Giant adrenal myelolipoma with a spontaneous rupture: report of a case

Mielolipoma gigante de glândula adrenal com ruptura espontânea: relato de um caso

Sergio Renato Pais Costa[1], Sergio Henrique Couto Horta[2], Alexandre Cruz Henriques[3]

ABSTRACT

Myelolipoma is a benign tumor of the adrenal cortex, which is non-functioning and often asymptomatic. It is generally diagnosed in imaging tests by chance. Rupture and bleeding of myelolipoma is an infrequent complication, and may lead to the formation of a hematoma or, less usually, result in a massive retroperitoneal hemorrhage. The compression of adjacent structures by retroperitoneal hematoma of adrenal origin is possible but not frequent. Indications for surgery continue to be a difficult and controversial decision, since a conservative treatment and a watchful waiting may be indicated in some cases. However, the surgical procedure has been more precisely indicated to symptomatic patients (pain or infection) or even to uncertain diagnosis (malignant neoplasm). The authors present a case of a giant myelolipoma of the adrenal gland that presented both complicated spontaneous rupture (abdominal pain) and uncertain diagnosis by radiological images (computed tomography). The patient underwent a surgical resection with favorable postoperative outcome and pain relief.

Keywords: Myelolipoma; Adrenal gland neoplasms; Case reports

RESUMO

O mielolipoma é um tumor benigno raro oriundo do córtex adrenal, não funcionante e normalmente assintomático. É geralmente descoberto de incidentalmente por meio de exames de imagem. A ruptura e o sangramento são complicações pouco habituais que podem evoluir tanto para uma hemorragia retroperitonial maciça, quanto para um hematoma. A compressão de estruturas adjacentes por um hematoma retroperitonial de origem adrenal é um evento pouco frequente. Atualmente, a resolução cirúrgica dessa complicação continua sendo uma decisão difícil e controversa, haja vista que o tratamento expectante pode ser realizado em alguns casos. Contudo, a exploração cirúrgica tem sido indicada para os casos sintomáticos (dor ou infecção) ou em que haja dúvida diagnóstica com neoplasia maligna. Os autores apresentam um caso de mielolipoma gigante de glândula adrenal, o qual evoluiu com ruptura espontânea complicada por hematoma organizado e dor abdominal. Concomitantemente, havia dúvida diagnóstica quanto à neoplasia retroperitoneal pelo exame de imagem (tomografia computadorizada). O paciente foi operado e evolui satisfatoriamente, com resolução da síndrome dolorosa abdominal.

Descritores: Mielolipoma; Neoplasias das glândulas supra-renais; Relatos de casos

INTRODUCTION

Adrenal gland myelolipoma is a benign uncommon tumor that grows slowly. Its origin is in the mature adipose tissue entwined with hematopoietic elements. It has a low incidence ranging from 0.08 to 0.2%[1]. In some occasions, it has been associated with other endocrinopathies, such as obesity, Cushing's syndrome or Addison's disease[2]. In general, these tumors are small and they seldom exceed 5 cm in diameter. Usually, they are asymptomatic and diagnosis is made by chance in most cases. Occasionally, in case of a bulky tumor, abdominal pain, bowel obstruction or even vomits are the symptoms observed[3]. Less frequently, it may cause spontaneous retroperitoneal hemorrhage, which occurs in larger tumors[4].

The authors of this study report a case of a giant myelolipoma in the left adrenal gland with spontaneous rupture, resulting in an organized retroperitoneal hematoma. The patient complained of abdominal pain irradiating to the lumbar region and bowel obstruction. Preoperative computed computed tomography (CT) scan suggested retroperitoneal sarcoma (liposarcoma). Therefore, the patient was submitted to a multivisceral resection with a favorable postoperative follow-up and abdominal pain relief.

Study carried out at Serviço de Cirurgia Geral do Hospital de Ensino of Faculdade de Medicina do ABC – FMABC, Santo André (SP), Brasil.

[1] Master's degree, Assistant physician at the Department of General Surgery of Hospital de Ensino da Faculdade de Medicina do ABC – FMABC, Santo André (SP), Brazil.

[2] Assistant physician, Department of General Surgery at Hospital de Ensino da Faculdade de Medicina do ABC – FMABC, Santo André (SP), Brazil.

[3] Head of the Department of General Surgery at Hospital de Ensino da Faculdade de Medicina do ABC – FMABC, Santo André (SP), Brazil.

