Solitary fibrous tumor of the pelvis: an uncommon soft-tissue tumor.
A case report

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Solitary fibrous tumor of the pelvis :An uncommon soft-tissue tumor.A case report

A rare case of Solitary fibrous tumor (SFT) of the pelvis is reported. A 76-years-old man presented with a low abdominal pain, acute urine retention and constipation. Imaging studies (US, CT, MR) showed an 17 x 10 x 9 ovoid mass in the pelvis, dislocating bladder and rectum. Finally, trans-rectal needle biopsy suggested the diagnosis of SFT. En bloc excision of tumor and rectum (because of strong adhesions) was performed. Histological examination showed spindle and fibroblastic-like cells dispersed in collagenous areas with positive stains for CD34, bcl-2, CD99 and it confirmed diagnosis of SFT. No postoperative complications occurred, only vesico-sphincter dyssynergia was found by urodinamics. After 5 years, patient is disease-free. SFT is, usually, benign tumor with slow growth and excellent prognosis. Complete surgical resection is the only curative treatment. However, 10-15% of SFT are malignant and histological findings cannot always predict clinical behaviour. For this reason, careful and long term follow-up is necessary after surgery.

KEY WORDS: Pelvis, Pelvic space, SFT, Solitary Fibrous Tumor, Spindle cells.

Introduction

Solitary fibrous tumor (SFT) is an uncommon soft-tissue tumor, usually arising from pleura, also if, in the last years, extrathoracic SFTs have been reported with increased frequency.
Few case of pelvic SFT, usually with short follow-up, are reported in literature [1-3]. We describe a SFT originating from the pelvic space with 5-year follow-up and analyze the histological, clinical and therapeutic features of this tumor.

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Case presentation

In November 2004, a 76-years-old man presented with a low abdominal pain, acute urine retention and constipation. Physical examination revealed a palpable abdominal mass in hypogastrium (Fig. 1).
Abdominal ultrasonography revealed a bilateral hydronephrosis and an well-defined hypoechoic mass with small central anechoic areas in pelvic space.
Abdominal Computed Tomography (CT) showed a pelvic-occupying mass compressing bladder and rectum. Pelvic Magnetic Resonance (RM) confirmed the presence of ovoid mass (Fig. 2), having iso-signal intensity on the T1-weighted image and high signal intensity on the fat-saturated T2-weighted image. A normal levels of oncologic markers (AFP, CA 19.9, CEA, PSA, TPA) were noted. Finally, trans-rectal needle biopsy suggested the diagnosis of SFT.
The patient underwent surgery. Because of strong adhesions, successful en bloc resection of the mass with the rectum was performed and end-to-end anastomosis was realized.

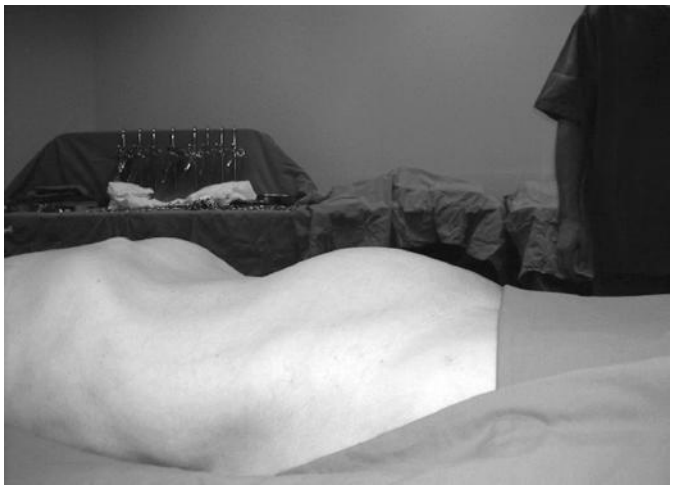

Fig. 1: Palpable abdominal mass in our patient affected from SFT.

