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Case Reports

# Retroperitoneal benign leiomyoma is a challenging diagnosis in gynecology: report of two cases

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**Summary**

*Background:* Leiomyomas represent rare primary retroperitoneal tumors. Preoperative diagnosis is challenging due to the low incidence, non-specific clinical presentation, and atypical radiological features. *Case Report:* The authors describe two cases of retroperitoneal tumors that resulted to be rare retroperitoneal leiomyomas. The degenerating process of the tumors created atypical conditions that explained the difficult diagnosis at imaging and the absence of compression syndrome. MRI has not been useful in this case to highlight preoperative diagnosis. Diagnosis was only made postoperatively based on final anatomopathological results. Complete surgical excision was achieved in both cases and patients are free of recurrence after a very long period of follow up. *Conclusion:* The very low incidence and non-specific clinical presentation of retroperitoneal leiomyomas make the diagnosis challenging. Preoperative imaging can be misleading mostly when tissue degeneration exists. The treatment consists in complete surgical excision. Histopathological analysis is essential to diagnosis. *Conclusions:* Retroperitoneal leiomyomas are rare retroperitoneal tumors. Diagnosis is challenging due to the low incidence, poor clinical presentation, and atypical radiologic features. Tissue degeneration is common in retroperitoneal leiomyomas. This partially explains the absence of compression syndrome and atypical features on radiology.

*Key words:* Leiomyosarcoma; MRI; Retroperitoneal leiomyoma.

## Introduction

Leiomyomas of the retroperitoneum are rare smooth muscle tumors. Preoperative diagnosis and differential diagnosis with more common retroperitoneal tumors like sarcoma may be challenging because of this low incidence, non-specific clinical presentation, and atypical radiological features. The authors present two cases of retroperitoneal tumors that resulted to be benign retroperitoneal leiomyoma.

## Case Report

*Case 1*

A 55-year-old G3P3 menopausal patient consulted for vaginal bleeding. She had no relevant medical history. Physical examination was normal. Pap smear and endometrial biopsy were normal.

Ultrasound showed abnormal limits of the uterus. MRI revealed a right lateral pelvic mass of 14×10×8.5 cm in size (Figure 1A). The differential diagnosis was: gastrointestinal stromal tumor (GIST) versus leiomyoma or sarcoma. Blood test including CA 125 was normal. CT scan showed a pelvic mass, with cystadenoma or cystadenocarcinoma as differential diagnosis. A CT scanner-guided biopsy showed some benign fibrosclerous components. Explorative laparoscopy showed a large and soft lateral retroperitoneal pelvic mass without ascites. Laparotomy was decided.

The mass was resected from the broad ligament and dissected from iliac vessel, ureter, sigmoid, and sacral bone. The extemporaneous histopathological examination ruled out malignancy. Concomitant hysterectomy and adnexectomy were performed. No other explanation for vaginal bleeding was found. Final anatomo-pathological results concluded to be a retroperitoneal leiomyoma of 773 grams showing signs of myxoid transformation (Figures 1B and 1C).

The mass was attached to the right ovary and tube, without infiltration of the tissues. Mitotic activity was low. There was no necrosis. Some cellular atypia was found, which was explained by the presence of myxoid degeneration rather than by malignancy. Immunohistochemistry showed positivity for desmin, actin, and caldesmon. S100 was negative. Ki-67 was low. No concomitant uterine leiomyoma were observed. After 59 months of follow up, the patient was free of recurrence, unfortunately after she died of pulmonary disease.

*Case 2*

A 50-year-old, G2P2, non-menopausal women consulted for removing her IUD. The patient complained of non-specific abdominal pain during the past two years. There was no relevant medical history or actual treatment. Physical examination showed a pelvic mass with extrinsic compression of the rectal ampulla. Ultrasound showed a para-uterine heterogeneous mass. CT scan revealed a pelvic mass of 26×20×9 cm in size (Figures 2A and 2B) with leiomyoma versus ovarian solid cyst (cystadenoma, cystadenocarcinoma) as differential diagnosis.

Blood test was normal except for CA 125 which was two-fold increased (58 mUI/ml). Laparotomy showed a retroperitoneal large mass which was dissected from pelvic structures. Complete surgical excision was achieved. Concomitant hysterectomy and adnexectomy was done. The extemporaneous anatomopathological examination was not conclusive. Final anatomopathological results confirmed a 2,534-gram benign retroperitoneal leiomyoma showing hyaline de-