Corresponding author: Sergio Renato Pais Costa – Avenida Pacaembu, 1.400 – CEP 01234-200 – São Paulo (SP), Brasil – Tel.: 11 3666-2299 – e-mail: sergiorenatopais@terra.com.br

Received on Apr 12, 2009 – Accepted on Jul 16, 2009

CASE

A 45-year-old, Caucasian male patient. He was suffering from systemic arterial hypertension (SAH), type II *diabetes mellitus* (DM) and obesity. SAH and DM were under control with oral medication.

The patient complained of abdominal pain during the last three months. The pain was in the left hypocondrium irradiating to the lumbar region. During hospitalization, he presented vomits and changes in the bowel movement (intestinal obstruction). On physical examination, he presented obesity (BMI = 36 kg/m^2) associated with a palpable fiber elastic mass measuring approximately 20 x 10 cm, poorly defined, in the left hypocondrium and epigastrium. For initial workup, he was submitted to an upper digestive endoscopy and total abdomen ultrasound, but both had no changes. Later on, an abdomen computed tomography (CT) scan showed a mass that was predominantly solid with a hypoattenuating central area (cystic) and peripheral heterogeneous enhancement, with IV contrast medium administration. This lesion measured 15 x 10 cm and was in the retroperitoneum, in the left adrenal kidney topography (with no proper definition of its origin). There was some doubt whether the lesion dislocated or involved the distal portion of the pancreas and the splenic angle of the colon (Figures 1 and 2).

Serum tumor markers (CEA and CA 19,9) were measured, as well as vanil mandelic acid, metanefrines and cathecolamines in a 24-hour urine sample. All the exams were within the normality range. In other biochemical tests, the patient presented only a discreet anemia (hemoglobin = 11.2 g/dl; hematocrit = 36 g/dl).

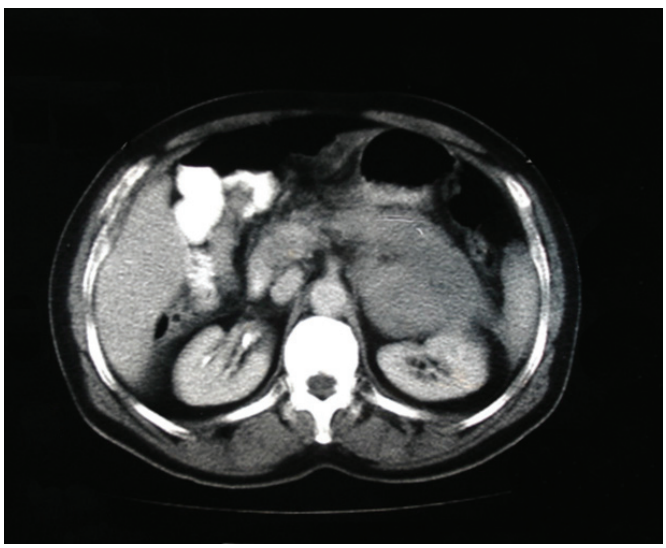

Figure 1. Preoperative CT scan: large solid heterogeneous lesion in the retroperitoneum

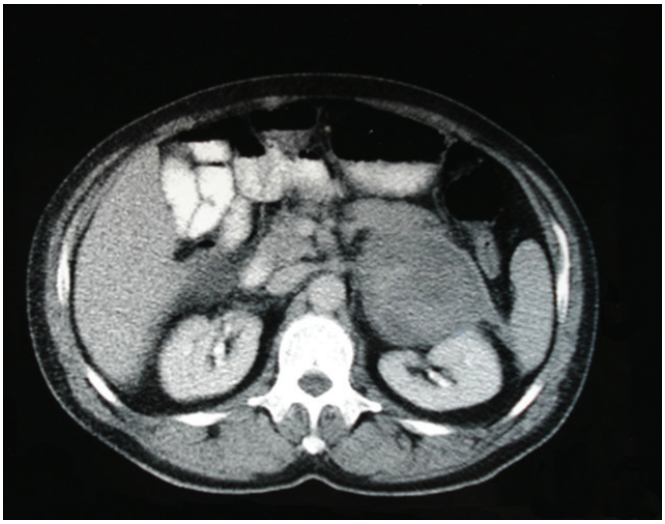

Figure 2. Preoperative CT aspect: lesion showing diffuse hypoattenuated areas after the administration of intravenous contrast

