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Case report: Management of A Giant Retro-Peritoneal Leiomyoma

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Abstract

Retro-peritoneum is a rarely reported location of leiyomyoma. Symptoms are not specific and the tumor can be confused with a renal tumor in tomography. Surgery must consider the tumor's vascularization, that may provide from the aorta or the renal artey. A pre-operative embolization may facilitate surgery and reduce bleeding in voluminous tumors. Follow-up is mandatory to detect reccurrence and the eventual malignant transformation.

Keywords: Interrogation, Tumor Size, Vascularization, Hypervascularized, Doppler.

Introduction

Leiomyomas are benign tumors wich are mainly observed in adult women. Retro-peritoneal location is rare ; almost a hundred of cases have been reported. As retro-peritoneal leiomyomas are pauci-symptomatic and the tumor size at diagnosis is relatively large, surgical management is challenging. Authors report a case of a 22 centimeter retro-peritoneal leiomyoma in a 52 years old woman. A pre-operative embolization was performed two days before surgery. Clinical, therapeutic, and evolutive aspects of this rare entity are discussed.

Case Report

A 52 years old woman consulted surgical emergency with a suddent left flank pain with a palpable tender mass. She hasn't any past medical history. Interrogation revealed a chronic constipation. She was apyretic, and didn't present vomiting or a recent disturbance in bowel motility. There were no urinary of gynecological disorders. The palpable mass was located in the left flank and the iliac fossa, with a posterior lumbar extension. Biological exploration was normal, there wasn't particularly a biological inflammatory syndrom. The CA-125 and the carcinoembryonic antigen were in their normal rates. A voluminous left-sided heterogeneous tissular abdomino-pelvic mass was described in abdominal ultrasonography, with a mass effect on the left kidney. The mass was non-vascularized on the Doppler. On the abdomino-pelvic CT-scan, the mass measured 16*17*22 centimeter. It was well-limited, heterogeneous and hypervascularized. The vascularization seemed to arise exclusively from the renal artery. The mass effect concerned the left kidney and its hilum, the left ureter, the left colon and the inferior mesenteric vascular axis. The renal vein and the inferior vena cava were permeable (Figure 1).Angiography confirmed the CT findings, as the tumor vessels emerged exclusively from the renal artery. An embolization of the renal artery was performed, using a trans-femoral approach, with a consequent total disappearance of the tumor vascularization (Figure 2). After two days of opioid analgesia, the patient woas operated. A midline incision

allowed an exploration of the abdominal cavity. The operative exploration was similar to the scan findings. The left kidney was pushed to the median line. The mass seemed to be firmly adherent to the kidney and its hilum; the dissection plane wasn't obvious. The tumor had smaller size after embolization. Its manipulation was'nt particularly haemorrhagic. The first step was the identification of the ureter and the clamping of the renal artey and vein. A thourough dissection of the tumor from adjacent structures was performed, allowing a one-piece resection of the mass with the left kidney (Figure 3). Post operative course was uneventful. The patient was discharged at the fourth post-operative day.Histological examination concluded to a benign mesenchymal proliferation, involving regular smooth muscle cells proliferating in a hyper-vascularized stroma. Focal hyalinized spots were observed. Mitotic index wasn't high. There weren't any fat cells and necrotic lesions. The tumoral proliferation didn't concern the renal parenchyma. Immunohistochemical study was negatif to HMB45, CD 117, Dog 1, PS 100 and CD 34. Ki-67 proliferation index was also negative. Tumor cells were postive to desmine, caldemone and actine, confirming the diagnosis of retro-peritoneal leiomyoma. The kidney and its capsule were free from any histological lesions. Follow-up was regular for two years. Clincal examination was normal. Renal function was conserved. Abdominal CT-scan ruled out any local or distant recurrence.

Discussion

Leiomyomas are developped from smooth muscle cells, and are rarely located in the retro-peritoneum. To the best of our knowledge, slightly more than a hundred of cases have been described in the english litterature, fourty of them contain sufficient data for study [1]. Retro-peritoneal smooth muscle cell tumors are often diagnosed fortuitously, symtoms are related to adjecent structures compression and the mean size at diagnosis is 12 centimeter [2]. 80% of them are malignant, represented by leiomyosarcoma [3]. Pathogenesis of retro-peritoneal leiomyoma is still unclear. The role of gonadotropic hormones seems to be influent. The uterine location, the most decribed one, mainly occurs during the genital acticity period in women. Up to 40% of retro-peritoneal cases are associated to synchronous or operated uterine myomas [1]. This therory is particularly plausible, as only nine cases were reported in men [4], and the density of oestrogen and mainly of pregesteron receptors is remarkably important [5]. Stutterecker et al. evoked the eventuality of the development of embryonic vessels remnant musculature. This idea is supported by the previous cases describing pulmonary and heart locations [6]. Diagnosis is rarely established pre-operatively [1]. Symptoms are not specific, and are related to the mass effect of the tumors [7]. It's important to mention that most of the retro-peritoneal leiomyomas are independent from the uterus, in the pelvic floor [1]. They develop in the upper part of the retro-peritoneum in 30% of the cases [8]. In four patients, leiomyoma was in the anterior retro-peritoneum, in the Retzuis or in uterus adnexa [5,9].The ultrasound and the CT scan aspects aren't evocative. The tumor is usually decribed as an voluminous, heterogen and hypervascularized mass [4]. The pre-operative imagery is mainly interesting to define precisely the anatomical limits of the tumor. MRI may be interesting to differenciate leiomyoma from leiomyosarcoma, like in uterine myomas [10]. Tumor biomarkers, mainly the CA-125 and the carcinoembryonic antigen, may be elevated in huge tumors. Once elevation is documented pre-operatively, it can be interesting in the further follow-up [11]. Surgery is the only curative option. Open surgery was the option in most of the previously reported retro-peritonal

