Extrarenal retroperitoneal angiomyolipoma with unusual evolution

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Abstract
The aim of this paper is to present the case of an extrarenal retroperitoneal angiomyolipoma with unusual evolution, due to the herniation through the inguinal canal, determining an extraperitoneal hernia. A ureteral duplication and associated hydronephrosis contributed to the peculiarity of the case. The case was operated (en block tumor and right kidney removal), the postoperative evolution being favorable at seven years after the surgery.

Keywords: primary retroperitoneal angiomyolipoma, ureteric obstruction, ureteral duplication, extraperitoneal hernia.

Introduction
Angiomyolipomas are rare mesenchymal tumors, developed mainly from the renal parenchyma, but can also occur in other locations (liver, nasal cavity, oral cavity, colon, lung, skin, adrenal glands and bladder) [1–4]. The retroperitoneum, as a primary site, is the third most common location [5, 6], less than 60 cases being reported until now in medical literature [7], most of them as single case reports. Although most of the cases were benign, some malignant cases were also reported; in fact, in many cases, due to the large size of the tumor, the presence of the malignancy is difficult to be excluded and represents an important problem of the differential diagnosis [5–9].

A retroperitoneal angiomyolipoma exteriorized through the right inguinal canal along with a right ureteral duplicity, both ureters being involved in the tumoral mass, is also an exceptional clinical circumstance that has not been published yet in the medical literature.

Aim
The aim of this paper is to present the diagnostic and treatment difficulties induced by the development of an asymptomatic, rare retroperitoneal tumor, associated with a ureteral duplicity, and determining an inguinal hernia.

Case report
Patient D.G., male, 65-year-old, was admitted in our surgical clinic in September 2006, with an irreducible tumor in the right inguinofemoral region, developed for over four years, with no subjective complaints. Associated pathology included grade 3 obesity and grade 2 arterial hypertension.

Preoperative diagnosis was right irreducible inguinal hernia; usual biological tests, EKG and thoracic X-ray were normal, other imaging investigations being considered irrelevant in case of an irreducible hernia.

Intraoperative in the inguinal canal a lipomatous mass was found, with continuity in the right scrotum and also contiguous through the deep inguinal orifice with a retroperitoneal tumor. The presence of two ureters in the right inguinal canal and the absence of a peritoneal sac were also recorded (Figure 1).

The lipomatous mass was removed en-block with the right testicle, but the section of the tumor was limited at the level of the deep inguinal orifice; both ureters were reduced in the retroperitoneal space.

Postoperatively an abdominal CT revealed a 15/25 cm inhomogeneous retroperitoneal tumor, developed from the inferior renal pole to the inguinal arch; the tumor presented fat tissue densities, intratumoral hemorrhage areas, calcification, and sclerosis. The tumor pushed superiorly and laterally the right kidney and includes two ureteric ducts; a thickening of the pararenal fascia, the presence of the cleavage plane with the inferior vena cava and abdominal wall, and the right kidney hydro-nephrosis were also noticed. An intravenous urography showed a grade two right hydronephrosis, and a single right ureter amputated at the ilium crest level (Figure 2).

A second operation using a median laparotomy was employed three weeks later, revealing a 15/25 cm lipomatous tumor, developed caudally from the inferior pole of the right kidney. The tumor encased two ureteric ducts:
one of the ureters (the one highlighted by urography) originated from the right renal pelvis and the other is developed from superior renal pole with no relation to renal pelvis (Figure 3). This explains why the urography revealed only one ureter, the other being nonfunctional. The tumor presented with smooth, elastic areas, specific to a lipoma, but it becomes hardened in the distal half; at this level, the ureteral dissection is impossible. The impossibility of the ureteral dissection from the tumoral mass becomes clear after the entire tumor mobilization, both ureteral ducts being embedded in a process of sclerosis and calcification determining ureteral obstruction; consequently, the tumor was removed en-bloc with both right ureters and the right kidney.

The resection specimen, including the right kidney, consisted in a 15/25 cm mass with lipomatous appearance and hemorrhagic areas alternating with areas of necrosis and hardened areas with sclerosis and calcifications; both right ureters were stuck tight in the fibrosclerosis process (Figure 4).

The postoperative evolution was uneventful, the patient being alive and with no signs of recurrence seven years after the surgery.

The pathology revealed a mature adipose tissue mixed with smooth muscle tissue proliferation, with rare giant cells, surrounding the medium caliber blood vessels. The microscopic aspect was highly conclusive with the angiomyolipoma diagnosis (Figure 5).

**Discussion**

The primitive retroperitoneal extrarenal site of angio-

myolipomas is rarely encountered, less than 60 cases being reported [7] since their initially description by Friis & Hjortrup in 1982 [6].

Due to their development in the retroperitoneal space, these tumors may evolve for a long time without showing any clinical symptoms, even if they reach a significant size [8–12]. Patients suffering from retroperitoneal angiomyo-

lipomas, similar with other primary retroperitoneal tumors, may experience non-specific abdominal pain, enlargement of the abdomen, and less commonly, symptoms caused by the ureteric obstruction [10–13]. Retroperitoneal hemorrhage and hemorrhagic shock (especially in angiomatous subtype) may
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represent severe complications [7, 9, 13–15]. However, in many cases, angiomyolipomas are incidentalomas and they are almost impossible to be diagnosed in case of obesity [7].

