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Giant solitary fibrous tumor of the pelvis: A case report and review of literature

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ABSTRACT

INTRODUCTION: Solitary fibrous tumors (SFTs) are rare spindle cells neoplasms most likely arising from mesenchymal cells. Usually they involve the pleura. Even if extra-thoracic SFTs are rare, lately they are diagnosed with increased frequency.

CASE PRESENTATION: We describe the case of giant pelvic and retroperitoneal neoplasm, a rare solitary fibrous tumor, in a 51-year-old man that was admitted for abdominal pain.

DISCUSSION: On CT a SFT appears usually as a smooth, lobulated mass with occasional calcifications, but the imaging differential diagnosis with other mesenchymal tumors is very difficult, if not impossible.

CONCLUSION: The histological and immune-histochemical features of SFTs are helpful for the differential diagnosis. The malignant potential of this cancer is low, but it is very important to perform an optimized surgery and a close follow up in the patient. We believe that this case is particularly interesting and complex because of the difficulty of predicting the future biological behavior.

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1. Introduction

This work has been reported in line with the SCARE criteria [1].

SFTs were described for first time in the 18th century. They locate mainly in parietal and visceral pleura [2–4], lung parenchyma and pericardium. There are numerous reports that document their extrapleural location but mesenteric localization is extremely unusual [5]. In literature there are also described other location of STFs: pleura [6], orbits [7], tentorium cerebella [8], vagina [9], Liver [10], lesser omentum [11], thyroid [12]. Solitary fibrous tumor is a rare mesenchymal tumor [2,3] which accounted for less than two percent of all soft tissue tumors [13]. SFTs reported in literature mostly occurred in the thoracic cavity and pleura, but there were cases of SFTs involving extrathoracic organs [14,15]. More than 80% of SFTs are benign, asymptomatic and slow-growing tumors [16,17]. However, excessive involvement of important structures may lead to local symptoms [14,18–20]. The diagnosis is usually made by histopathological and immunohistochemical examination of the excised sample. Surgery is considered

as the primary treatment of SFTs, but chemotherapy and radiation therapy could also be indicated in some cases [13]. However, there is no standard treatment for these tumors. Therefore, information from case reports could be useful for clinicians to define better treatment approach for each tumor location.

2. Presentation of case

We describe the case of a 51-year-old man with abdominal pain that was admitted with a giant pelvic and retroperitoneal neoplasm, a rare solitary fibrous tumor. Preoperative diagnosis required complex pathological studies to exclude a high grade sarcoma that was initially suspected (EU, TC). The abdominal scan showed a mass of 18 × 17 × 12 cm (Fig. 1) with a double content: a solid part with a significant enhancement to the contrast and numerous pseudocystic images containing liquid. It was located in meso and hypogastric and reached the pelvis in front of the rectum (Fig. 2). Cranially the origin was at the level of kidney hilum. The CT report suspected a leiomyosarcoma. We sent the patient for operative laparotomy. During surgery we confirmed the presence of massive retroperitoneal tumor extended to the pelvis and rectal tissues. After the identification and dissociation of the ureters from the mass and the detachment from the rearward aortic and caval floor, we performed the complete excision. The tumor appeared to originate from lateral rectal right tissues in close proximity to the right hypogastric axis. We also sent the distal fragments for histopathological examination in order to assess the radical removal of the lesion as it was difficult to evaluate it macroscopically. We marked

Abbreviations: SFT, solitary fibrous tumour; GIST, gastro-intestinal stromal tumour; HPF, high power field; EU, endoscopic ultrasonography.

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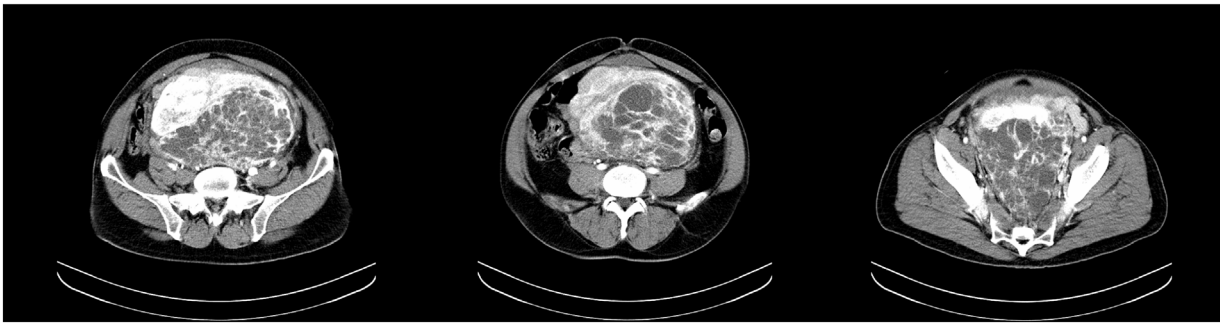


