Giant retroperitoneal leiomyoma
A case report and review of the literature

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Giant retroperitoneal leiomyoma. A case report and review of the literature.

Retroperitoneal leiomyomata are infrequent, and their prevalence among primary retroperitoneal tumours has been estimated as 0.5-1.2%. The authors report a case of symptomatic retroperitoneal leiomyoma with a favourable prognosis. A 53-year-old woman presented for abdominal pain associated to an inflammatory syndrome. A contrast-enhanced computed tomography revealed a large abdominopelvic mass and patient underwent open surgical excision. Definitive diagnosis was done after immunohistochemical assessment. Immunoreactivity was strong for smooth muscular actin. Presence of oestrogen and progesterone receptor proteins was also detected. Prognosis of these well-differentiated smooth muscle tumours is generally favourable but a postoperative surveillance is always recommended.

KEY WORDS: Leiomyoma, Retroperitoneal space, Surgery

Introduction

Primitive retroperitoneal tumours represent 1% of tumours and 15-20% of soft tissue tumours. Frequently (65-86%) are malignant [1-3]. Embryologically originates from the mesoderm [4]. Histologically, there are muscular adipose, and fibrous histiocytaire tumour [5-8].
Retroperitoneal tumours are generally paucisymptomatic (abdominal discomfort, pain radiating to the back, weight loss) or yet be totally asymptomatic. These neoplasms are frequently discovered incidentally during routine check-up or autopsy [9].

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Case report

We report the case of a 53-year-old nullipara woman admitted in emergency for an increasing left side abdominal pain evolving from 15 days. Her clinical history included hypertension, overweight with a body mass index (BMI) at 36 kg/m^2. She had undergone a laparotomic appendectomy 24 years ago, and a transvaginal hysterectomy with enterocele repair 5 years ago. Physical examination revealed a solitary painful tender mass in the left abdominal lower quadrant. Blood tests found an inflammatory syndrome with a C-reactive proteine level at 25 mg/L, without leukocytosis. Abdominal ultrasound scan revealed a solid, heterogeneous echoic abdominopelvic lesion. Contrast-enhanced abdominal computed tomography (CT) showed a large mass of the retroperitoneum (maximum diameter of 24 cm), extended also in the pelvic space and displacing the left colon anteriorly (Fig. 1). Neither lymph node involvement nor visceral metastases was found. A transparietal ultrasonography-guided fine-needle aspiration biopsy suggested a smooth muscle cell origin, but it was low contributive and uncertain in order to exclude the diagnosis of sarcoma. The patient underwent a laparotomic

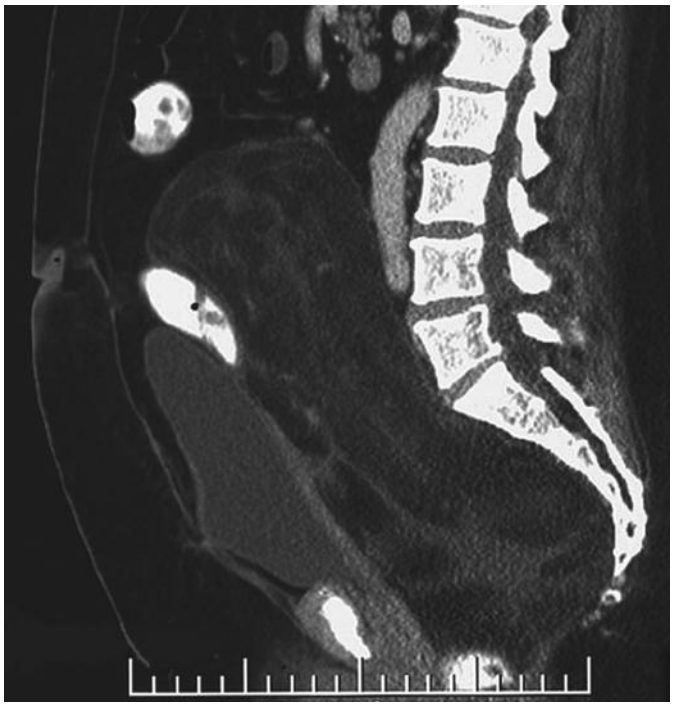

Fig. 1: Contrast-enhanced Computed Tomography: large retroperitoneal tumour (sagittal view).