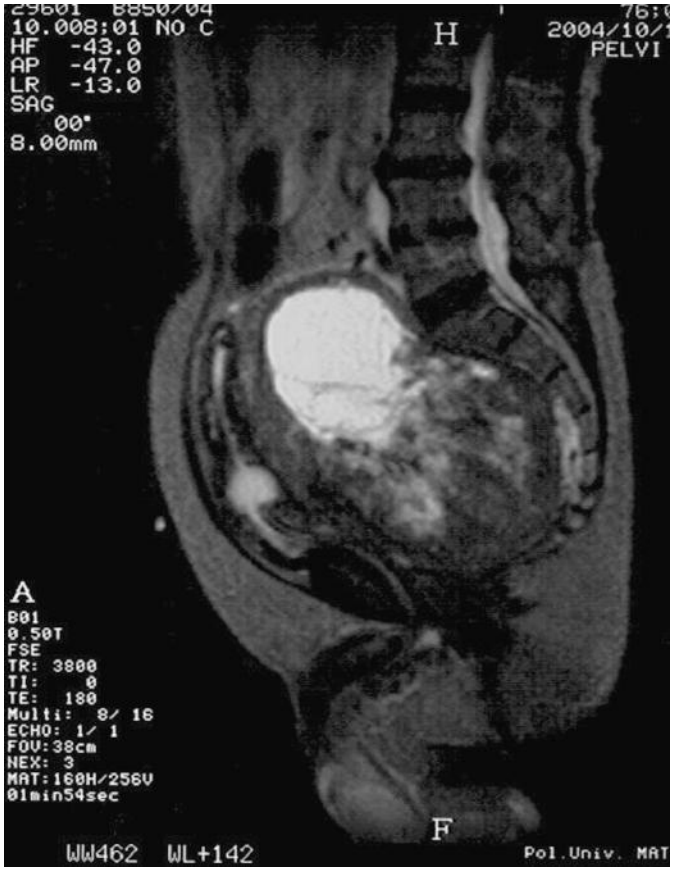

Figure 2. Magnetic resonance of abdomen showing SFT, compressing bladder.

Gross examination revealed a well-encapsulated 17 x 10 x 9 cm rubbery mass, pink-white on cut surface and weighed 1200 g (Fig. 3).
Histological examination revealed spindle and fibroblastic-like cells dispersed in collagenous areas, with multiple small vascular spaces and same mitotic figures.

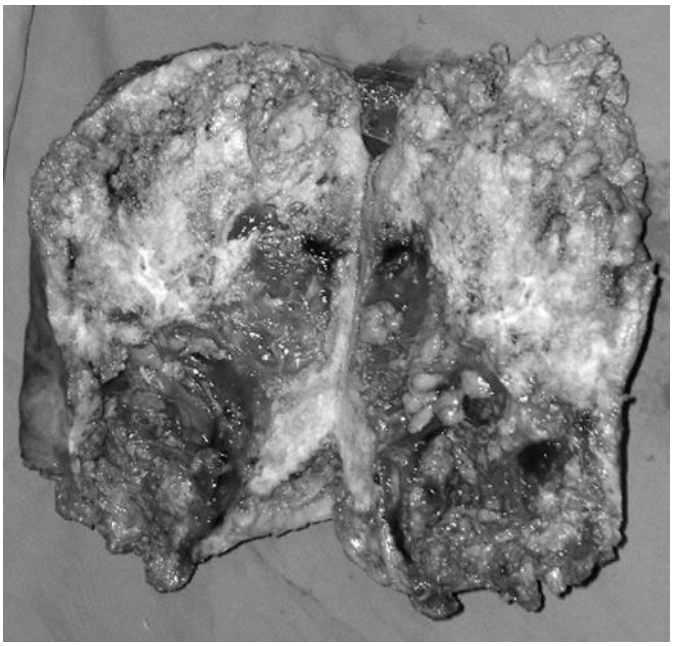

Fig. 3: Gross appearance of well-demarcated SFT of the pelvis

Immunohistochemical analysis showed strong positive marking for CD34, bcl-2, CD99, but negative stain for SMA and S-100. The final diagnosis was made as benign solitary fibrous tumor of the pelvis.
No postoperative complications were observed, also if urodinamics revealed vesico-sphincter dyssynergia. To date, after follow-up of 5 years, patient is disease-free.