Fig. 1. A pre-operative contrast enhanced abdominal CT scan axial section, shows a giant poli-lobulated mass occupying the pelvis.

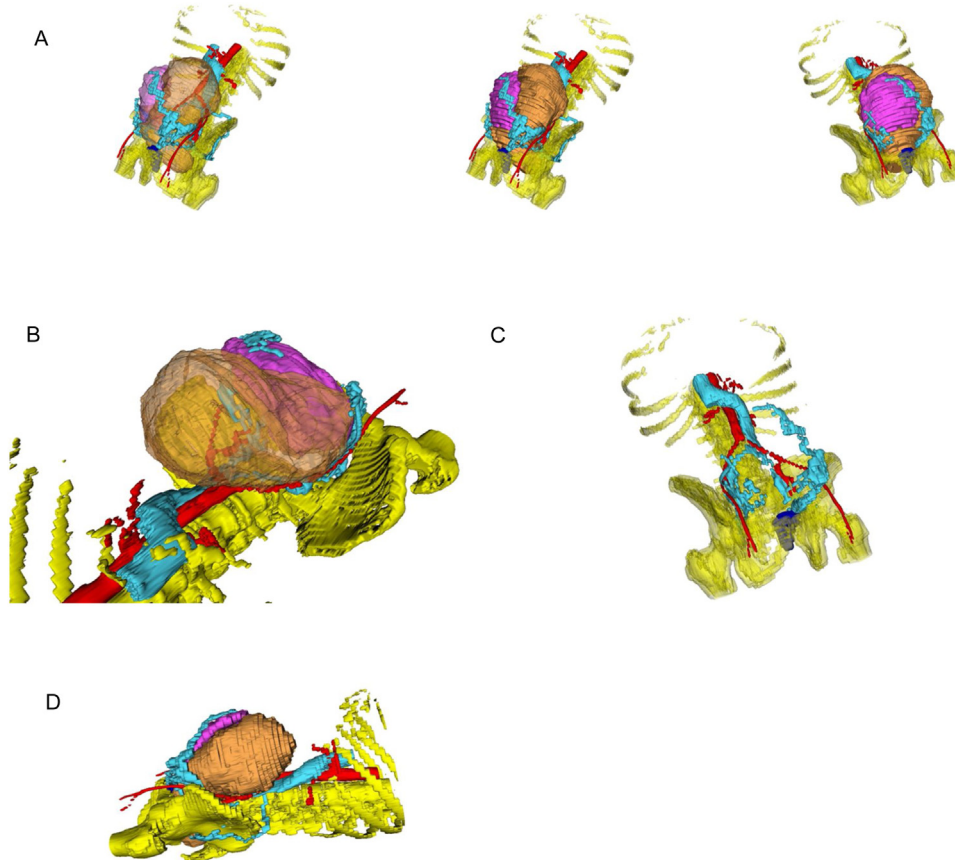


Fig. 2. A CT scan-based 3D reconstruction using Vesalius software for pre-operative planning. Color code: bone yellow, arterial red, vein light blue, orange brown, gall bladder violet, prostate gland dark blue. A Mass and its relationship with vascular structures, right and left lateral view. B Cranial view. C Inferior vena cava congenital abnormality; the vein crosses the midline passing from right to left side. D Collateral left vein arising from inferior vena cava circulation due to compression of the tumour, running posteriorly to left iliac crest.

