



Giant primary retroperitoneal myxoid leiomyoma: a case report

Gigantski primarni retroperitonealni miksoidni lejomiom

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Abstract

Introduction. Leiomyomas are benign smooth muscle tumors that usually arise from the uterus. **Case report.** We present a patient with a 6-month history of vague abdominal discomfort, occasional nausea, vomiting and urinary incontinence. On examination, there was an extremely large firm unpainful palpable abdominal mass. Laboratory investigation revealed mild leukocytosis and blood creatinine elevation. Abdominopelvic ultrasonography and computed tomography revealed a massive well bordered, encapsulated intraabdominal tumor, extending from the pelvis to epigastrium and almost completely fulfilling the pelvic and abdominal cavity. At laparotomy, tumor arising from the retroperitoneum was excised *in toto*. Histopathological examination disclosed that the tumor was composed mainly of smooth muscle cells and very rare fibrous connective tissue elements with myxomatous alteration and with no mitotic activity. The negative results of numerous additional parameters analyzed (pancytokeratin, epithelial membrane antigen, S100 protein, CD68, CD34, desmin, aktin) ruled out different origin of a tumor. One year after resection the patient had no complaints and no radiological evidence of tumor recurrence. **Conclusion.** Considering current limitations in radiological diagnosis, *in toto* resection of these tumors is necessary to rule out malignancy.

Key words:

leiomyoma; myxoma; retroperitoneal space; digestive system surgical procedures.

Apstrakt

Uvod. Lejomiomi su benigni tumori glatkih mišića koji se najčešće javljaju u uterusu. **Prikaz bolesnika.** U radu smo prikazali bolesnicu sa neodređenim bolovima u trbuhu, povremenim mučninama, povraćanjem i urinarnom inkontinencijom koji su trajali prethodnih šest meseci. Fizikalnim pregledom utvrđeno je prisustvo ekstremno velikog, tvrdog, bezbolnog abdominalnog tumefakta. Laboratorijska ispitivanja otkrila su blagu leukocitozu i povišenu vrednost kreatinina u serumu. Ultrazvučni pregled i kompjuterizovana tomografija abdomena i karlice utvrdili su postojanje masivnog, jasno ograničenog, inkapsuliranog intraabdominalnog tumora koji se prostirao od karlice do epigastrijuma, ispunjavajući skoro u potpunosti karličnu i trbušnu duplju. Nakon laparotomije, tumor poreklom iz retroperitoneuma odstranjen je u celini. Histopatološkom analizom utvrđeno je da se sastojao uglavnom iz glatkomišićnih ćelija i veoma retkih elemenata fibroznog vezivnog tkiva sa miksomatoznom alteracijom i bez mitotske aktivnosti. Negativni rezultati brojnih, dodatno analiziranih parametara (pancytokeratin, epitelijalni membranski antigen, S100 protein, CD68, CD34, dezmin, aktin) isključili su drugačije poreklo tumora. Godinu dana nakon resekcije bolesnica nije imala tegobe niti radiološke pokazatelje recidiva tumora. **Zaključak.** S obzirom na ograničenja radiološke dijagnostike, *in toto* resekcija lejomioma je neophodna da bi se isključio malignitet.

Ključne reči:

lejomiom; miksom; retroperitonealni prostor; hirurgija digestivnog sistema, procedure.

Introduction

Leiomyomas are benign smooth muscle tumors that usually arise from the uterus being the most common benign tumors in women¹. However, primary leiomyoma of the retroperitoneum without the co-existence of uterine leiomyoma or disseminated disease is very rare². We presented a case of

barely symptomatic extremely large primary retroperitoneal leiomyoma (RL).

Case report

A 68-year-old multiparous woman was referred to our department with the 6-month history of vague abdominal discom-

