



Solitary Fibrous Tumour

A rare cause of acute abdomen



Ann Ital Chir, Digital Edition 2017, 6
pii: S2239253X17026767 - Epub, March 29
free reading: www.annitalchir.com

Nicole Grech*, Ernest Ellul*, Neville Spiteri*, Miljan Milic**

*Department of Surgery, Mater Dei Hospital, Msida, Malta

**Department of Histopathology, Mater Dei Hospital, Msida Malta

Solitary Fibrous Tumour. A rare cause of acute abdomen

Solitary Fibrous Tumours (SFT), previously known as haemangiopericytoma, are rarely encountered in surgery. They arise from mesenchyme tissue and can occur at several sites in the body – head and neck, extremities, thorax, abdomen and retroperitoneal space. In the thorax, where they arise from the pleura, and abdomen they may attain a large size before giving rise to symptoms.

Most SFT behave in a benign manner. However a number of them recur locally or metastasize. Recurrences can occur several years after excision of the primary tumour.

Complete surgical excision remains the primary modality of treatment. But, in sites where complete excision is not possible, other modalities have been tried with varying success.

Here, we describe a SFT of the mesentery of the small intestine, an uncommon manifestation of the tumour, recurring after a period of 19 years in a 55 year old female, and presenting to the Emergency Department as an acute abdomen caused by acute intestinal obstruction. Surgical excision of the tumour was performed together with primary anastomosis of the small intestine.

KEY WORDS: Acute abdomen, Late recurrence, Mesentery, Solitary Fibrous tumour

Introduction

Solitary fibrous tumour (SFT) is a term used to describe a heterogenous group of benign and malignant neoplasms of soft tissues¹. It is a tumour met rarely, with a reported incidence of 0.2/100000/years². It shares several histological features with haemangiopericytoma, a vascular sarcomatous tumour described in 1942 by Scott and

Murray³, although the same tumour had been described by Klemperer and Rabin in 1931 when the term 'solitary fibrous tumour' was first used⁴. In 2002, the World Health Organisation (WHO) defined the term 'fibrous tumour' as a broad term encompassing several mesenchymal tumours, including haemangiopericytoma^{5,6}.

We report here a recurrent solitary fibrous tumour occurring in the abdominal cavity 19 years after first presentation, and presenting as an acute abdomen.

Case Report

A 55-year-old lady, with a history notable for excision of small bowel SFT 19 years previously, presented to the Emergency Department complaining of a 3-day history of increasingly severe generalised abdominal pain, later localising to the lower abdomen. She also complained of

Pervenuto in Redazione Dicembre 2016. Accettato per la pubblicazione Febbraio 2017

Correspondence to: Ernest Ellul, Mater Day Hospital, Surgical Ward 5, Msida, Malta MSD 2090 (E-mail: ernest.ellul@gov.mt)

one bout of melaena but on further questioning no other symptoms such as nausea, vomiting, epigastric burning or lower urinary tract symptoms were elicited. She had been monitored for 10 years following surgery for possible recurrence of the tumour.

On examination, the abdomen was found to be slightly distended with generalized tenderness, and rebound tenderness in the lower abdomen. Bowel sounds were absent. Laboratory investigations showed low haemoglobin (10.3 g/dl) together with a normal leucocyte count, normal liver and renal function, normal glucose (6.88 mmol/l). Plain Abdominal X-Ray film suggested small intestinal obstruction. A contrast enhanced Computed Tomography(CT) Scan of the abdomen and pelvis was therefore performed. In the portovenous phase, this showed a heterogeneous enhancing mass in the right lower quadrant of the abdomen involving the distal ileum and causing distal small bowel obstruction. The patient underwent laparotomy during which a large mass in the small bowel mesentery encroaching on the ileum was excised. Wide excision was attempted with subsequent primary stapled small bowel anastomosis (GIA 80 Covidien ©). Postoperative recovery was uneventful with the patient well enough to be discharged home on the eighth postoperative day.

