DOI: https://doi.org/10.2298/SARH180621090A
UDC: 616.688-006-079.4

CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Spermatic cord angiomyolipoma misdiagnosed as inguinoscrotal hernia

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SUMMARY

Introduction Angiomyolipomas represent neoplasms of mesenchymal origin, made up of abnormal thick-walled blood vessels, smooth spindle muscle cells, and mature adipose cells. The most common site of origin are kidneys, and other localizations are extremely rare. We represent a case of a spermatic cord angiomyolipoma misdiagnosed as incarcerated inguinoscrotal hernia, and to our prudence this is second described case of an angiomyolipoma localized in the spermatic cord.

Case outline We present a case of a 63-year-old man presented with high fever and difficulty in walking due to pain and swelling in the right groin. According to the clinical examination and laboratory tests, presumptive diagnosis was incarcerated inguinoscrotal hernia, so the patient was immediately operated on. The exploration of the inguinal canal showed a timorous mass, 9 × 9 cm in size, with the origin from the spermatic cord, so radical inguinal orchiectomy was performed with the removal of the tumor mass. Histopathological and immunohistochemistry examination suggested angiomyolipoma of the spermatic cord. The postoperative course was uneventful.

Conclusion Although rare, an angiomyolipoma of the spermatic cord must be included in the differential diagnosis of scrotal masses. Also, we advocate additional diagnostic procedures (ultrasound or computed tomography) for every inguinoscrotal mass before undertaking surgery, since a variety of different causes can be found. After definitive angiomyolipoma diagnosis is obtained, further investigation is needed, especially brain computed tomography due to possible tuberous sclerosis coexistence.

Keywords: angiomyolipoma; extrarenal neoplasm; spermatic cord tumor

INTRODUCTION

Angiomyolipomas (AML) are neoplasms of mesenchymal origin, made up of abnormal thick-walled blood vessels, smooth spindle muscle cells, and mature adipose cells, probably derived from perivascular epithelioid cells [1]. The kidneys are the most common site of origin, and AML represent the most usual benign resectable kidney tumors [2]. Other localizations are exceedingly uncommon [3].

We present a rare case of a spermatic cord AML misdiagnosed as incarcerated inguinoscrotal hernia.

CASE REPORT

We present a case of a 63-year-old man presented to our general surgery emergency department with high fever and difficulty in walking due to pain in the right groin for a few days. The clinical examination revealed irreducible inguinoscrotal swelling on the right side and tenderness to touch, measuring about 6 × 4 cm with overlying erythematous skin changes. The patient noticed an inguinal mass about one year ago, but without any other symptoms. Laboratory tests showed leukocytosis (14,200 leukocytosis/μL). Presumptive diagnosis was of an incarcerated inguinoscrotal hernia. The patient's medical history was significant for type 2 diabetes mellitus and arterial hypertension. The patient was immediately taken to the operating room. Surgery started with a right inguinal approach. The exploration of the inguinal canal was performed. The spermatic cord was dissected and showed a tumorous mass that was 9 × 9 cm in size. The urologist was immediately called to the operating room, and radical inguinal orchiectomy was performed, with high ligation of the right cord and removal of the entire tumor mass. Posterior inguinal wall weakness was observed and the defect was repaired using the Bassini technique. Postoperative histopathological examination (Figure 1) and immunohistochemistry (Figure 2) suggested an angiomyolipoma of the spermatic cord. After surgery, whole-body computed tomography (CT) was performed in order to exclude other tumors, and all findings were normal. Also, serum tumor markers (lactate dehydrogenase, alpha-fetoprotein, and beta human chorionic gonadotropin) were within normal limits. The postoperative course was uneventful and the patient was discharged on the fifth postoperative day.

Received • Примљено: June 21, 2018
Revised • Ревизија: July 11, 2019
Accepted • Прихваћено: August 2, 2019
Online first: August 14, 2019

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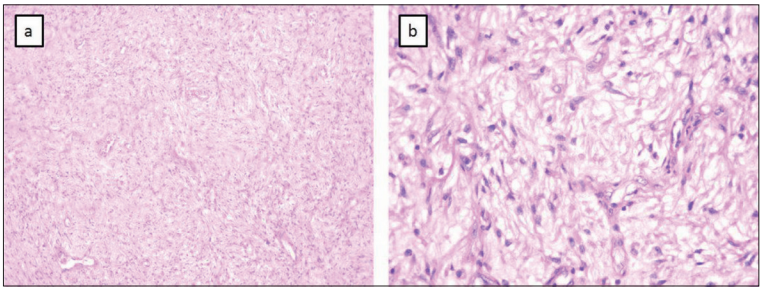