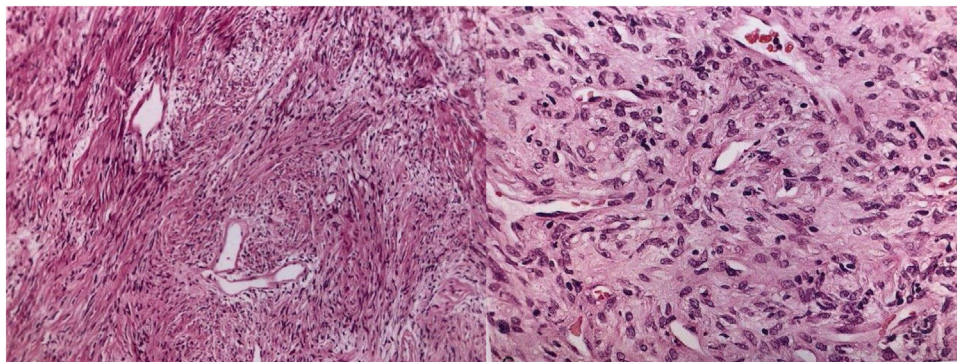


Fig. 3. Mildly atypical fused cell proliferation, organized in bundles, separated by abundant hyaline fibrous matrix, with CD-34 positive immune-phenotype. The proliferation index documented with the Ki-67 antibody was very low, absence of evident atypical mitosis (less than 4/10 HPF).

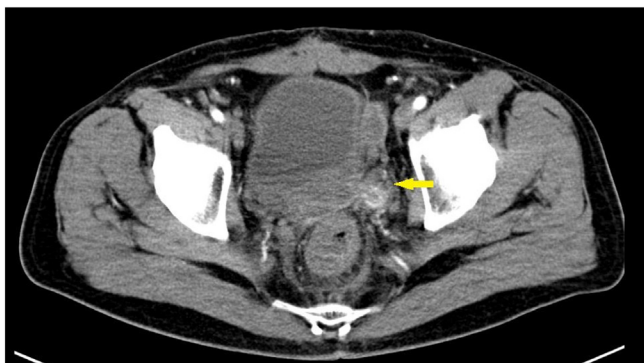


Fig. 4. 3 cm nodular lesion adherent to the left hypogastric internal artery.

with metalclips the presumed stalk for eventual X-ray therapy. We decided to perform an Hartmann procedure as we were concerned of the rectum vascularization as the mass was in close proximity even if without infiltration. We left a drain in the pouch of Douglas then sutured the retroperitoneum. The postoperative course was uneventful. The histopathological examination showed an extrathoracic solitary fibrous tumor with prevalent “hemangioprictoma like” aspects with retroperitoneal localization (Fig. 3).

After three months there was a radiological suspicion of disease recurrence, so we decided to perform an explorative laparotomy. During the surgery we obtained multiple biopsies (Fig. 4). Intraoperative frozen sections were negative for all samples.

We decided to avoid performing the colo-rectal anastomosis to avoid the risk of anastomotic leakage.

In third surgery we performed the recanalization with colo-rectal anastomosis (Knight-Griffen technique) and temporary ileostomy. This maneuver was completed with the use of the colonoscope that showed a regular anastomosis. Rectal stump and bladder were merged together. We dissected bladder wall, through introduction of the finger in the bladder, that allowed the progressive isolation of the rectal stump from the bladder. We did the test filling of the bladder with methylene blue that did not show abnormal spillage. After about two years, the patient returned to our attention for a recto-vesical fistula. He underwent cystoscopy with placement of a left ureteral stent. At the same time, xipho-pubic laparotomy was performed, opening of the Retzius recess, isolation of the bladder with visualization of the recto-vesical fistula located on the posterior bladder surface. The bladder was washed with physiological and methylene blue which allowed to highlight a small leak in the fistula. The fistulous orifice was sutured with detached stitches. The posterior fibrosis of the bladder made impossible the isolation of the patent sub-bladder rectal stump in order to perform an anastomosis to recanalize the large intestine. Therefore, having assessed the non-negligible risks of iatrogenic bladder, prostate and ureteral injury, we opted to stop the attempt to isolate the rectum and we decided to pack a permanent terminal colostomy, disconnection of the ileostomy and removal of the paraileostomy hernial sac. In order to prevent a parastomal incisional hernia we decide to insert a reabsorbable polypropylene prosthesis. The postoperative course was uneventful. No oncological therapies were needed.

After ten year follow up during last outpatient visit, we reported a middle abdominal and peristomal incisional hernia with prolapse. The patient is currently on elective operating list.

3. Discussion

Approximately over 800 cases of SFTs of pleural and extrapleural origin have been reported in literatures recently [31]. Approximately 30% of SFT arise in extra- pleural locations [32].