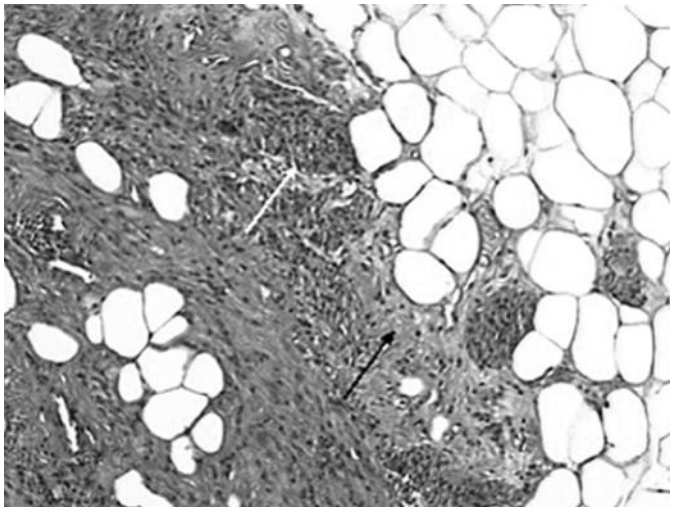

Fig. 2: Histological examination: mature adipocytes (black arrow) and spindle cells (white arrow) (hematoxylin-eosin, x100 magnification).

excision of the retroperitoneal mass, which effected in complete tumour removal. Dissection was laborious, and a supplementary left colectomy was also done due to tumoral size and tight contacts with left mesocolon and mesentery root. Total operative time was 160 minutes. Macroscopically the specimen showed a fatty tissue mass (29x24x9 cm, 2350g) that appeared well-circumscribed, lobulated, and scarred with several whitish bands of fibrous tissue.

The microscopic findings on hematoxylin-eosin staining, showed a not-well-encapsulated tumour, with a prevalent monomorphic spindle cells cluster, trabecularly arranged, associated to a contingent of mature fat cells without lipoblasts, coagulative necrosis, cytologic atypia, and mitosis (< 1 mitosis /50 high power field [HPF]) (Fig. 2). There was no vascular pattern abnormalities. On immunostaining, smooth muscle actin (SMA), desmin, and caldesmon positivity along with negativity for human melanoma black 45 monoclonal antibody (HMB45), CD34, CD117, and S-100 protein (Fig. 3 a,b) confirmed the diagnosis of leiomyoma. The oestrogen receptor (ER) was positive in 70%, progesterone receptor (PR) in 60%. Postoperative course was uneventful, and she was discharged on 13th postoperative day. No complementary treatments were administrated. The patient underwent six monthly reviews and when last seen, at 24 months, was free of symptoms, had a normal laboratory tests and neither local recurrence nor distant metastases.

Discussion

Smooth muscle tumours occurring in deep soft tissue is a rare entity. Leiomyomata and leiomyosarcomata of soft tissue account for less than 4% of benign soft tissue lesions, and 10% of soft tissue sarcomata, respectively [5]. Leiomyomata are frequent localised in the uterus and in the parasitic uterine structures but occur rarely in the retroperitoneal space [6]. Generally, retroperitoneal smooth muscle tumour presented a malignant behavior [10]. Retroperitoneal leiomyomata (RL) have been described in the literature, and their prevalence among primary retroperitoneal tumours has been estimated as 0.5-1.2% [6,11]. These occurred predominantly in females, in the fourth-fifth decades of life, and histological aspects are similar to uterin leiomyomas [12]. A clinicopathologic evaluation on long-term follow-up with immunohistochemical analysis of RLs, to the best of our knowledge, was recently assessed [6]. Commonly these tumours are ER and PR positive [6,13]. The etiology and pathogenesis of retroperitoneal leiomyomata are still not well understood; is unclear then their can arise from visceral o vascular wall smooth muscle cells [12,14]. Histologic features highlighted the presence of composed fascicles of spindle mature smooth muscle cells, stromal hyalinisation, and fatty change associated to the absence of cellular atipya, coagulative necrosis, and low mitotic rate. Immunohistochemically, these tumours showed positivity to SMA and desmine, and negativity for CD34, S-100, and HMB45 [5,6]. Distant or local recurrence is infrequent [5]. Current management is radical excison, especially in symptomatic patients [7]. All avaible cases in the literature of RL were treated with open surgical approaches [15]. Incomplete excision of the primary tumour might have induced recurrence [6]. Moreover,