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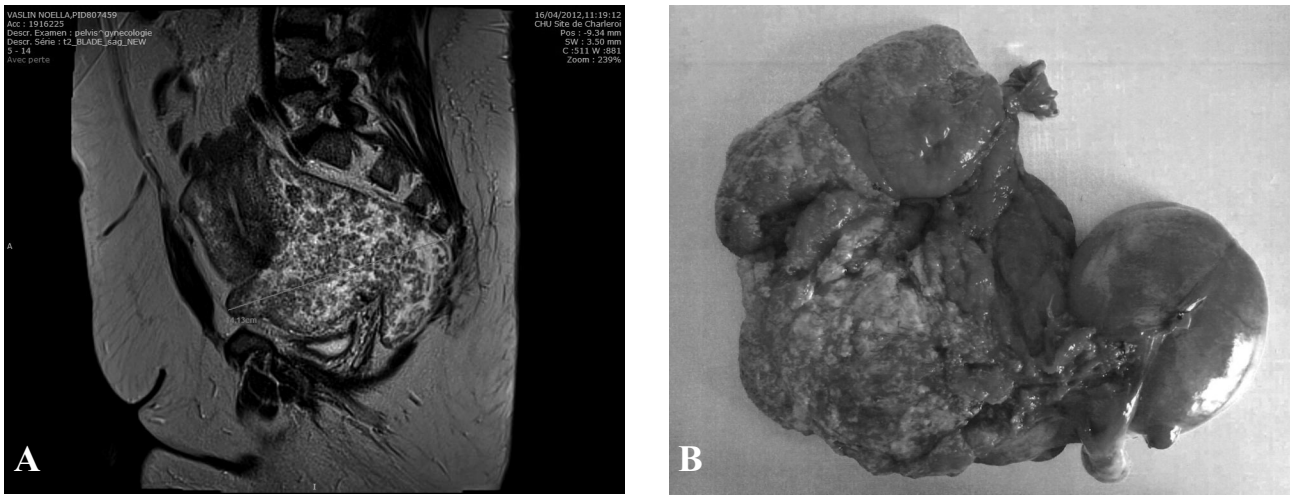
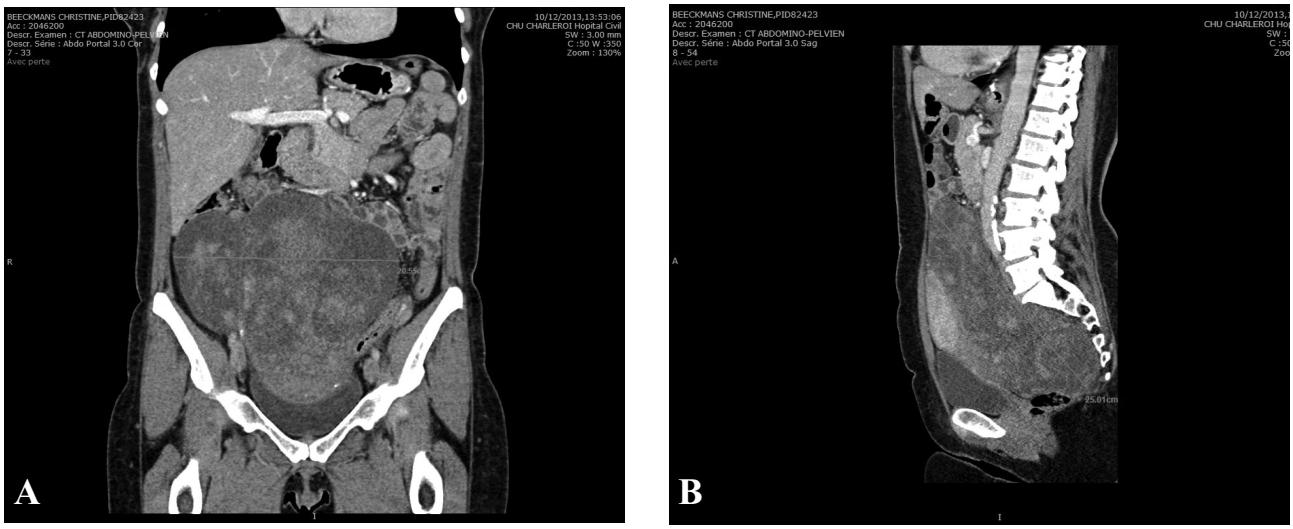



Figure 1. — A) Sagittal T2- weighted MR image shows a polylobed mass located in the pelvis. B) Macroscopic anatomopathological picture of R.L. (myxoid degenerating).



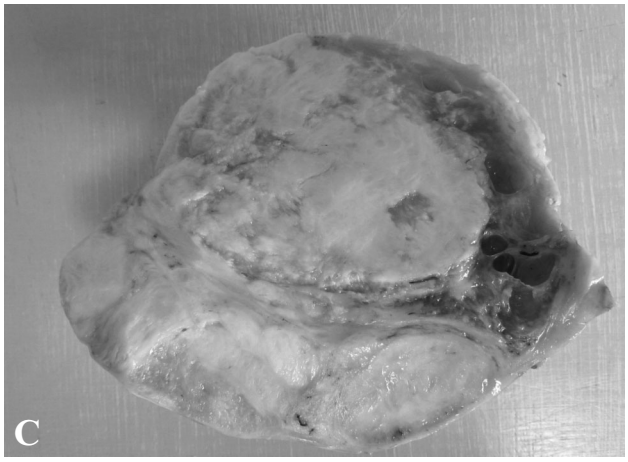




Figure 2. — A) Coronal view of contrast CT revealing the voluminous tumor. B) Sagittal view of contrast CT reveals the tumor pushing the uterus and bladder forward. C) Macroscopic view of retroperitoneal mass.

generation (Figure 2C). Several small uterine leiomyoma were also found. The mass was attached to the uterus at the level of the uterine isthmus, without infiltration of the uterine wall. Immunohistochemistry showed positivity for desmin, smooth muscle actin, caldesmon, a low Ki-67, and negativity for S100 and C-kit. There was no cellular atypia and no necrosis. Mitotic index was low. After 42 months of follow-up, the patient was free of recurrence.