On gross pathology, the specimen was found to consist of a soft polypoid mass 80mm in diameter pushing against an adjacent loop of small intestine. Haemorrhage and necrosis were also present in part of the tumour mass. Small intestine proximal to the mass was dilated. Histological examination revealed solitary fibrous tumour with clear resection margins and no lymph node involvement. Neoplastic cells expressed Vimentin and CD34 antigens but not CD 117 and DOG-1, thus distinguishing it from gastrointestinal stromal tumour. Nuclear pleomorphism appeared to be moderate, with approxi-

mately 10 mitoses per 10 high power field (10 mitoses/10 HPF) noted, together with areas of necrosis and haemorrhage. This was considered a high-risk tumour ⁵.

In comparison, the tumour previously excised 19 years ago was 180mm in diameter. It showed moderate nuclear pleomorphism and contained haemorrhagic areas. Neither mitoses nor areas of necrosis were seen. Excision was deemed complete.

She was re-admitted 3 days later complaining of pyrexia, abdominal pain and a foul-smelling purulent discharge from the lower part of the laparotomy wound. An anaemia of 9.6 g/dl together with a leucocytosis of $29.7 \times 10^9/l$ was reported. C-Reactive Protein was raised at 385mg/l.

A fresh abdominal CT Scan showed features of an enterocutaneous fistula arising from the site of the anastomosis to the abdominal wall, with a loculated abscess in the abdominal wall itself. Wound toilet with drainage of the small abscess was performed. Conservative management of the fistula consisting of total parenteral nutrition, antibiotic therapy and subcutaneous Octreotide, was instituted.

After four weeks of conservative management, leakage from the abdominal wound had ceased completely, the wound healed and a third CT scan confirmed healing of the enterocutaneous fistula.

She was therefore discharged home, and is being followed up as an outpatient together with the Department of Oncology.

Discussion

SFT is an uncommon tumour, representing only about 1-2% of all soft tissue tumours. It behaves in a benign



Fig. 1: large mass in right lower quadrant, dilated small bowel loops



Fig. 2: contrast enhancement of mass in the portovenous phase.

fashion in about 80% of cases. Classically, three types of SFT are recognized: pleural, meningeal and soft tissue¹. The mean age of presentation of extra-pleural SFTs is 54 years with a slightly higher incidence in males⁸. The exact pathogenesis of these tumours is as yet unknown^{16,17} although it has been postulated that they arise from submesothelial mesenchymal connective tissue⁸. Recently, it has been recognized as a translocation-associated neoplasm arising from recurrent rearrangements in chromosome 12q⁹.

Abdominal SFTs are then more common than tumours in the extremities⁵. Abdominopelvic SFT are commonly painless and only come to light either as incidental findings¹⁰, or as result of secondary symptoms due to mass effect causing obstruction, distension, constipation, or urinary tract obstruction¹¹. They can therefore reach a large size prior to diagnosis. Abdominopelvic and pleural SFT are consequently significantly larger at diagnosis than those in the head and neck, or extremities^{5,11}. The frequency of metastases and clinical outcome of thoracic and abdominal SFTs is, however, similar to that of SFTs anywhere else in the body^{5,13}.

Another clinical presentation of SFT is that of hypoglycaemia. This is due to a paraneoplastic syndrome where the tumour secretes insulin-like growth factor-2 (IGF II)¹².

Although they must grow to a larger size to cause symptoms, pleural SFT are diagnosed more frequently; this is postulated to be due to the frequency of requesting chest radiographs for unrelated pathology, when a mass in the chest - a SFT - is detected as an incidental finding¹³. In the literature, very few cases of SFTs related to bowel have been reported so far - Bassi¹⁴ described an incidental finding of one such tumour in the retroperitoneal area of a previously healthy 27-year-old male while being investigated for pneumonia. The tumour was at first managed expectantly but later required resection due to mechanical obstruction of the inferior vena cava. The mass was adjacent to the duodenum, renal hilum, aorta and pancreas but was not invasive.