However, for a complete diagnosis the value of laparotomy and histopathological examination are undeniable, since the malignant retroperitoneal tumors are four times more frequent than benign ones [10–12].

In our case, although the right ureteric obstruction has caused hydronephrosis, there was a lack of symptoms, possible due to the chronic progressive obstruction, the only clinical manifestation being the right inguinal hernia.

Figure 5 – Microscopic aspects of the angiomyolipoma: (A) Mature adipose tissue mixed with smooth muscular tissue proliferation and rare giant cells, around medium caliber blood vessels (Hematoxylin–Eosin – HE – staining, ×100); (B) Mature adipose tissue mixed with smooth muscular tissue proliferation and vacuolar cells disposed between the adipose cells (HE staining, ×100).

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Ureteral hernia was reported in approximately 140 cases in the literature. There are two types of ureteral hernias: paraperitoneal (80% of cases – the ureter herniates alongside a peritoneal sac) and extraperitoneal (20% of cases – the peritoneal sac is absent) [16, 17]. In paraperitoneal hernias, the ureter herniates due to peritoneal adhesions and in some cases abdominal viscera may form one wall of the hernia sac [16, 17]; by contrast, in the extraperitoneal form herniation of the ureter occurs without a peritoneal sac, the ureter being pushed (“got in head”) by a retroperitoneal tumor, as in our case.

It is important to notice that extraperitoneal hernia may associate congenital renal or ureteric malformations (in our case right ureteral duplicity) [16, 17].

If the only clinical manifestation is the hernia, it is important to recognize and identify intraoperatively the ureter, in order to avoid its damage [16, 17], but this may be difficult in obese patients with no signs or symptoms of urinary involvement and if additional diagnostic imaging tests were not performed.

In most cases, hydronephrosis is caused by angulation of the ureter or its compression by the hernia contents [16, 17]; in our case, chronic obstruction was produced by the progressive sclerosis and calcification around the ureter, secondary to intratumoral hemorrhage and necrosis.

Imaging investigations are crucial both for diagnosis and for guiding surgical attitude; the usual imaging modalities include ultrasound imaging, CT scan and MRI. Urography is required in most case after CT scan and MRI. Along with the exclusion of the renal origin of the tumor, these imaging modalities highlight the relations between the tumor, kidney and ureter and, very important, it certifies the contralateral kidney functionality, essential in case of nephrectomy [10, 11]. Also, the abdominal CT may help in differentiating between the angiomyolipomas and other retroperitoneal tumors (especially liposarcomas), in spite of lack of pathognomonic signs [18].

If possible, the tumor may be resected without an associated nephrectomy, but usually (30% of cases in the study of Tseng et al.) [20], the removal of the kidney en-bloc with the tumor is necessary [7], due to the impossible ureteral dissection (the ureter is encased in the tumoral mass) and the associated right hydronephrosis. Normal function of the contralateral kidney is mandatory, otherwise a modality of solving the case without nephrectomy must be employed: reimplantation of the proximal ureter in the contralateral ureter or in the digestive tract, often the reimplantation into the bladder being impossible due to the small length of the ureteral stump; kidney transplantation in the right iliac fossa and proximal ureter implantation into the bladder represents another modality of solving these cases [10].
Pathological diagnosis is difficult and must distinguish between various types of retroperitoneal tumors, especially liposarcomas [4, 5, 8], but also other retroperitoneal (atyypical lipomas) or renal and adrenal tumors.

Histologically, angiomyolipomas are described as having three tissue components: convoluted thick walled blood vessels, smooth muscle cells, and mature adipose tissue; the proportion of the tissue components is variable, one or the other may prevail, thus three subtypes were described: angiomatous, myomatous, or lipomatous type [5, 8]. Their origin appears to be in the perivascular epithelioid cells, therefore they are HMB-45-positive; the immunohistochemical diagnosis, although not mandatory for the diagnosis, helps in differentiating from retroperitoneal liposarcomas or myolipomas [3–5, 7].

Regarding their behavior, most of the angiomyolipomas are benign, and very rarely they can degenerate malignant, becoming recurrent or metastasizing to the liver, bone and mediastinum [4, 7, 21]. These cases require aggressive surgery that would give patients the maximum chances of long-term survival. Often, vascular resections with vascular reconstructions (vena cava, liver, bone and mediastinum [4, 7, 21]. These cases require aggressive surgery that would give patients the maximum chances of long-term survival. Often, vascular resections with vascular reconstructions (vena cava, common iliac pedicle) and multiorgan resections may be necessary for the diagnosis, helps in differentiating from retroperitoneal liposarcomas or myolipomas [3–5, 7].

Extrarenal retroperitoneal angiomyolipomas are rare mesenchymal tumors, the retroperitoneum being an unusual location. The diagnosis is difficult due to the non-specific symptoms; sometimes the angiomyolipomas may be part of an extraperitoneal hernia and cause hydronephrosis. Renal and ureteric malformations may be associated. The treatment consists in surgical resection, complete removal of the tumor, with or without associated nephrectomy, being the main objective. Although the tumoral behavior is usually benign, the possibility of malignancy must be taken into consideration.

Conflict of interests
The authors declare that they have no conflict of interests.

Author contribution
All authors have contributed equally to the present work.

References

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