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RARE CASE OF RETROPERITONEAL LEIOMYOMA: CASE REPORT

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Abstract: *Retroperitoneal leiomyoma is a rare pathology, given the high prevalence of malignant tumours of the retroperitoneal space and the absence of surgical history of uterine fibromatosis. We present the case of a 47-year-old patient, with no significant medical history, accusing pelvic pain, in which the CT scan showed voluminous pelvic tumour and uterine fibroma. Histological examination of the tumour showed retroperitoneal leiomyoma.*

INTRODUCTION

Leiomyomas are a frequent gynecological pathology, being the main cause for surgical interventions on the uterus. Although in the vast majority of cases they are attached to the uterus, it is possible for them to occur with unusual growth patterns, especially in women of reproductive age, such as the retroperitoneum. This makes them susceptible to be interpreted as other gynaecological pathologies, such as ovarian malignancies. In most cases, tumours originated from the smooth muscles of this region are malignant, namely leiomyosarcomas.

Diagnosis of such pathologies can prove to be difficult, as tumour markers have low specificity, and imaging cannot always fully state the type of tumour. As preoperative biopsies are not always an available option, the only certain diagnostic is often histological examination of the resected specimen.

Clinically, the symptoms are most of the time non-specific, beginning to manifest when the tumour reaches large sizes and causes compression on adjacent organs, increasing the risk and difficulty of the surgical procedure. Also, the pelvic retroperitoneal placement of these tumours can pose a challenge regarding surgical approach and technique.

For these reasons, pelvic retroperitoneal leiomyomas can cause difficulties regarding diagnosis, treatment and post-operative management.

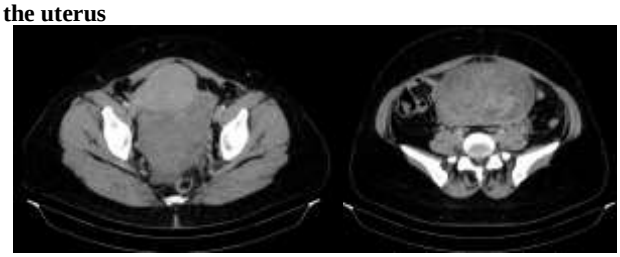
CASE REPORT

We present the case of a 47-year-old patient, with no significant medical history, accusing menometrorrhagia, with pelvic and abdominal pain.

A CT scan of the abdomen and pelvis was performed, showing a voluminous pelvic tumour of 10/15/20 cm, with well-defined margins, in contact with the posterior side of the uterus, as well as an intramural uterine fibroid (figure no. 1).

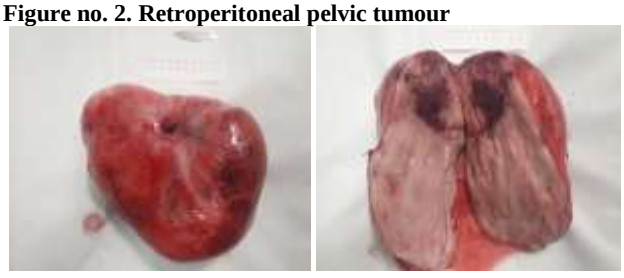
Blood tests show a high level of CA-125, respectively 111 U/ml, with no additional pathological findings. Surgery is performed, with the excision of a voluminous retroperitoneal pelvic tumour (figure no. 2) along with total hysterectomy for the uterine fibroid.

Figure no. 1. Pelvic-abdominal tumour of 10/15/20cm, with well-defined margins, in contact with the posterior side of the uterus



Histopathological examination of the tumour has shown benign mesenchymal proliferation suggestive for leiomyoma composed of smooth muscle fibres, with no nuclear atypia, associating vascular congestion and areas of necrosis. Postoperative evolution was favourable, with no pathological findings in follow-up examinations.

Figure no. 2. Retroperitoneal pelvic tumour



DISCUSSIONS

Retroperitoneal tumours can have varying origins (pancreatic, renal, lymphoproliferative), both malignant (primary or metastatic) and benign, leiomyomas being a rare occurrence. Statistically, pelvic retroperitoneal tumours of malignant origin are 4 times more frequent than benign ones,