Lau¹⁵ reported a case of a 53-year-old gentleman presenting with a 10-day history of nausea, abdominal pain, lethargy and constitutional symptoms. On examination a large mass in the right lower quadrant that was tender on deep palpation was elicited. Tumour markers for carcinoma were unremarkable. In this case, the tumour arose from the mesentery of the ascending colon and was resected as part of limited right hemicolectomy with subsequent primary anastomosis. Histological examination of the specimen confirmed SFT.

Histologically, criteria for diagnosis of SFT include small cells separated by strands of collagen, with hyperchromatic nuclei and scanty cytoplasm. Cellular arrangement is haphazard and often exhibits various areas of hypercellularity with adjacent areas of hypocellularity¹⁸.

CD34 expression is common in SFTs. Most SFTs are positive for CD34 but negative for CD117 and DOG-

1, a feature that helps to distinguish them from gastrointestinal stromal tumours (GIST)^{19,20}. CD34 expression also helps distinguish SFTs from other uncommon tumours such as desmoplastic mesothelioma²¹.

In studies involving numerous cases of solitary fibrous tumours, Vimentin was expressed in all histological sections of the tumour^{22,23}.

An anaplastic form known as Mosquera exists and is associated with a poorer prognosis if larger than 8cm in diameter¹⁸.

Surgical resection with negative margins remains the advocated treatment for solitary fibrous tumours²⁴. Although the correlation between positive microscopic resection margins and subsequent metastasis was found to be strong¹³, in a series of recurrent or metastatic tumours⁵ margin status was not found to be a relevant factor. It is important to note that only 10% of recurrences in this series came from abdominopelvic tumours. In both the thoracic and peritoneal cavities, where minimally invasive techniques have become well-established, SFT have also been successfully resected laparoscopically with microscopic negative margins, giving obvious benefit to the patient in terms of postoperative recovery, and seemingly without an added risk of recurrence^{25,26}.

Cytotoxic chemotherapy has not proved to be of significant benefit to the patient². Radiotherapy is mainly used as adjuvant in the treatment of head and neck, and retroperitoneal tumours whose resection margins did not show complete resection of tumour. It has also been reported to be of benefit in treatment of recurrent disease in the pleura²⁷ and in advanced pelvic tumour²⁸. Tyrosine kinase inhibitors (Sunitinib) and insulin-like Growth factor I receptor (IGFIR) (Figitumab) are now being studied in the treatment of locally advanced or metastatic tumours resistant to systemic chemotherapy²⁹. Solitary fibrous tumours are rare encounters in surgery, with those arising in the bowel mesentery comprising only a small minority of these tumours. It is known that the majority behave in a "benign" fashion. Local recurrence or metastatic disease worsen prognosis. Indeed, in a review of a small series of recurrences² the histological picture was always found to be more aggressive than that at first presentation. Strict surveillance of patients treated for solitary fibrous tumour is therefore advised³⁰.

Riassunto

I Tumori Fibrosi Solitari (SFT), precedentemente conosciuti come emangiopericitoma, si incontrano assai raramente in chirurgia. Essi nascono dal tessuto mesenchima, pertanto possono essere presenti in diversi parti del corpo - nel cranio e nel collo, negli arti, nel torace, nell'addome e nello spazio retroperitoneale. Nel torace, essi provengono dalla pleura; nell'addome possono raggiungere grandi dimensioni prima di dar luogo a sinto-

mi. Spesso vengono scoperti in un modo del tutto accidentale.

L'escissione chirurgica completa rimane la principale modalità di trattamento. Tuttavia, in siti dove l'escissione completa non è possibile, altre modalità sono state provate con risultati variabili.

Descriviamo un SFT nel mesentere dell'intestino tenue, un sito insolito per l'insorgenza di questo tumore, recidivo dopo un periodo di 19 anni in una donna cinquantacinquenne giunta al Pronto Soccorso di quest'ospedale. Accusava dolori addominali da un paio di giorni. Il suo quadro clinico si presentava suggestivo per una occlusione acuta dell'intestino tenue.