The main preoperative presumptive diagnosis was retroperitoneal liposarcoma. With progressive worsening of abdominal pain and signs of peritoneal irritation, the patient was submitted to exploratory laparotomy. During surgery, a predominantly solid lesion was observed, with cystic and hemorrhagic areas, measuring 15 x 10 cm, involving the left kidney and the left adrenal gland with firm adherences to pancreatic body, tail and splenic flexure of the colon. Since there was no certainty regarding the origin of the tumor, it was decided to make a monobloc resection of the affected structures. Hence, radical left nephrectomy extended with distal pancreatectomy and splenectomy, and left hemicolectomy (with primary anastomosis) were performed. The patient had a follow-up without complications and was discharged seven days after surgery. In the macroscopic histological examination, there was a tumor measuring 15 x 10 cm in the larger axes. Microscopic examination revealed a mature lipomatous neoplasm with no areas of atypia, presence of hematopoietic cells (erythroblasts, granulocytes and megakaryocytes) associated with recent and organized hemorrhage. The pancreatic segment was partially autolyzed, with areas of recent hemorrhage in the surrounding adipose tissue.

The kidney presented arteriosclerosis, nephrosclerosis, nephrocalcinosis with foci of chronic pyelonephritis and intense inflammatory process with recent hemorrhage. Based on these findings, the diagnosis of adrenal myelolipoma with spontaneous rupture was made.

Figure 3 shows the late postoperative CT image. Two years after resection, the patient has no relapses and remains asymptomatic.

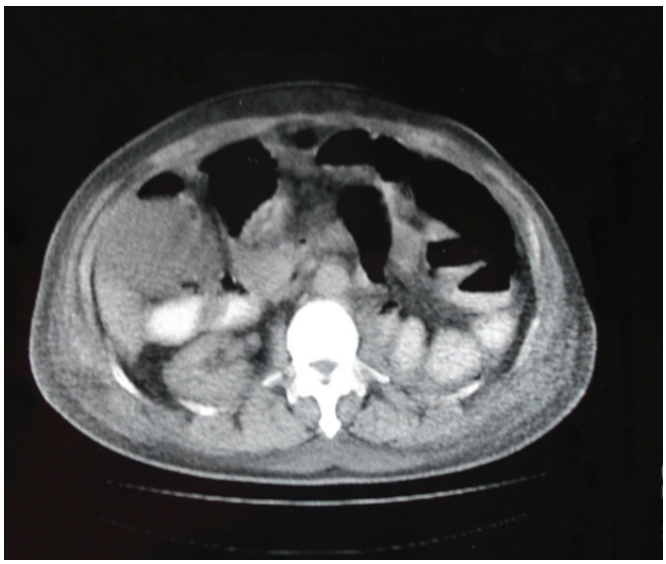

Figure 3. Postoperative CT aspect: the surgical site was filled by the colon (superior cut)

DISCUSSION

Adrenal myelolipoma is a benign tumor, described by Gierke in 1905. However, the current designation was given by Oberling, who named it as myelolipoma in 1922. This tumor has similar distribution in both sexes and although there are reports in children and elderly, it affects more frequently individuals between the fifth and seventh decades of life. Usually, it measures less than 5 cm, but there are reports of tumors reaching 34 cm and weighing nearly 6 kg[5].

Patients are generally asymptomatic. In most cases, the diagnosis is made by chance due to a finding in imaging tests or even during a surgical procedure due to other reasons. Nevertheless, when they reach large dimensions, they can present symptoms related to the compression of neighboring organs, hemorrhage or even tumoral necrosis. Abdominal pain, hematuria and constipation are the most frequent symptoms observed[5-7]. In some rare cases, large size tumors may cause spontaneous rupture leading to severe hemorrhage. More frequently, a retroperitoneal hematoma is formed and, as a consequence, it may become organized, as in this case[5,7,8].

The differential diagnosis for myelolipoma includes especially angiomyolipoma, retroperitoneal sarcomas (liposarcoma) and adrenal carcinoma. Imaging tests, in particular CT scan or magnetic resonance imaging (MRI), may differentiate the above-mentioned lesions from the myelolipoma. Some criteria, such as tumor density (fat), lesion heterogeneity and existence of regular edges are decisive for a proper therapeutic approach[9,10]. MRI is not better than CT scan, but should be used in cases with uncertain diagnosis[10]. Partial resection of retroperitoneal sarcomas is associated with high rates of recurrence and poor survival rates, because of the possibility of a tumor implant in the cavity, therefore diminishing its curability. Both radiological and macroscopic aspects of the tumor during the intraoperative assessment are not totally reliable.