Figure 1. H&E staining, magnification: (a) ×40, (b) ×400

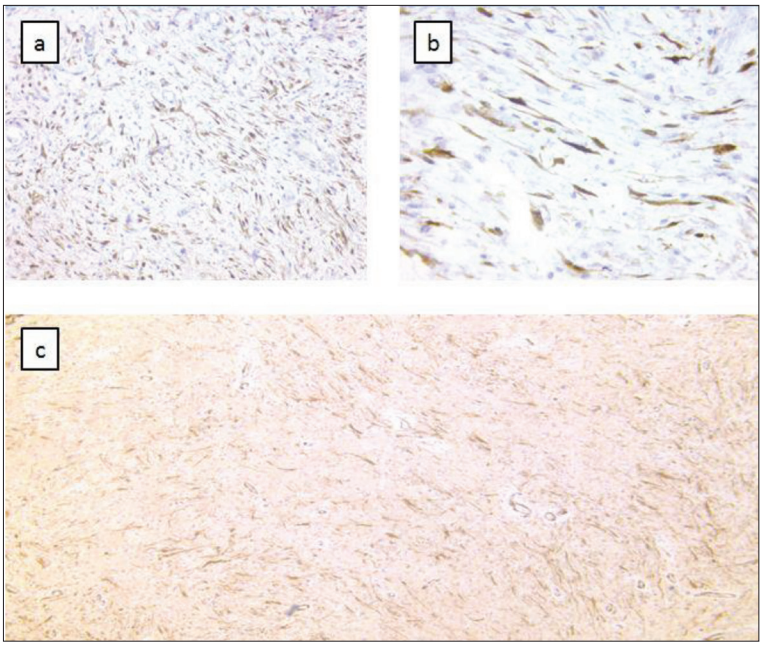

Figure 2. Immunochemical staining: (a) CDK4 ×400; (b) p16 ×400; (c) alpha-SMA ×100

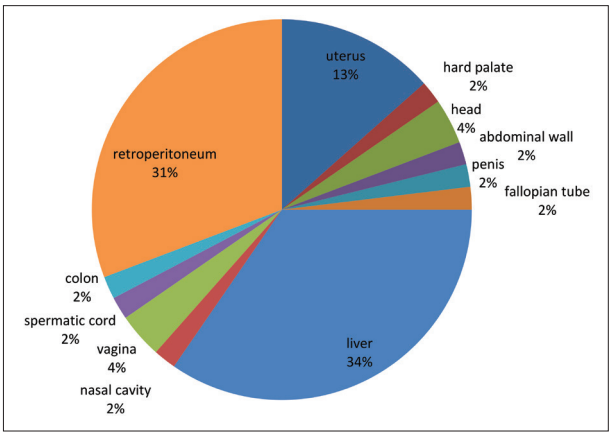

Figure 3. Extrarenal localization of AML according to Minja et al. [3]

DISCUSSION

AML are rare benign neoplasms that usually occur in the kidneys, and account for 1% of all renal masses. Extrarenal localization of AML is exceedingly uncommon [4]. After kidneys, the liver and the skin are the next most common sites of AML origin. Minja et al. [3] conducted a literature review in 2012 and found only 52 cases of AML with extrarenal localization. Summarized findings of the Minja et al. [3] review are given in Figure 3. The liver localization was the most common (18 cases), followed by the uterus (seven cases), and retroperitoneum (four cases). Out of all presented cases, only one patient had a spermatic cord AML (3), i.e. in 1989 this case was described by Castillenti and Bertin [5]. According to our knowledge, our case is the second AML located in the spermatic cord.