Sottoposta a TAC dell'addome, si confermava il sospetto diagnostico con la presenza di una massa derivante dall'ileo nel quadrante inferiore destro dell'addome. All'intervento chirurgico è stata eseguita l'asportazione del tumore con anastomosi primaria dell'intestino.

La maggior parte degli SFT si comportano in maniera benigna. Tuttavia, un certo numero di loro ricorrono a livello locale o con metastasi. Le recidive possono verificarsi vari anni dopo l'asportazione del tumore primario.

Pare che i tumori ad alto rischio di metastasi e di mortalità siano quelli che occorrono in pazienti al di sopra dei 55 anni, quelli con dimensioni $\geq 15\text{cm}$, e in cui le mitosi sono $\geq 4/10$ HPF.

References

1. Penel N, Amela EY, Decanter G, Robin YM, Marec-Berard P: *Solitary fibrous tumors and so-called hemangiopericytoma*. *Sarcoma*, 2012; 2012: 690251.
2. Baldi GG, Stacchiotti S, Mauro V, Dei Tos AP, Gronchi A, Pastorino U, Duranti L, Provenzano S, Marrari A, Libertini M, Pilotti S, Casali PG: *Solitary fibrous tumour of all sites: outcome of late recurrences in 14 patients* *Clinical Sarcoma Research*, 2013 3: 4.
3. Stout AP, Murray MR: *Hemangiopericytoma: A vascular tumor featuring Zimmermann's pericytes*. *Ann Surg*, 1943; 16(1):26-33.
4. Klemperer P, Rabin CB: *Arch. Pathol.* (Chicago), 1931;11: 385.
5. Demicco E, Park M, Araujo D, Fox P, Bassett R, Pollock R, et al.: *Solitary fibrous tumor: A clinicopathological study of 110 cases and proposed risk assessment model*. *Mod Pathol*, 2012; 25(9):1298-306.
6. Gengler C, Guillou L: *Solitary fibrous tumour and haemangiopericytoma: Evolution of a concept*. *Histopathology*. 2006; 48(1):63-74.
7. Young RH, Clement PB, McCaughey WT: *Solitary fibrous tumors ('fibrous mesotheliomas') of the peritoneum: A report of three cases and a review of the literature*. *Arch Pathol Lab Med*, 1990;114: 493-95.
8. Dervan PA, Tobin B, O'Connor M: *Solitary (localized) fibrous mesothelioma: Evidence against mesothelial cell origin*. *Histopathology*, 1986; 10(8):867-75.
9. Khin Thway, Wen Ng, Noujaim J, Jones RL, Fisher C: *The current status of solitary fibrous tumor: diagnostic features, variants, and genetics*. *Int J Surg Pathol*, 2016; 24:281-92.
10. Liu YQ, Yue JQ: *Intramural solitary fibrous tumor of the ileum. A case report and review of the literature* *J Can Res Ther*, 2013; 9: 724-76.
11. Wang H, Chen P, Zhao W, Shi L, Gu X, Xu Q: *Clinicopathological findings in a case series of abdominopelvic solitary fibrous tumors* *Oncol Lett*, 2014; 7(4):1067-72.
12. Froesch ER, Zapf J, Widner U: *Hypoglycaemia associated with nonislet cell tumour and insulin-like growth factors*. *New Engl J Med*, 1982; 306:1178-179.
13. Gold JS, Antonescu CR, Hajdu C, Ferrone CR, Hussain M, Lewis JJ, Brennan MF, Coit DG: *Clinicopathological correlates of solitary fibrous tumours*. *Cancer*, 2002; 94:1057-68.
14. Bassi G, Velasquez S, Orozco R, Garcia-Gallont R.: *An incidental retroperitoneal solitary fibrous tumor*. *Pathology Discovery*, 2014; 2(1):1.
15. Lau M, Foo F, Sissons M, Kiruparan P: *Solitary fibrous tumor of small bowel mesentery: A case report and review of the literature*. *Tumori*, 2010; 96:1035-39.
16. Amo-Takyi BK, Günther K, Peters I, Mittermayer C, Eblenkamp M, Tietze L: *Benign solitary fibrous pleural tumour. Evidence of primitive features and complex genomic imbalances, including loss of 20q*. *APMIS*, 2001; 109:601-06.
17. Debiec-Rychter M, de Wever I, Hagemeijer A, Sciort R: *Is 4q13 a recurring breakpoint in solitary fibrous tumors?* *Cancer Genet Cytogenet*, 2001;131:69-73.
18. Surgpathcriteria.stanford.edu. *Solitary Fibrous Tumor - Surgical Pathology Criteria* - Stanford University School of Medicine [Internet]. 2016 [cited 8 February 2016]. Available from: http://surgpathcriteria.stanford.edu/softfib/solitary_fibrous_tumor/
19. Abraham SC: *Distinguishing gastrointestinal stromal tumors from their mimics: An update*. *Adv Anat Pathol*, 2007; 14:178-88.
20. Joensuu H, Fletcher C, Dimitrijevic S, Silberman S, Roberts P, Demetri G: *Management of malignant gastrointestinal stromal tumours*. *Lancet Oncol*, 2002; 3:655-66.
21. Flint A, Weiss SW: *CD-34 and keratin expression distinguishes solitary fibrous tumor (fibrous mesothelioma) of pleura from desmoplastic mesothelioma*. *Hum Pathol*, 1995; 26(4):428-31.
22. Hanau CA, Markku M: *Solitary fibrous tumour: Histological and immunohistochemical spectrum of benign and malignant variants presenting at different sites*. *Hum Pathol*, 1995; 26 (4):440-49.
23. Nielsen GP, O'Connell JX, Dickersin GR, Rosenberg AE: *Solitary fibrous tumor of soft tissue: A report of 15 cases, including 5 malignant examples with light microscopic, immunohistochemical, and ultrastructural data*. *Mod Pathol*, 1997; 10(10):1028-37.
24. Bouhabel S, Leblanc G, Ferreira J, Leclerc YE, Dubé P, Sidéris L: *Solitary fibrous tumor arising in the mesenter: A case report* *World Journal of Surgical Oncology*, 2011; 9:140.
25. Patriti A, Rondelli F, Gullà N, Donini A: *Laparoscopic treatment of solitary fibrous tumor of the greater omentum presenting as spontaneous haemoperitoneum* *Ann Ital Chir*, 2006; 77(4):351-53.
26. Mazzella A, Izzo A, Amore D, Cennamo A, Cerqua FS, Perrotta