Discussion and Conclusions

Even tough STS was first described as pleural neoplasm[4], in the last years the number of extra-pleural SFT reported increased exponentially, probably because until 20 years ago, most of the SFTs, for lack of clear criteria, were diagnosed from pathologist like hemangiopericytomas [5].
Typically, this tumor presents as well-defined and slow-growing mass dislocating and compressing neighbour organs, also if there were reported same cases associated with secretion of insuline-like growth factor 2 [6,7].
Microscopically SFT shows a classic pattern ("patternless pattern") with spindle cells in collagenous background. Immunohystochemically it commonly expresses CD34, bcl-2, vimentin, CD99 and, sometimes, also epithelial membrane antigen (EMA) and smooth muscle actin (SMA). It is usually negative for S100 protein, desmin and cytokeratins [2,8].
In the pelvic space, preoperative diagnosis can be established by transrectal needle biopsy [9,10].
About 10-15% of solitary fibrous tumor are malignant [11, 12]. Criteria of malignancy include high cellularity,

nuclear pleomorphism, area of tumor necrosis and high mitotic activity [>4/10 high-power fields (HPF)] [11,13]. CD34 expression can be lost in high-grade tumours, similar to the other soft-tissue tumours [14,15]. Metastases occur in lungs, liver and bone.
Clinical behaviour is, however, unpredictable [16]: even tough in most of the cases prognosis after complete resections is good, histological findings cannot always predict clinical behaviour and careful long-term follow-up is necessary [17-19].
Complete surgical excision is the treatment of choice, while adjuvant chemotherapy and radiotherapy for malignant forms, due to their rarity, are not evaluable.

Riassunto

Gli Autori descrivono un raro caso di Tumore Fibroso Solitario (TFS) a localizzazione pelvica in un paziente di anni 76 esordito con ritenzione acuta di urina, dolore addominale ai quadranti inferiori e stipsi.
Gli esami strumentali effettuati (ecografia pelvica e transrettale, TC con m.d.c RM) evidenziavano la presenza di massa espansiva a verosimile localizzazione retroperitoneale, grossolanamente rotondeggiante, di circa 12,5 cm di diametro, ad ecostruttura disomogenea. Nella norma l'Rx torace, gli esami ematochimici ed i marcatori tumorali esaminati (AFP, CA 19.9, CEA, PSA, TPA).
Una biopsia tran-rettale eco-guidata della lesione evidenziava un quadro morfologico ed immunofenotipico coerente con la diagnosi di Tumore Fibroso Solitario.
All'intervento chirurgico per l'exeresi della neoformazione la massa ovalariforme di cm 17x10x9, capsulata, a sede retrovescicale fino al piano degli elevatori, dislocante anteriormente la vescica, risultò essere ben clivabile dal piano prostato-vescicale ma nettamente aderente al retto. Pertanto l'exeresi fu estesa in blocco dalla neoplasia, al retto fino al piano degli elevatori, del mesoretto e del sigma, con conseguente anastomosi termino-terminale.
decorso postoperatorio privo di complicanze, salvo una instabilità detrusoriale con alterato svuotamento vescicole da dissinergia vescico-sfinteriale documentata con esame urodinamico. L'esame istologico definitivo confermava la diagnosi di TFS.
L'alta variabilità del Tumore Fibroso Solitario (TFS) è responsabile di una notevole difficoltà diagnostica, soprattutto quando la neoplasia si manifesta in siti extratoracici. La diagnosi differenziale va posta con l'emangiopericitoma, ma anche con il fibroistiocitoma maligno, il leiomioma, il leiomiosarcoma, lo schwannoma, il carcinosarcoma ed i tumori stromali gastro-intestinali (GIST), che l'istochimica può chiarire.
Come per le lesioni intratoraciche, il comportamento dei TFS extratoracici è imprevedibile: approssimativamente il 10% dei TFS extratoracici sono considerati maligni. Le principali caratteristiche di malignità sono l'ipercellularità, il pleomorfismo nucleare, la presenza di estese aree di necrosi e soprattutto un'elevata attività mitotica, maggiore di 4/10 HPF. Al contrario, fattori prognostici positivi sono una scarsa attività mitotica, la presenza di una capsula, di un peduncolo e la resecabilità.
La completa escissione chirurgica della neoplasia risulta essere di solito curativa e non vi è necessità di radioterapia o chemioterapia postoperatoria, considerando che le sedi extrapleuriche sono diventate un'entità nosologica nota solo da pochi anni è, a nostro avviso, razionale sottoporre i pazienti ad un follow-up accurato a lungo termine.

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