

Small bowel mesentery solitary fibrous tumor. A rare neoplasia in a young male

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SUMMARY: Small bowel mesentery solitary fibrous tumor. A rare neoplasia in a young male.

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Extrapleural Solitary Fibrous Tumors (SFT), in particular small bowel mesentery SFTs, are extremely rare neoplasms.

We describe the case of a young male hospitalized for unspecific abdominal symptoms and evidence of a well-circumscribed mass arising from the small bowel mesentery. Histopathological and immunohistochemical analysis on the surgical specimen confirmed the diagnosis of SFT.

A Pubmed search revealed only another case of small bowel mesentery SFT, confirming the extremely rarity of this tumor.

RIASSUNTO: Tumore fibroso solitario del mesentere. Una neoplasia rara in un giovane uomo.

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I tumori fibrosi solitari extrapleurici, in particolare quelli interessanti il mesentere dell'intestino tenue, rappresentano una entità rara.

Riportiamo il caso di un giovane uomo ricoverato per una sintomatologia addominale aspecifica associata alla presenza di un processo espansivo, ben delimitato, a partenza dal mesentere dell'intestino tenue. L'esame istologico con valutazione immunohistochemica della neoplasia ha permesso di porre diagnosi di tumore fibroso solitario del mesentere.

Una ricerca di PubMed ha evidenziato solo un altro caso di tumore fibroso solitario del mesentere, confermando l'estrema rarità della neoplasia.

KEY WORDS: Extrapleural solitary fibrous tumor - Small bowel mesenchymal tumor - CD34 staining.
Tumore fibroso solitario extrapleurico - Tumore mesenchimale dell'intestino tenue - Positività per CD34.

Introduction

Recognized for the first time as a distinctive pleural lesion in 1931, SFT is considered an uncommon spindle cell neoplasm. In subsequent decades, additional intrathoracic locations, including intrapulmonary, mediastinal, and epi/pericardial tumors were described, although the most common site for SFT remains pleural surface (1, 2). Literature review reveal different extrathoracic site: peritoneal cavity, retroperitoneum, head and neck (3). Extrapleural SFT occur slightly more frequently in men than women, the mean age of presentation is 54 years (4).

Clinical manifestations are related to the size and localization of the lesion. Characteristic microscopic features are described as a "patternless" growth pattern, with bland spindle cell cytology, alternating hyper-hypocellular areas, keloid-like hyalinization and frequently a prominent branching vasculature often described as hemangiopericytoma-like (5).

Complete local excision is curative in most cases, although local recurrence and distant metastases are known (6). Hystologic features associated with local or distant recurrence include large tumor, high cellularity, mitotic activity (> 4/10 HPF), nuclear pleomorphism and necrosis (7). Often extrathoracic SFTs appear to be uneasily distinguishable from other mesenchymal tumors such as GISTs or leiomyosarcoma (8, 9). The differential diagnosis also include mesenteric fibromatosis, sclerosing mesenteritis and inflammatory pseudotumor (10). Considering that radiologic appearance of benign fibrous tumor and tumor-like lesions of the mesentery overlap with those of lymphoma, metastatic carcinoid, soft-tis-

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sue sarcoma and primary gastrointestinal malignancies (10), immunohistochemical analysis result mandatory for a differential diagnosis (11, 12). A reliable prediction of behaviour in these lesions is still difficult (6).

Case report

A 26-year-old male was admitted at our General and Emergency Surgery Department with ultrasound diagnosis of a 10 cm retrovesical lobulated mass. Complained symptoms were abdominal fullness and urinary frequency in the last month. There were no major medical or surgery history in the past, neither a familiar history of intestinal cancer.

On admission physical examination revealed a non-painful, firm mass in the lower central abdominal quadrant. Blood exams and tumor markers (CEA, CA 19-9, alpha-fetoprotein) were all within normal range.

MRI demonstrated a well-circumscribed mass, with heterogeneous high-signal intensity on T2-weighted images and homogeneous intermediate signal intensity on T1-weighted images. After gadolinium injection there was intensive enhancement of the lesion, but the center remained unenhanced with an intermediate intensity on T1-weighted images. A thoracic-abdominal CT scan didn't reveal any signs of metastatic spread or peritoneal dissemination of the tumor. The neoplasm appeared as a well defined mass with solid and cystic features.

The patient was scheduled for an exploratory laparotomy during a multidisciplinary staff. A midline incision was performed. At inspection a 12 cm firm, capsulated mass arising from the proximal ileum mesentery was detected. No pathologic mesentery lymph nodes were revealed at inspection. A 30 cm ileum resection was performed with primary anastomosis. At macroscopic examination the tumor appeared as a well-defined, firm mass with a pale cut surface, without necrosis foci. Microscopic pathological examination described a mesenchymal tumor with hyalinized collagen and spindle-shaped cells organized in a fascicular pattern. Mesenteric lymph nodes were negative for atypical cells. Mitotic count was < 4/10 HPF, with no nuclear pleomorphism. Immunohistochemical profile revealed strong and diffuse staining for CD34, CD99 and Bcl2 while CD117 and S100 were negative. The pathologic findings were evocative of solitary fibrous tumor of the mesentery.

The post operative course was uneventful, patient was discharged in VII p.o day. Regarding the pathological findings of an intermediate tumor behaviour the patient underwent clinical and radiologic follow-up every three months. At 18 months from surgery no evidence of recurrence or metastatic disease have been detected.

Discussion and conclusions

SFT is an uncommon mesenchymal neoplasm involving both intra and extrathoracic sites (1-4).

In literature a male preponderance is described, with a median age of 54 years (13). Symptoms of the tumor

may be unspecific, they might be related to organs compression or dislocation when dealing with intrabdominal SFTs.

Pathological pattern can mime other mesenchymal tumors such as GIST, synovial sarcoma, leiomyosarcoma, spindle cell thymoma, spindle cell carcinoma, dedifferentiated liposarcoma, malignant peripheral nerve sheath tumor and desmoplastic mesothelioma (14). Immunohistochemical analysis are crucial in the differential diagnosis especially by the positive staining of CD34 and CD99. Despite CD34 is not entirely specific for SFT, the combination of CD34 with B-cell leukaemia/lymphoma-2 (bcl-2) oncogene seems to be diagnostic of solitary fibrous tumor (11). Moreover bFGF and Ki-67 staining are strong indicators of a malignant behaviour of SFT (12).

Radiologic features appear not to be highly specific for SFT diagnosis (10).

The clinical behaviour is usually benign. Nevertheless is described an aggressive behaviour (15, 16). SFTs have the potential to locally recur in the peritoneal surface and to metastasise to liver and lungs (6). Vallat-Decouvelaere et al. underlined in their paper that nuclear atipia, hypercellularity greater than 4 mitoses/10 HPFs, and necrosis may be seen in up to 10% of extrathoracic SFTs, and are associated with an aggressive clinical behaviour (7). Tumor size represent an independent prognostic factor, with a 5-year survival of 20% in patients with SFTs larger than 10 cm (4, 17). Distinctive features signalling malignant behaviour are the presence of extracapsular extension and the elevated number of mitoses associated with the finding of hypoglycemia in both the initial presentation of the disease and on subsequent recurrent episodes (18, 19).

Radical surgery represent the first choice treatment of this neoplasm. The use of integrated treatment such as cytoreductive surgery plus hipec in peritoneal disease is described in literature. Systemic chemotherapy may be advisable in case of metastatic spread. In these cases the prognosis appears to be poor (6).

In conclusion the unpredictable behaviour of extrathoracic SFTs require a careful, close, long term follow up.

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