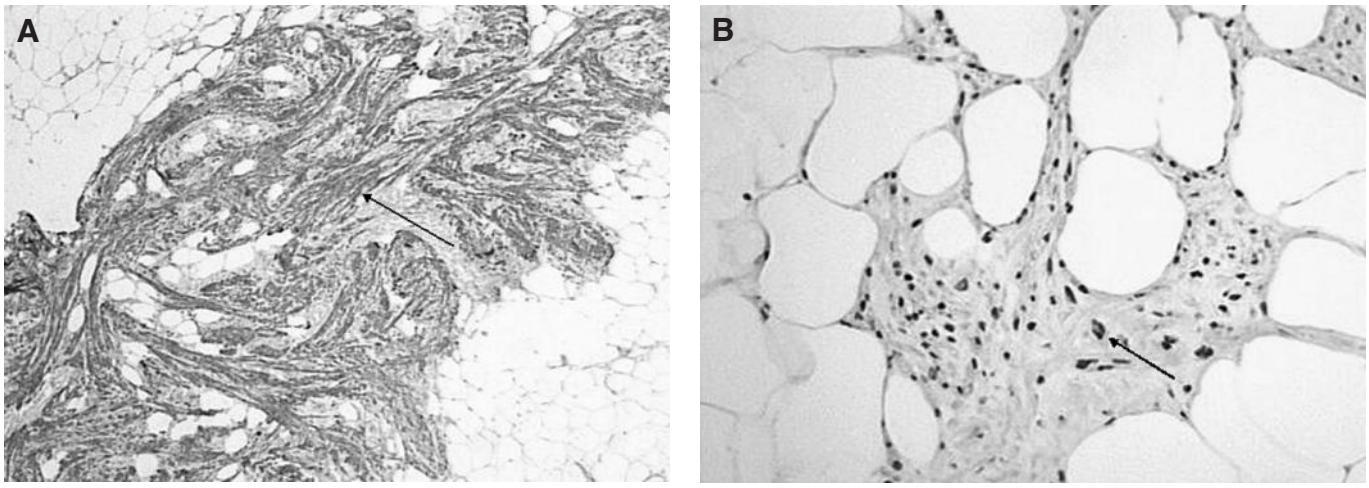

Fig. 3: Immunohistochemical assessment: A) spindle cells strong (>80%) positivity (arrow) for anti-SMA antibodies (x50); B) positivity (arrow) for anti-oestrogen receptor protein antibodies (x400).

retroperitoneal masses may displace, compress or involve adjacent organs or visceral, vascular, and nervous structures [12,16]. Definitive diagnostic is rarely assessed before surgical excision [17]. In this case the strong expression of AML, desmine and caldesmon of fusiform cells, in absence of lipoblast and malignancy aspects, allowed the diagnosis of RL. Prognosis of these well-differentiated smooth muscle tumours can be considered as probably favourable [8]. However a malignancy potential cannot excluded [5,6,15]. A careful surveillance is required [15]. The differential diagnosis of RL includes, firstly, leiomyosarcomata, liposarcomata, angiomyxomata, hemangiopericytomata, and metastatic tumours [6,17-20]. We report a new case of symptomatic benign retroperitoneal leiomyoma develloped after hysterectomy. She is alive and disease-free on a 24-months follow-up.

Riassunto

I leiomiomi retroperitoneali sono rari e la loro prevalenza tra i tumori primitivi del retroperitoneo è stata valutata come compresa tra 0.5 e 1.2%. Gli autori riportano un caso di leiomioma retroperitoneale sintomatico con prognosi favorevole. Una donna di 53 anni giunse in ospedale per dolore addominale associato a sindrome infiammatoria. Una tomografia computerizzata con mezzo di contrasto evidenziò una voluminosa massa addomino-pelvica e la paziente venne sottoposta ad escissione chirurgica per via laparotomica. La diagnosi definitiva fu posta dopo l'esame immunoistochimico. L'immunoreattività fu forte per l'actina muscolare liscia. La presenza di recettori per gli estrogeni ed il progesterone fu altresì riscontrata. La prognosi di questi tumori muscolari lisci ben differenziati è generalmente benigna ma una sorveglianza postoperatoria è sempre consigliata.

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