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sarcomas representing one third of them. For benign tumours, neurogenic origin is the most frequent (neurofibromas, schwannomas), along with fibroids and lipomas.(1,2) Fibromas not connected to the uterus are a rare occurrence, as most frequent extrauterine locations are found in the genitourinary tract (ovaries, vulva, urethra and urinary bladder). Of these rare occurrences, even more unusual locations have been cited, such as disseminated peritoneal leiomyomatosis, parasitic leiomyomas, intravenous leiomyomatosis, and retroperitoneal mass.(3,4) Retroperitoneal tumours of leiomyomatous origin have an incidence of approximately 1.2%.(5,6) There is no clear consensus on the origin of these tumours, but there are theories that claim they originate either from smooth muscle tissues that are susceptible to hormone influence (7), from embryonic remnants of Wolff or Muller ducts (8), or of iatrogenic origin, as one study revealed that between 67 and 80% of patients with extra uterine leiomyoma had surgical history of hysterectomy or laparoscopic myomectomy with morcellation of the fibroma.(9) The presence of retroperitoneal fibromas has been associated with medical history of uterine fibromatosis (22.2%), or with uterine fibroma synchronous with the retroperitoneal one. (29.2%).(5) In the case of our presented patient, there was no history of surgical procedures, and has presented uterine fibroma synchronous with the diagnosis of the retroperitoneal tumour.

Imaging holds a crucial role in the management of pelvic retroperitoneal tumours. Although it is not possible to differentiate between malignant or benign origin based on imaging alone, certain features like invasive growth, heterogeneous appearance or extensive central necrosis can be suggestive for leiomyosarcoma. Although CT scans can successfully show the anatomical limits of the tumour and describe homogenous attenuation for leiomyomas, these types of tumours are best described by MR imaging. Leiomyomas have signal that is isointense to that of muscle on T1-weighted images and hypointense to that of muscle on T2-weighted images, with homogenous enhancement. There is one cited prospective study done on 45 patients that found MRI to have a 95% sensitivity and 72% specificity for diagnosing an uncomplicated leiomyoma and a 10% sensitivity and 100% specificity for a cellular leiomyoma.(10,11) Our patient underwent a CT scan of the abdomen and pelvis, revealing the tumor to have well-defined margins, a feature mostly associated with benign pathology.

Regarding tumour markers, the patient had an elevation of over 3 times the normal limit of CA-125. While this marker is more specific for ovarian malignancies and not benign pathologies, there are studies that reveal a correlation between a high level of the marker and the size of the leiomyomas (12), as well as a decrease in the level of the marker following a decrease in the size of the fibroma after conservative treatments were applied.(13) Thus, it could be explained the high level of CA-125 in the context of a 20cm large fibroma. This marker could be used for monitoring the evolution of large-sized leiomyomas.(14)

Symptoms of pelvic retroperitoneal tumours are not specific most of the time, beginning to manifest when compression of the adjacent organs occurs. In a study of 56 patients over a 10-year period, all retroperitoneal pelvic tumours have been diagnosed in their symptomatic stage, most frequent symptoms including abdominal pain (83%), which was the case of our patient, urinary (51%) and gastrointestinal (42%) symptoms.(15,16)

A common practice in the management of retroperitoneal tumours is performing a minimally invasive biopsy. This practice has certain advantages regarding precise diagnosis and facilitating a multidisciplinary approach. However, some studies suggest that local recurrence and tumour seeding following biopsy is low, but not negligible.(17) Because the patient presents symptoms due to the size and positioning of the tumour, we have considered biopsy is not necessary due to the clear surgical indication. Also, precise diagnosis can prove to be difficult with a small biopsy sample, and a much more detailed and exact description can be provided examining the whole resected tumour.(18,19)

Elective treatment of these tumours is surgical resection, given that symptoms are present and the histological type is unknown. The most common surgical approach to the resection of these tumours is through laparotomy. Laparoscopic approach has been shown to have many benefits, such as shorter hospitalisation, less postoperative pain and reduced intraoperative bleeding.(18,20) However, laparoscopy is not widely applicable in this kind of pathology, mainly because of the difficult approach of a pelvic retroperitoneal tumour. There are studies that describe recurrence of malignant retroperitoneal tumours at the level of the trocar sites following laparoscopic approach.(21,22) We have chosen laparotomy because of the size of the tumour, its positioning, and given its unknown histology, morcellation would imply a great oncological risk. Although conservative treatments with curative aim of these tumours have not been found, there is one case in which vascular embolization of a retroperitoneal leiomyoma was performed. The main blood supply of the tumour was the left renal artery, which after its embolization led to the decrease in size of the tumour.(23) Hormonal treatments that are used in the management of uterine fibromas have shown risk of recurrence once treatment is stopped.(24)

CONCLUSIONS

Retroperitoneal position of leiomyomas is a rare occurrence, with no specific symptoms, and thus distinguishing them from other pathologies becomes difficult, more so as there are no markers specific for diagnosis. Because symptoms occur most of the time after the tumour has grown significantly, large tumour size can raise issues regarding diagnosis and surgical management.

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