



## Giant primary retroperitoneal leiomyoma - a case report

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**Abstract :** Leiomyomas are benign smooth muscle tumors that usually arise from the uterus. Isolated retroperitoneal leiomyoma is very rare. It usually coexists with uterine leiomyoma or part of a disseminated disease. We report a 41-year-old multiparous women presented with abdominal discomfort and left Loin pain for six months. Abdominopelvic Sonography , computed tomography and MRI showed a large cystic mass that filled the left retroperitoneum around the ureter and pushing the kidney upwards. With the preoperative diagnosis of a malignant tumor, an exploratory laparotomy was planned. Intraoperatively, a cystic mass originated from the retroperitoneum surrounding the ureter was observed. it was completely resected and sent for histopathological examination, which came as retroperitoneal leiomyoma, confirmed with immunohistochemical staining.

**Keyword :** leiomyoma, retroperitoneum, immunohistochemical staining

### Introduction

Leiomyomas are benign smooth muscle tumors that usually arise from the uterus. Leiomyomas are the most common benign tumors in women(1). However, primary leiomyoma of the retroperitoneum without the co-existence of uterine leiomyoma or disseminated disease is very rare.

### Case report

A 41-year-old multiparous woman was referred to our department with 6-month history of vague abdominal discomfort and continuous dullaching left loin pain. She gave a past history of transabdominal hysterectomy with bilateral salphingoopherectomy done 2 years ago for dysfunctional uterine bleeding. Her general condition and vital signs were normal. On examination, there was midline scar and no mass was palpable. Laboratory investigations revealed mild anemia and all other basic investigations and renal parameters were normal. Chest and abdominal radiography examination findings were within normal limits. Abdominopelvic ultrasonography (USG) revealed a large 14X11 cm cystic tumor, located in the left retroperitoneum around the ureter and pushing the kidney upwards with mild hydroureteronephrosis .Contrast enhanced computed tomography (CECT) and MRI (figure 1 and 2)

confirmed USG finding revealing a massive , well bordered, encapsulated retroperitoneal tumor 14X11 cm in size, extending from the renal pelvis into bony pelvis and almost completely encircling the left ureter. There was no ascites.

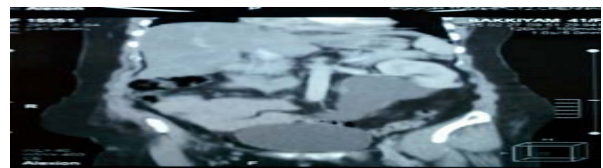


Figure 1 - CECT KUB

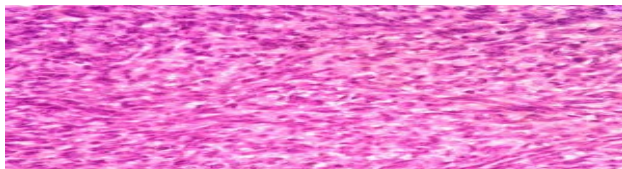


Figure 2 - MRI KUB

At exploration through left flank approach, a giant encapsulated tumor with smooth surface was found, arising from the left retroperitoneal region , extending from the renal pelvis into bony pelvis and almost completely encircling the left ureter. There was no connection of the tumor to the genital tract. The tumor was excised *in toto* after dissecting the tumor from the left ureter and ligating the main feeding arteries and complete hemostasis was achieved . The resected tumor weighed 1.3 kg . The postoperative course was uneventful. Histopathological examination disclosed that the tumor was spindle cell neoplasm composed mainly of smooth muscle cells on immuno histochemical staining revealed retroperitoneal leiomyoma with strong positivity for SMA (CD-117, S-100 & CD-10 negative) (figure 3 and 4).



**Figure 3 - Resected specimen**



**Figure 4 - Microscopic appearance on immunohistochemical staining**

#### Discussion

Etiopathogenesis of leiomyomas are still poorly understood. These tumors arise from smooth muscle cells and can originate wherever smooth muscle cells exist. The most common site is the uterus during the fourth and fifth decade of life. It has been estimated that leiomyomas affect 25% of all women during their reproductive life (1). However, leiomyomas occasionally occur in atypical extrauterine locations like genitourinary tract (vulva, ovaries, urethra, urinary bladder, kidney), lung, rectum etc.(2,3) and may show unusual growth patterns. 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 menopause (1). 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. 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 retroperitoneal leiomyomas have a concurrent uterine leiomyoma or a history of hysterectomy for leiomyoma. Retroperitoneal leiomyomas may grow very long and become considerably large and still remain asymptomatic. They may be detected incidentally during the examination for other reasons on autopsy (1). The most frequent clinical feature of these tumors is palpation of abdominal/pelvic mass present in 90% of patients. 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 hydronephrosis, 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 initial 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 difficult(4). 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 prognosis(2). 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 leiomyosarcoma(5). The differential diagnosis of RL includes leiomyosarcomas, nonovarian teratomas, paragangliomas, neurilemmomas-schwannomas, angioyxomas, hemangiopericytomas, pheochromocytomas, liposarcomas, lymphomas and metastatic tumors(6).

#### 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, retroperitoneal leiomyomas 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, in expert hands, laparoscopic treatment of these tumors is possible.

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