- F: *Single port VATS resection of a sessile solitary fibrous tumour of the visceral pleura. A case report.* Ann Ital Chir, 2015; 86(ePub). pii: S2239253X15024457.
27. Saynak M, Bayir-Angin G, Kocak Z, Oz-Puyan F, Hayar M, Cosar-Alas R, Karamustafaoglu A, Yurut-Caloglu V, Caloglu M: *Recurrent solitary fibrous tumor of the pleura: Significant response to radiotherapy.* Medical Oncology, 2010; 27(1):45-48.
28. Kawamura S, Nakamura T, Oya T, Ishizawa S, Sakai Y, Tanaka T, et al.: *Advanced malignant solitary fibrous tumor in pelvis responding to radiation therapy.* Pathol Int, 2007; 57:213-18.
29. Stacchiotti S, Negri T, Palassini E, Conca E, Gronchi A, Morosi C, Messina A, Pastorino U, Pierotti MA, Casali PG, Pilotti S: *Sunitinib malate and figitumumab in solitary fibrous tumor: Patterns and molecular bases of tumor response.* Mol Can Ther, 2010; 9:1286-297.
30. Hasegawa T, Matsuno Y, Shimoda T, Hasegawa F, Sano T, Hirohashi S: *Extrathoracic solitary fibrous tumors: their histological variability and potentially aggressive behaviour.* Hum Pathol, 1999; 30: 1464-73.

READ-ONLY COPY
PRINTING PROHIBITED