fort, occasional nausea, vomiting and urinary incontinence. The previous medical history including gynecological was negative. Her general condition and vital signs were normal. On examination, there was an extremely large firm unpainful palpable abdominal mass with smooth surface, extending from the suprapubic region to the epigastrium and completely fulfilling the abdominal cavity. Laboratory investigation revealed mild leukocytosis (11 000/dL) and blood creatinine elevation (173.0 $\mu\text{mol/L}$). All other parameters including tumor markers (CEA, CA19-9, CA125 and AFP) were normal. Systemic examination, including gynecological, did not reveal any abnormalities. Chest and abdominal radiography examination findings were within normal limits. Abdominopelvic ultrasonography (USG) revealed a large 33 \times 28 cm in diameter cystic tumor, located ventrally and fulfilling most of the abdominal cavity with mild hydronephrosis on the right side. Abdominal computed tomography (CT) confirmed USG finding revealing a massive well bordered, encapsulated intraabdominal tumor 40.8 \times 36.9 \times 19.1 cm in size, extending from the pelvis to epigastrium and almost completely fulfilling the pelvic and abdominal cavity and comprising adjacent organs, mainly the right kidney, aorta, urinary bladder and intestines (Figure 1). There were no signs of ascites and genital pathology.

feeding arteries and hemostasis was achieved without difficulties. The abdomen was closed in layers. The resected tumor weighed 6.3 kg (Figure 2). The postoperative course

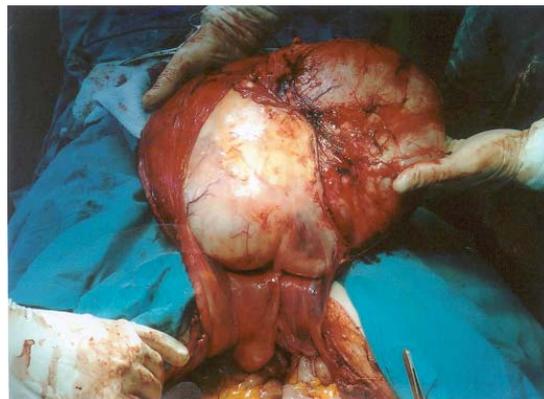


Fig. 2 – Resection and gross appearance of a giant retroperitoneal leiomyoma.

was uneventful. Histopathological examination disclosed that the tumor was RL composed mainly of smooth muscle cells and very rare fibrous connective tissue elements with myxomatous alteration and with no mitotic activity (Figure 3).



Fig. 1 – Computed tomography (CT) presentation of a giant retroperitoneal leiomyoma.

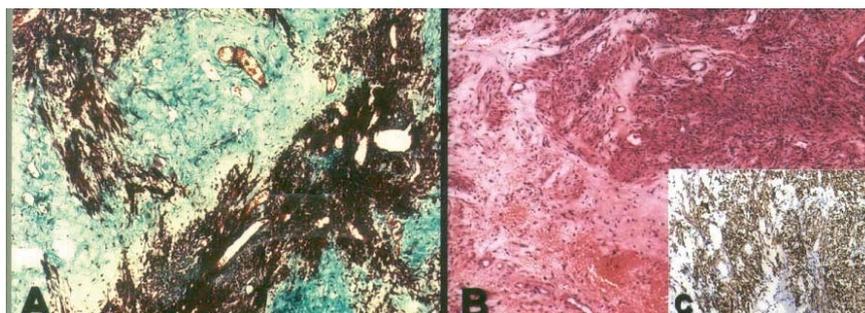


Fig. 3 – Myxoid leiomyoma

[A) Masson's trichrome stain, 10 \times ; B) Haematoxylin & eosin stain, 10 \times ; C) Desmin stain, 20 \times].

At laparotomy, a giant encapsulated firm tumor with smooth surface was found, arising from the right retroperitoneal region at the level of paraaortic (interaortocaval) area and extending throughout the whole abdominopelvic cavity. The remainder of the peritoneal cavity was normal. There was no connection of the tumor to the genital tract. The tumor was easily excised *in toto* after ligating the main

The negative results of numerous additional parameters analyzed (pancytokeratin, epithelial membrane antigen, S100 protein, CD68, CD34, desmin, aktin) ruled out a different origin of the tumor. One year after resection the patient had no complaints and no radiological evidence of RL recurrence.

Discussion

Etiology and pathogenesis of leiomyomas are still poorly understood. Since these tumors probably arise from smooth muscle cells, including those in blood vessel walls, they can originate wherever smooth muscle cells exist. The most common site of their location is the uterine corpus during the fourth and fifth decade of life. It has been estimated that leiomyomas affect 25% of all women during their reproductive life¹. However, leiomyomas occasionally occur in atypical extrauterine locations like genitourinary tract (vulva, ovaries, urethra, urinary bladder, kidney), lung, rectum etc.³⁻⁵ and may show unusual growth patterns: benign metastasizing leiomyoma, disseminated peritoneal leiomyomatosis, intravenous leiomyomatosis, parasitic leiomyoma etc.⁶. The growth of uterine leiomyomas is most probably hormonally (estrogen) dependent since their frequency is increased after menarche, they enlarge during pregnancy and their regression occur after the menopause¹. Also, it has been demonstrated that a number of different growth factors may be involved in the pathogenesis of leiomyomas: epidermal growth factor, basic fibroblast growth factor, heparin-binding growth factor, transforming growth factor beta, granulocyte-macrophage colony-stimulating factor and insulin-like growth factors^{7,8}.