AML are previously thought to be hamartomatous lesions. It is now known that AML arise from the perivascular epithelioid cells (PEC) and belong to a family of tumors named "PEComas." These tumors show phenotypic (smooth muscle, epithelioid, and lipid-rich histology), and immunohistochemical coexpression of myogenic (smooth muscle actin) and melanocytic (HMB-45, melan A, microphthalmia transcription factor, tyrosinase) modulation. In other words, pathohistological findings of AML typically tend to have triphasic features: myoid spindle cells, islands of fat tissue, and dysmorphic blood vessels that have thick walls and do not have elastic lamina [6, 7]. In AML immunohistochemical studies show positive staining for various markers such as MART1/Melan-A, muscle-specific actin (HHF35), HBM45, calponin, NKI-C3. Also, AML tend to be negative for renin and keratin [6, 8]. In our case, histopathological and immunohistochemical examination revealed typical AML. On the other hand, scrotal region tumors are mostly testicular germ cell tumors, which are associated with an increase in serum tumor markers. Since in our case the tumor grew from the spermatic cord, before obtaining definitive diagnosis by histopathology, our assumption was that germ cell tumor is the probable diagnosis. However, the tumor markers were within the normal range. Since AML represents a rare entity which can be clinically present in many guises, a pathohistological and especially an immunohistochemical examination are the gold standard in the definitive diagnosis of AML.

Extrarenal retroperitoneal AML may present in a variety of ways, such as incidental radiological finding, abdominal pain, loin and back pain, felling of fullness in the abdomen, diffuse pain and bleeding, hematuria, vomiting, constipation, weight loss, abdominal mass [6]. Pain in the right groin, high fever, and a local finding indicative of incarcerated inguinoscrotal hernia were present in our patient. Thus, AML of extrarenal localization can appear in many possible scenarios and diagnosis prior to obtaining a pathohistological finding is almost impossible.

According to Minja et al. [3], CT scan is the commonest used radiologic technique in the investigation of AML. Also, brain CT is recommended in patients with AMS, because about 40% of them have CT features of tuberous sclerosis [6]. High proportion of fat content, which is

found in the majority of AML, produces a characteristic pattern on CT scan. Variants of AML with poor fat content (about 5% of cases) or cystic AML, as well as atypical (epithelioid or monophasic) AML may present diagnostic challenges on radiological studies, and in such cases, it is very difficult to distinguish an AML from a renal cell carcinoma. This is also case in the patients with tuberous sclerosis coexistence, where up to one third of patients do not demonstrate macroscopic fat on CT. Also, calcifications are rare in AML [9]. MR imaging is excellent at evaluating lesions containing fat tissue, and two main sequences are used. First, fat saturated sequences demonstrate high signal intensity on non-fat-saturated sequences and loss of signal following saturation of fat. The second technique is to use in-phase and out-of-phase imaging which generates "India ink artifact" at the interface between fat and non-fat components. This can occur either at the interface between the AML and surrounding tissue or between fat and other components of the tumor [10]. Also, AML are hypervascular lesions demonstrating often characteristic findings on angiography images: a sharply marginated hypervascular lesion with a dense arterial network, with tortuous vessels (arterial phase), spiral "onion peel" appearance of peripheral vessels (venous phase), absent arteriovenous shunting, and intraumoral aneurysms [11]. In our case preoperative diagnosis was not performed because of the patient's general state and pernicious local finding, so patient was immediately taken to operating room. However, postoperatively we performed whole-body CT including brain CT scan, and all findings were normal.

In 1982, Friis and Hjortrup [12] reported the first extrarenal AML in a 22-year-old female patient who presented with abdominal pain. Exploratory laparotomy revealed retroperitoneal peripancreatic AML weighing 11 kg. They concluded that AML should always be treated surgically because preoperative diagnosis is uncertain. In our case, presumptive diagnosis was of an incarcerated inguinoscrotal hernia. Guided by the experience of Friss and Hjortrup [12], as well as the fact that AML has a tendency of recurrence if the entire mass is not removed, if differential diagnosis includes AML, surgery remains the best treatment option. Since AML of extrarenal origin are rare, guidelines for renal AML must be taken with caution. Renal AML found incidentally (small, asymptomatic) usually require no therapy, although a follow-up is recommended to assess possible growth. AML larger than 4 cm in size or symptomatic AML can be electively embolized and/or resected with a partial nephrectomy. AML presented with hemorrhage often require emergency embolization or resection as a life-saving procedure. mTOR inhibitors are shown to decrease the size of AML; however, larger studies are needed [13]. After reviewing the literature and taking into account our experience, we strongly advocate surgery in patients with a presumed diagnosis of AML of extrarenal origin.

This case report was approved by the institutional ethics committee, and written consent was obtained from the patient for the publication of this case report and any accompanying images.

Conflict of interest: None declared.