leiomyomas. Laparoscopic approach was decribed in only two cases [1]. The difficulty in laparoscopic or robot-assisted approaches are mainly related to the relatively large size at diagnosis, and the adherences to adjacent structures [12]. The principle is to ensure a one-piece excision of the tumor, and to conserve the integrity of the surrounding organs and large vessels [13]. Once benignity is confirmed histologically, recurrence and malignant transformation are extremely rare [1]. Nevetheless, the few reported cases justify a thourough clinical and radiological follow-up. Other therapeutic options can be discussed on a individual basis. LHRH-analogue have been proposed, despite the lack of effisciency and the recurrence of evolutive symptoms on the treatment discontinuation [14]. Arterial embolization was also described, as a pre-operative option in huge retro-peritoneal tumors. It's mainly useful to reduce operative haemorrhagic incidents, and to slightly reduce the tumor size. Surgery should be perfomed within two days, and analgesia must be strengthened due to the consequent intense pain [5]. Even if some incidents have already been described, like arterial dissection or systemic embolization, tumor arteries embolization is usually a safe technique to facilitate surgical approach in voluminous or symptomatic retro-peitoneal tumors [15]. In the case we report, the tumor was totally vascularized from the renal artery, while most of the described embolization procedures of the retro-peritoneal tumors concerned lumbar arteries and secondarily the renal artey.

Conlusions

Retro-peritoneal leiyomyoma is a challenging diangostic and therapueitc situation. Histological diagnosis is usually post-operative, as radiological features aren't conclusive of benignity. Surgery must consider vascularization of the tumor, as it may provide from the renal artery in some cases. A thourough clinical and radiological follow-up is required due to the potential malignant transformation.

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Figures

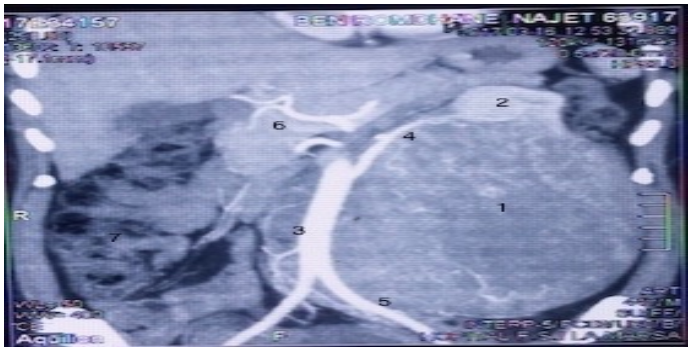

Figure 1 : A retro-peritoneal tumor with a mass effect on the left kidney, the aorta and the bowel. 1 : Tumor, 2 Left kidney, 3 : Aorta, 4 : Left kdney's artery, 5 : Ilaiac artery, 6 : Pancreas.

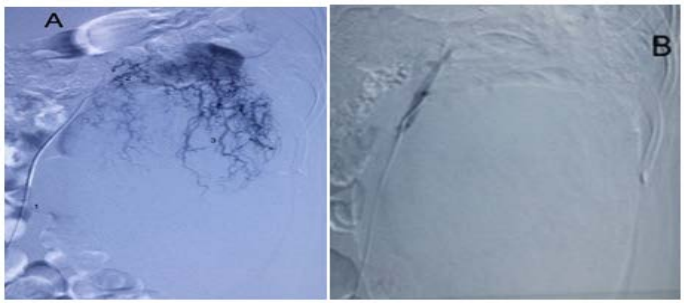

Figure 2 : Figure 2 A : An abundant and anarchic vascularization of the tumor, providing exclusively from the left renal artery.

Figure 2 B : Embolization of the renal artery, with a complete disappearence of the tumor vascularization.

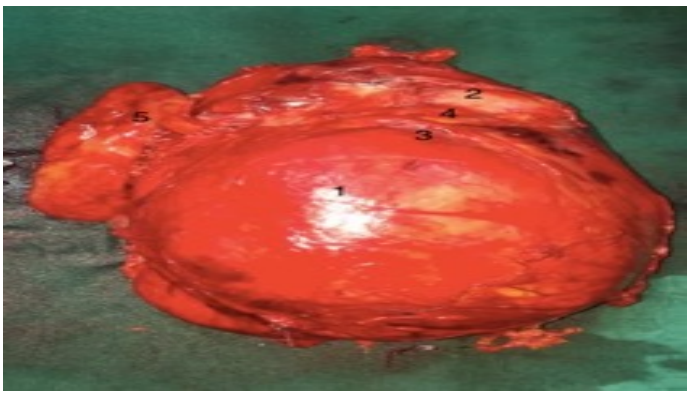

Figure 3 : Operative specimen. 1 : The tumor, 2 : Left kidney, 3 : Tumor capsule, 4 : The left kidney's capsule, 5 : The kidney's hilum.