## Discussion

Leiomyomas of the retroperitoneum are smooth muscle tumors [1, 2]. Among primary retroperitoneal tumors, the incidence of leiomyoma is extremely low: 0.5-1.2% [1, 3-5]. The majority of them are malignant sarcomas [1, 2]. The authors report two cases of retroperitoneal tumors that resulted to be benign retroperitoneal leiomyomas.

Diagnosis of retroperitoneal leiomyomas represents a challenge related to the non-specific clinical presentation. Most of the cases are asymptomatic. Symptoms described in litera-

ture are: abdominal, pelvic or back pain, dyspareunia, urinary or gastrointestinal compressive complaints, weight loss or gain and fatigue. An abdominal mass is sometimes palpated [1, 2, 4-7]. The presented cases confirmed that retroperitoneal leiomyomas are associated with poor clinical presentation. Despite the voluminous masses described here (773 and 2,534 grams), patients presented only non-specific symptoms and compression of retroperitoneal structures was not observed. A possible explanation might be the myxoid and hyaline degeneration, observed in both tumors, making the tumor tissue soft and less likely to compress surrounding structures.

An elevated CA 125 was found in association with retroperitoneal leiomyomas in some cases. Peritoneal stimulation in large retroperitoneal leiomyomas is a hypothesized explanation in literature [1]. The second case, with higher tumor volume, included an elevated CA 125 level.

With regards to imaging, radiological presentation is also a challenge. Ultrasonography, CT scan, and MRI can assist in the diagnosis of retroperitoneal leiomyomas [1, 2, 3, 6]. Of these, MRI seems to be the most reliable technique for diagnosis and differential diagnosis with leiomyosarcoma. Leiomyomas typically show a muscle-specific signal and homogenous enhancement with MRI [6, 8]. Unfortunately, degeneration process can produce non-specific findings. Radiologic findings were non-specific in the present cases. In the present authors' opinion, the degeneration process may explain the atypical imaging and then perioperative diagnosis was not been possible.

Pathogenesis of retroperitoneal leiomyomas remains unclear. Literature reports several hypotheses: they may originate from embryonal remnants, from local vessel musculature, parasitic leiomyomata (that detach from the uterus), iatrogenic or metaplastic leiomyoma [1, 4, 6-8]. Although 40% has concurrent or a personal history of uterine leiomyoma, tumors are seldom connected to the uterus [1, 4, 6]. The second case showed the presence of concurrent uterine leiomyoma. Although the mass was attached to the uterus, no infiltration of the uterine wall was observed.

Anatomopathological analysis is the gold standard for diagnosis and exclusion of malignancy, although low-grade sarcoma may be challenging to distinguish. Preoperative biopsy or extemporaneous anatomopathology may contribute but definitive diagnosis is often only made postoperatively [1]. Both cases presented here failed to be diagnosed on biopsy and extemporaneous analysis.

Retroperitoneal leiomyomas present as well-differentiated smooth muscle cell tumors [1, 3, 4]. They show no nuclear atypia, no coagulative necrosis, and have a low mitotic index [1, 3, 4]. Immunohistochemically, they are positive for alpha-SMA and desmin and negative for CD34 and CD117 [1, 3, 4]. They are mostly hormone-sensitive [1, 3, 4, 6]. Up to 50% of cases present fibroid degeneration, mostly hyaline, but also myxoid and cystic degenerations are described [1, 4].

Anatomopathological and immunohistochemical results in the present cases were similar to the cases described in literature, besides some cellular atypia in the second case, explained by the degeneration process. The frequent fibroid degeneration is confirmed in both cases.

Treatment of choice is complete surgical excision, which is mostly curative [1-3, 5, 7, 8].

Retroperitoneal leiomyomas has a favorable prognosis with very low local recurrence rate [1, 4], as was confirmed in the present two cases. Moreover, the authors published two cases with very long term follow ups (59 and 42 months).

The incidence of sarcomatous transformation of uterine leiomyoma is extremely low (0.04-0.7%) [1]. Furthermore, long term follow up is mandatory to exclude recurrences in cases of initial misdiagnosis.

## Conclusion

The authors report two new cases of rare retroperitoneal leiomyoma that were in accordance with the previous published cases. Preoperative diagnosis was not made, highlighting this challenging condition. The two tumors were fully surgically resected and the very long follow up did indicate any recurrence. Further studies are needed to confirm that long term follow up is probably not required. Facing primary retroperitoneal tumors with poor clinical manifestation and with no compression syndrome of surrounding structures, benign retroperitoneal leiomyoma must be considered.

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