REFERENCES

1. Flum AS, Hamoui N, Said MA, Yang XJ, Casalino DD, McGuire BB, et al. Update on the Diagnosis and Management of Renal Angiomyolipoma. J Urol. 2016; 195(4 Pt 1):834–46.
2. Kwazneski Ii D, Merrill M, Young J, Sell H Jr. Angiomyolipoma and Malignant PEComa: Discussion of Two Rare Adrenal Tumors. Case Rep Oncol Med. 2016; 2016:5204092.
3. Minja EJ, Pellerin M, Saviano N, Chamberlain RS. Retroperitoneal extrarenal angiomyolipomas: an evidence-based approach to a rare clinical entity. Case Rep Nephrol. 2012; 2012:374107.
4. Tseng CA, Pan YS, Su YC, Wu DC, Jan CM, Wang WM. Extrarenal retroperitoneal angiomyolipoma: case report and review of the literature. Abdom Imaging. 2004; 29(6):721–3.
5. Castillenti TA, Bertin AP. Angiomyolipoma of the spermatic cord: case report and literature review. J Urol. 1989; 142(5):1308–9.
6. Venyo AK. A Review of the Literature on Extrarenal Retroperitoneal Angiomyolipoma. Int J Surg Oncol. 2016; 2016:6347136.
7. Tsutsumi M, Yamauchi A, Tsukamoto S, Ishikawa S. A case of angiomyolipoma presenting as a huge retroperitoneal mass. Int J Urol. 2001; 8(8):470–1.
8. Ziadie M. Kidney tumor – cysts, children, adult benign Benign (usually) adult tumors Angiomyolipoma. Pathology Outlines. com. 2015. http://www.pathologyoutlines.com/topic/kidneytumorangiomyolipoma.html.
9. Shetty AS, Sipe AL, Zulfiqar M, Tsai R, Raptis DA, Raptis CA, et al. In-Phase and Opposed-Phase Imaging: Applications of Chemical Shift and Magnetic Susceptibility in the Chest and Abdomen. 2019; 39(1):115–35.
10. Israel GM, Hindman N, Hecht E, Krinsky G. The use of opposed-phase chemical shift MRI in the diagnosis of renal angiomyolipomas. AJR Am J Roentgenol. 2005; 184(6):1868–72.
11. Logue LG, Acker RE, Sienko AE. Best cases from the AFIP: angiomyolipomas in tuberous sclerosis. Radiographics. 2003; 23(1):241–6.
12. Friis J, Hjortrup A. Extrarenal angiomyolipoma: diagnosis and management. The J Urol. 1982; 127(3):528–9.
13. Maclean DF, Sultana R, Radwan R, McKnight L, Khastgir J. Is the follow-up of small renal angiomyolipomas a necessary precaution? Clin Radiol. 2014; 69(8):822–6.

Ангиомиолипом сперматичне врпце погрешно дијагностикован као ингвиноскротална кила

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САЖЕТАК

Увод Ангиомиолипоми представљају туморе мезенхимног порекла састављене од абнормалних крвних судова дебелих зидова, глатких мишићних ћелија и зрелих масних ћелија. Најчешће настају у бубрезима, а друге локализације су изузетно ретке. Приказујемо случај ангиомиолипома сперматичне врпце погрешно дијагностикованог као уклештена ингвиноскротална кила; по нашем сазнању, ово је други описани случај ангиомиолипома ове локализације.

Приказ болесника Приказујемо случај мушкарца старог 63 године који се јавио у нашу установу због повишене телесне температуре и отежаног хода услед бола и отока у десној препони. Према клиничком налазу и лабораторијским претрагама претпостављена дијагноза била је уклештена ингвиноскротална кила и болесник је хитно оперисан. Експлорацијом ингвиналног канала уочена је туморска маса димензија 9 × 9 *cm* порекла сперматичне врпце, због чега је учињена радикална ингвинална орхиектомија са уклањањем целе туморске масе. Хистопатолошки и имунохистохемијски налаз су указали на ангиомиолипом сперматичне врпце. Постоперативни ток је протекао уредно.

Закључак Иако ретки, ангиомиолипоми сперматичне врпце морају бити укључени у диференцијалну дијанозу тумефакција у ингвиноскроталној регији. Такође сматрамо да би обављање допунских дијагностичких процедура (ултразвучни преглед или компјутеризована томографија) било значајно пре оперативног захвата, јер се могу наћи различити патолошки супстрати. После постављања дефинитивне дијагнозе ангиомиолипома потребна је даља дијагностика, а посебно компјутеризована томографија мозга, због могуће коегзистенције ангиомиолипома и туберозне склерозе.

Кључне речи: ангиомиолипоми; екстраренални тумори; тумор сперматичне врпце