Although there have been reports on various atypical localizations for leiomyomas, their growth in the retroperitoneum is extremely rare. The etiopathogenesis of primary RL is not fully elucidated. It could be related to uterine leiomyomas since more than 40% of patients with RL have a concurrent uterine leiomyoma or a history of hysterectomy for leiomyoma⁹. Zaitoon¹⁰ suggested that large uterine leiomyomas adhere to surrounding structures, acquire an auxiliary blood supply and detach from the uterus – “parasitic” leiomyomas. Also, Stutterecker et al.¹¹ suggested that RL may arise from embryonal remnants of Müllerian or Wolffian tubes or local blood vessels musculature.

RL may grow very long and become considerably large and still remain asymptomatic. They may be detected incidentally during the examination for other reasons or autopsy¹. The most frequent clinical feature of these tumors is palpation of abdominal/pelvic mass present in 90% of patients⁹. Rarely, they grow to cause clinically significant symptoms: abdominal discomfort, fatigue, weight loss and pain radiating to the back. Sometimes, they cause compressive effect on renal collecting system producing hydronephrosis¹⁰, like in the presented case or pressure and displace important retroperitoneal and vascular structures. Since retroperitoneal smooth muscle tumors are more often malignant than benign, prompt and accurate preoperative radiological assessment is necessary. Ultrasonography examination provides good ini-

tial orientation for retroperitoneal masses. CT and especially magnetic resonance imaging (MRI) are most useful screening tools in evaluating and distinguishing the exact nature of the tumor and its relationship with adjacent organs and vascular structures. However, no radiological diagnostic modality appears highly sensitive or specific in ruling out malignancy and differential diagnosis on the basis of radiological finding alone is difficult¹². Therefore, the definitive diagnosis of RL requires histopathological examination of the tumor. Sampling of the retroperitoneal mass under USG or CT-guidance preoperatively may allow microscopic examination, although the results may be unreliable due to small histologic specimen. Hence, the final determination of the tumor's nature is to be accomplished with a complete examination of resected specimen. Histologically, the distinction of benign leiomyoma and malignant leiomyosarcoma (especially low grade) may also be difficult. The histopathological parameters used for differential diagnosis include gross tumor size, the presence of nuclear atypia, pleomorphism and necrosis and the mitotic activity as the most useful guide to prognosis². On light microscopy, leiomyoma consists of monomorphic spindle cells arranged in interweaving fascicles which are separated by variable amounts of hyalinized collagen. Smooth muscle cells are elongated with eosinophilic cytoplasm and uniform, cigar-shaped nuclei. Usually, there is no cytologic atypia or necrosis and mitotic index is less than 5/10 in high-power fields. In addition, immunohistochemical staining with estrogen, progesterone receptors, desmin, calponin, h-caldesmon, CD10, CD34, c-kit, ki-67 and p53 may be helpful in differential diagnosis of leiomyoma from leiomyosarcoma¹³. The differential diagnosis of RL includes leiomyosarcomas, nonovarian teratomas, paragangliomas, neurilemmomas-schwannomas, angiomyxomas, hemangiopericytomas, pheochromocytomas, liposarcomas, lymphomas and metastatic tumors¹⁴.

Conclusion

A complete surgical excision is the only curative treatment for retroperitoneal smooth muscle tumors, regardless their benign or malignant nature. Considering current limitations in radiological diagnosis, *in toto* resection of these tumors is necessary to rule out malignancy. However, RL sometimes may be massive, adherent to important adjacent structures and covered with large vessels mimicking malignancy. Therefore, resection of the tumor might be incomplete. Nevertheless, the surgeon should excise the tumor as completely as possible, especially in symptomatic patients. Also, providing a necessary experience, laparoscopic treatment of these tumors is possible.

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