Moreover, intraoperative frozen biopsy of mesenchymal tumors, especially low-grade neoplasms such as well-differentiated liposarcomas, is frequently difficult to assess and may lead to mismanagement. In addition, retroperitoneal tumors are associated with high morbidity rates, so one may discuss the possibility of either performing (or not) a multivisceral resection of a retroperitoneal tumor in case of a suspected sarcoma that might be invading adjacent structures.

However, incision in the pseudocapsule of a retroperitoneal sarcoma has been associated with poor survival. Therefore, in this case, a resection with a curative purpose aiming at a better loco-regional control of the disease was performed. Moreover, in complicated myelolipomas with hemorrhage and an organizing hematoma, the etiologic radiological diagnosis may be even more difficult and inaccurate as observed in this patient. Thus, the preoperative radiological assessment itself may be impaired in these situations and a retroperitoneal malignant neoplasm cannot be totally ruled out by this method[5,7,8]. That is why, based on oncological principles, multivisceral resections, such as in this case, may be necessary. Pain and intestinal obstruction may occur together in complicated giant myelolipomas and are accurate indications for surgical resection, as described herein.

Although the management of adrenal myelolipoma may be "wait and watch" in the beginning, in order not to cause therapeutic failures, a definitive radiological diagnosis must be made, and fine needle aspiration biopsy might be performed[3,7]. On the other hand, there is consensus in literature that these tumors must be resected in cases of complications, such as pain, compression of adjacent structures (mass effect), hemorrhage, rupture and infection[3]. Some authors stated that tumors larger than 4 cm must be resected, even if asymptomatic, since they pose a higher risk of spontaneous rupture[11,12].

Although uncommon, myelolipoma should be considered in the expanding lesions of adrenal or retroperitoneal origin. These tumors have excellent prognosis as they are benign and can be treated in a conservative manner in uncomplicated, asymptomatic or small size (less than 4 cm) tumors. Nevertheless, for safe management, the preoperative radiological diagnosis must be precise.

REFERENCES

1. Wong KW, Lee IP, Sun WH. Case report: rupture and growth of adrenal myelolipoma in two patients. Br J Radiol. 1996;69(825):873-5.

2. 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(1):129-32.

3. Meaglia JP, Schmidt JD. Natural history of an adrenal myelolipoma. J Urol. 1992;147(4):1089-90.

4. Goldman HB, Howard RC, Patterson AL. Spontaneous retroperitoneal hemorrhage from a giant adrenal myelolipoma. J Urol. 1996;155(2):639.

5. Pérez Martínez J, Llamas F, López Rubio E, Serrano A, Salinas Sánchez A, Ruiz Mondéjar R, et al. [Giant adrenal myelolipoma: hypertension, renal failure and spontaneous rupture]. Nefrologia. 2006;26(1):132-5. Spanish.

6. Pareja Megia MJ, Barrero Candau R, Medina Perez M, Valero Puerta J. Mielolipoma adrenal gigante. Arch Esp Urol. 2005;58(4):363-5.

7. Suárez Artacho G, Rodríguez Muñoz J, Gómez Bravo MA, Campoy Martínez P, López Bernal F. [Adrenal gland hematoma due to rupture of myelolipoma. A case report]. Actas Urol Esp. 2004;28(10):785-8. Spanish.

8. Ares Valdés Y. [Adrenal myelolipoma. Case report and bibliographic review]. Arch Esp Urol. 2006;59(1):71-3. Spanish.

9. Kawashima A, Sandler CM, Ernst RD, Takahashi N, Roubidoux MA, Goldman SM, et al. Imaging of nontraumatic hemorrhage of the adrenal gland. Radiographics. 1999;19(4):949-63.

10. Vara Castrodeza A, Madrigal Rubiales B, Veiga González M, Cuesta Varela F, Sales Fernández C, Fresno Forcelledo M. [Adrenal gland myelolipoma: radiological view]. Arch Esp Urol. 2001;54(1):73-6. Spanish.

11. El Mejjad A, Fekak H, Dakir M, Sarf I, Manni A, Meziane F. [Giant adrenal myelolipoma]. Prog Urol. 2004;14(1):81-4. French.

12. Wagnerová H, Lazúrová I, Bober J, Sokol L, Zachar M. Adrenal myelolipoma. 6 cases and a review of the literature. Neoplasma. 2004;51(4): 300-5.