x4 cm and 160gr in weight. Microscopic study of tumor showed adrenal tissue mainly replaced by hematopoietic cells includeing megakariocytes, erythroid and myeloid lineage admixed by mature lipomatous tissue. (Fig. 2)

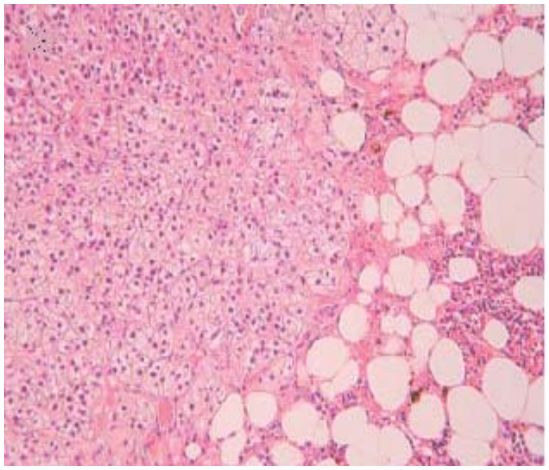

Fig. 2: Pathology of the tumor

Discussion

Myelolipoma is an uncommon benign tumor of the adrenal gland, containing mature adipose tissue, interspersed with hematopoietic tissue. It accounts for almost 4% of adrenal tumors[1], and has a varying prevalence ranging from 0.08 to 0.4% at autopsy. Estimated prevalence varies from 0.08 - 0.4%. It is usually under 5 cm in size, and giant myelolipomas are rare.[2,3]

Most tumors are small and found accidentally on CT scan or autopsy. Pain is one of its

symptoms, caused by hemorrhage, necrosis, or compression. Adrenal myelolipoma was first described by Gierke in 1905 and named by Oberling in 1929.[4,5] Male and female patients are affected equally and there is no laterality preference.[6-8] Majority of tumors are unilateral, however, a few cases with bilateral adrenal myelolipomas have been reported.[9]

Several explanations of this tumor's development have been offered. One theory by D.C. Collins suggests that a myelolipoma represents a site of extramedullary hematopoiesis.[10] The most widely accepted theory is the existence of metaplasia of the reticuloendothelial cells of blood capillaries in the adrenal gland in response to stimuli, such as necrosis.[7] It is also conceivable that increased erythropoietin levels may play a role in the pathogenesis of this tumors, simulating metaplasia of embryonic stem cells in myeloid tissue.[1]

Most myelolipomas are asymptomatic and hormonally nonfunctional. Occasionally, patients present with nonspecific abdominal or flank pain, secondary to intratumoral or peritumoral hemorrhage, tumor necrosis, or mechanical compression from tumor bulk. Other rare presenting symptoms include hematuria and abdominal mass. Endocrine dysfunction is also reported with adrenal myelolipoma, including conditions such as Cushing's syndrome, Conn syndrome, and congenital adrenal hyperplasia.[11]

Coincidence of adrenal myelolipoma with hematological disorders has rarely been described in literature; sickle cell anemia and thalassemia are two such disorders. Myelolipoma in thalassemic patients usually presents as giant or bilateral but no cause for this has been determined.[2,12]

The imaging modality of choice, is CT scan as it readily identifies the presence of mature fat, which has very low CT attenuation values (-30 to -100 Hounsfield units), within an adrenal mass. The typical size and lipoma appearance usually facilitate accurate diagnosis, and the presence of hematopoietic tissue makes its differentiation from other tumors possible.[4] Occasionally, myelolipomas have very little recognizable fat, and an MRI maybe helpful in distinguishing these lesions. The presence of fat is best demonstrated on T1-weighted images, as the fat containing area in a myelolipoma should be equal in signal intensity to that of subcutaneous and retroperitoneal fat at all pulse sequences.[4,13,14] The myeloid elements have a low signal intensity on T1-weighted images and moderate –signal intensity on T2-weighted images.[15]