The malignant variant generally consists of a large tumour (>50 mm in diameter) that is hypercellular and invasive, with nuclear pleomorphism and tissue necrosis. It has a high mitotic index (more than 4 mitoses per 10 HPF) [25,26]. In addition, a size of more than 10 cm and incomplete resection are also positively correlated with local recurrence and metastatic disease [33]. In our case the mass was voluminous, but there were no indexes of malignancy.

No occupying neoplasm was found in follow-up CT. The main differential diagnosis includes gastrointestinal stromal tumor (GIST) [34], synovial sarcoma [35,36], and reactive nodular fibrous pseudotumor (RNFP) of the gastrointestinal tract and mesentery [37].

A haemangiopericytomatous pattern is common in SFTs [21,22]. Immunohistochemically, SFTs commonly show strong and diffuse staining for CD34, bcl2 and vimentin, while epithelial membrane antigen (EMA) and SMA are occasionally expressed [23,24]. SFTs are rarely positive for S100 proteins, desmin, actin and cytokeratins on immunohistochemical analysis [21,22]. The majority of SFTs are histopathologically benign, but about 20% are malignant [25]. A scheme for defining the risk of aggressive behaviour in GISTs based on tumour size and mitotic count has been reached at the National Institutes of Health Workshops [27] and perhaps a similar consensus approach can be adopted for SFTs. A general theory of this particular scheme to predict tumor behavior is that a small tumor with a low mitotic index will have a better prognosis than a large tumor with a high mitotic index. Tumour size has also been shown to be an independent prognostic factor, with a 5-year survival of only 20% in patients with SFTs larger than 10 cm [28]. Positive immunohistochemical staining for CD34 has an important role in the diagnosis of SFT, although it is also found in other tumours [29]. About two thirds of GISTs express CD34 and 95% show positivity for CD117 [27]. SFTs are characteristically CD34 and CD99 positive [30] but lack staining for CD117 [29]. Hence, negative staining with antibody for CD117 is the best marker to distinguish SFT from GIST. SFT should be considered in the differential diagnosis of any mesenchymal lesion arising from the gastrointestinal tract. According to Nomura et al. in order to exclude other CD34 positive tumors, the histological and immunohistochemical features are helpful, where the presence of immunoreactivity for CD34 and absence of immunoreactivity for CD 117, desmin, and S-100, strongly favors a definite diagnosis of SFT.

4. Conclusion

SFTs are rare. The pleura is the most common site of development. Extrathoracic SFTs are not common [2–4]. Ours is the first case described in literature of a SFT arising from pelvis. They are usually located in pleura and peritoneum. The pre-operative diagnosis is difficult because clinical symptoms, physical examination and imaging (US, CT, MRI) are not diagnostic. The appearance of an SFT on CT scan is usually a smooth, lobulated mass with occasional calcifications [37], but the differential diagnosis with other mesenchymal tumors is very hard, if not impossible. The correct diagnosis requires pathological and immunohistochemical examination of the resected surgical specimens [4]. The positive immunoreactivity for CD34 in this tumors confirmed the diagnosis, and the negativity for CD-117 distinguish SFT from GIST. There is also a negativity for S-100. Resection was required. Prognosis for SFTs is usually good after complete resection [2–4]. Recurrence and metastases may be related to rare aggressive histological features including necrosis, greater than four mitoses/10 high power fields, hypercellularity, nuclear atypia [2]. Most of SFTs have a benign course, but the 20.8% have the potential to recur or metastasize. At the moment it's not possible to define each tumor's behavior in the future, so it's crucial a long term follow up for all patients

[2–4]. Histopathological examination of the voluminous mass in our case report, despite its size and location, showed no histological characters of certain malignancies, with mitotic proliferation index less than 4/10 HPF, absence of tumor necrosis and cellular pleomorphism failure. We believe that this case is particularly interesting and complex because of the difficulty of predicting the future biological behavior.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

Exempt from ethical approval.

Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images.

Author contribution

GP, CG and LC acquired the data, drafted and wrote the article. GC, MDD and PC critically revised the article. CG and PC performed the surgical operations. FB provided the 3D reconstruction images. All authors read and approved the final manuscript.

Registration of research studies

1. Name of the registry: none
2. Unique identifying number or registration ID: none
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): none

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