

CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Spermatic cord angiomyolipoma misdiagnosed as inguinoscrotal hernia

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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 tumorous 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 leukocytes/ μ L). 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.

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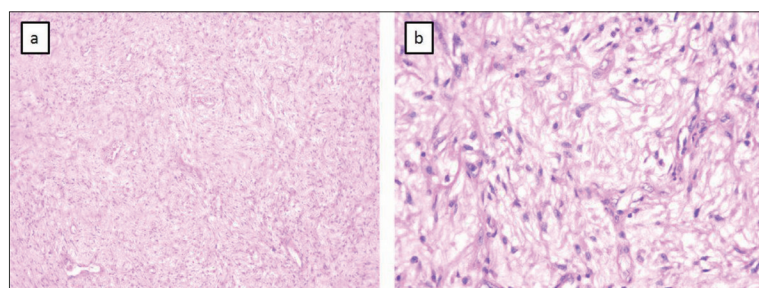


Figure 1. H&E staining, magnification: (a) $\times 40$, (b) $\times 400$

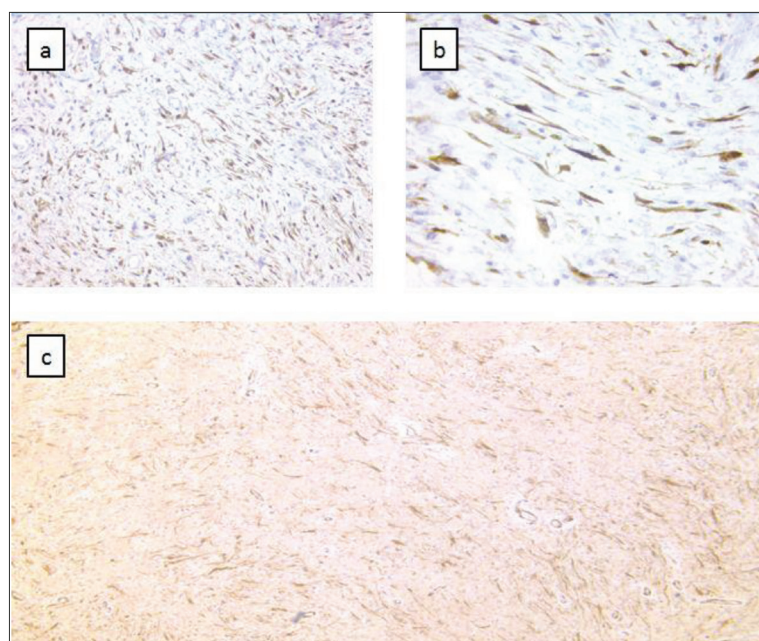


Figure 2. Immunohistochemical staining: (a) CDK4 $\times 400$; (b) p16 $\times 400$; (c) alpha-SMA $\times 100$

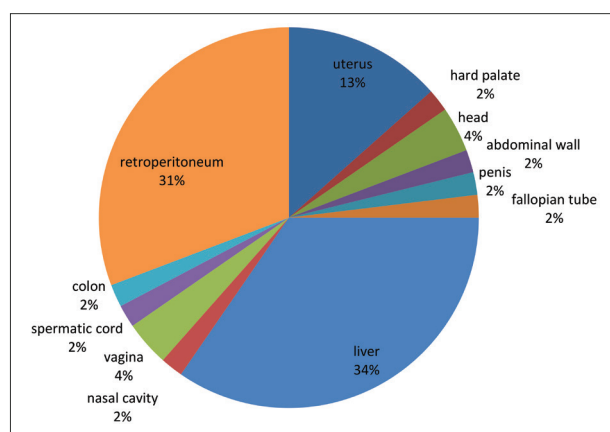


Figure 3. Extrarenal localization of AML according to Minja et al. [3]

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, feeling 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 tuberos 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 intra-tumoral 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.

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Conflict of interest: None declared.

Ангиомиолипом сперматичне врпце погрешно дијагностикован као ингвиноскротална кила

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САЖЕТАК

Увод Ангиомиолиполи представљају туморе мезенхимног порекла састављене од абнормалних крвних судова дебелих зидова, глатких мишићних ћелија и зрих масних ћелија. Најчешће настају у бубрезима, а друге локализације су изузетно ретке. Приказујемо случај ангиомиолиполима сперматичне врпце погрешно дијагностикованог као укљештена ингвиноскротална кила; по нашем сазнању, ово је други описани случај ангиомиолиполима 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 укљештена 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, ангиомиолиполи сперматичне врпце морају бити укључени у диференцијалну дијагнозу 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 ангиомиолиполима потребна је даља дијагностика, а посебно компјутеризована томографија мозга, због могуће коегзистенције ангиомиолиполима и туберозне склерозе.

Кључне речи: ангиомиолиполи; екстраренални тумори; тумор сперматичне врпце