On pathologic examination of these tumors, gross appearance of adrenal myelolipoma reveals yellow areas with the appearance of adipose tissue alternate with hemorrhagic foci composed of bone marrow tissue. Microscopic findings include bone marrow elements and mature fat. In contrast to other extra medullary foci of hematopoiesis in adults, adrenal myeolipoma is practically always accompanied by normal bone marrow.[16]

In conclusion, giant adrenal myelolipoma should be included in the differential diagnosis of adrenal masses. It is usually asymptomatic and maybe found only on autopsy. Adrenal myelolipoma is usually nonfunctional but endocrine disorders have been reported in some cases. Coincidence of thalassemia with myelolipoma support the theory that erythropoietin plays a role in the pathogenesis of myelolipoma.

References

1. Au WY, Tam PC, Ma SK, Lam KY. Giant myelolipoma in a patient with thalassemia intermedia. Am J Hematol 2000; 65: 265-6.
2. Hadjigeorgi C, Lafoyianni S, Pontikis Y, Van Vliet-Constantinidou C. Asymptomatic mye-

lolipoma of the adrenal. Pediatr Radiol 1992; 22: 465-6.
3. Tanaka D, Oyama T, Niwatsukino H, Nakajo M. A case of asymptomatic giant myelolipoma of the adrenal gland. Radiat Med 1998; 16: 213-6.
4. Cyran KM, Kenney PJ, Memel DS, Yacoub I. Adrenal myelolipoma. Am J Roentgenol 1996; 166: 395-400.
5. Yildiz L, Akpolat I, Erzurumlu K, Aydin O, Kandemir B. Giant adrenal myelolipoma: case report and review of the literature. Pathol Int 2000; 50: 502-4.
6. El-Mekresh MM, Abdel-Gawad M, El-Diasty T, El-Baz M, Ghoneim MA. Clinical, radiological and histological features of adrenal myelolipoma: review and experience with a further eight cases. Br J Urol 1996; 78: 345-50.
7. Meaglia JP, Schmidt JD. Natural history of an adrenal myelolipoma. J Urol 1992; 147: 1089-90.
8. Sharma MC, Kashyap S, Sharma R, Chumber S, Sood R, Chahal R. Symptomatic adrenal myelolipoma. Clinicopathological analysis of 7 cases and brief review of the literature. Urol Int 1997; 59: 119-24.
9. Spinelli C, Materazzi G, Berti P, Cecchi M, Morelli G, Miccoli P. Symptomatic adrenal myelolipoma: therapeutic considerations. Eur J Surg Oncol 1995; 21: 403-7.
10. Collins DC. Formation of bone marrow in the suprarenal gland. Am J Path 1932; 8: 97-106.
11. Hisamatsu H, Sakai H, Tsuda S, Shigematsu K, Kanetake H. Combined adrenal adenoma and myelolipoma in a patient with Cushing's syndrome: case report and review of the literature. Int J Urol 2004; 11: 416-8.
12. Kelekis NL, Alexopoulou E, Brountzos EN, Ladis V, Boussiotou A, Kelekis DA. Giant adrenal myelolipoma with minimal fat content in a patient with homozygous beta-thalassemia: appearance on MRI. J Magn Reson Imaging 2003; 18: 608-11.
13. Casey LR, Cohen AJ, Wile AG, Dietrich RB. Giant adrenal myelolipomas: CT and MRI findings. Abdom Imaging 1994; 19: 165-7.
14. Hoeffel CC, Kowalski S. Giant myelolipoma of the adrenal gland: natural history. Clin Radiol 2000; 55: 402-4.
15. Rao P, Kenney PJ, Wagner BJ, Davidson AJ. Imaging and pathologic features of myelolipoma. Radiographics 1997; 17: 1373-85.
16. Hunter SB, Schemankewitz EH, Patterson C, Varma VA. Extraadrenal myelolipoma. A report of two cases. Am J Clin Pathol 1992; 97: 402-4.