A 64-year-old woman with primary synovial sarcoma of the abdominal wall

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**Abstract**

Synovial sarcoma (SS) usually affects joints, bursae, and tendons of extremities and is very infrequent in the head and neck, abdomen, thorax, prostate and kidney, skin, blood vessels, and nerves. Primary intra-abdominal SS is exceeding uncommon and has non-specific symptoms or compress surrounding structures. The diagnosis is a challenge, and histopathological and immunohistochemical studies must confirm the hypothesis. We report the case of SS that has origin from peritoneal structures and a longstanding unsuspected course. The patient was a 64-year-old woman who claimed chronic pain in the left iliac fossa, without additional symptoms. She related laparoscopic oophorectomy, cholecystectomy, and abdominal hysterectomy in the previous three decades. There was neither local invasion nor lymph nodal, vascular or neural invasion, and her surgical treatment by open abdominal procedure was uneventful. The herein reported case aims to enhance the index of suspicion.

**Keywords:** intra-abdominal, primary tumor, soft tissue tumor, synovial sarcoma.

**Introduction**

Synovial sarcoma (SS) is an intermediate-to-high-grade malignant mesenchymal tumor more often affecting soft tissues in the extremities and related with synovial structures, joint capsules, bursae and tendon sheaths of children and adults often with a longstanding course. Infrequent sites are head and neck, abdominal wall, mediastinum, lung and pleura, prostate and kidney, skin, blood vessels, and nerves [1–11]. Primary intra-abdominal SS is very uncommon, causing non-specific symptoms or compression of adjacent structures [1–11]. Worthy of note is that the case reports of SS having origin from peritoneal structures are even rarer [2, 4–6, 10, 11]. Histopathological (HP) and immunohistochemical (IHC) studies of tumor samples are mandatory to establish the final diagnosis. SS may evolve unsuspected and should be considered an additional hypothesis during the investigation of intra-abdominal masses.

**Aim**

The present case report aims to enhance the awareness of this challenging malignant tumor.

**Case presentation**

A 64-year-old woman reported persistent pain located in the left iliac fossa, which had been more intense for six months. She had no intestinal and urinary changes or weight loss, was hypertensive using Losartan (50 mg/day), ex-smoker (45 packs/year), and non-alcoholic. Her surgical history was left laparoscopic oophorectomy 27 years ago, total abdominal hysterectomy 26 years ago, and laparoscopic cholecystectomy 10 years ago. It was confirmed the diagnosis of ovarian endometriosis and the absence of any malignancy. Physical examination showed a good general condition, body mass index (BMI) 37.8 kg/m² and normal clinical parameters; a flabby, painful abdomen with a slight resistance in the left iliac fossa; and normal intestinal sounds. Laboratory tests, including tumor markers, revealed normal data – hemoglobin: 15.9 g/dL; leukocytes: 9170/mm³; platelets: 204 000/mm³; urea: 49 mg/dL; creatinine: 1.2 mg/dL; Na+: 137 mEq/L; K+: 5.1 mEq/L; International Normalized Ratio (INR): 1.06; activated partial thromboplastin time (APTT): 27.6 s; C-reactive protein (CRP): 1.08 mg/L; carbohydrate antigen (CA) 19-9: 7.9 IU/mL; carcinoembryonic antigen (CEA): 1.3 ng/mL; CA 125: 6.6 IU/mL; CA 15.3: 14.3 IU/mL; beta-human chorionic gonadotropin (β-HCG): 6.3 mIU/mL; alpha-fetoprotein: 2.6 ng/mL. Abdominal contrast-enhanced computed tomography (CT) (July 12, 2019) showed a solid expansive image with regular contours and slight post-contrast enhancement in the left pelvic cavity and iliac fossa, measuring 5.8×5.3×4.7 cm (Figure 1). Upper digestive endoscopy and colonoscopy showed no changes. Magnetic resonance imaging (MRI) of the abdomen and pelvis (September 22, 2019) revealed a heterogeneous solid mass in the anterior abdominal wall of the left iliac fossa, measuring approximately 6.3 cm, with intense post-contrast enhancement displacing the intestinal loops, and lymph node enlargement in the external iliac chain on the left (Figure 2). The image-guided pelvic mass biopsy (November 19, 2019) showed tissue proliferation with a fusocellular pattern of indeterminate histogenesis. The IHC study found vimentin (V9) positive and Ki67 (MIB-1) positive (10%); however,
due to the scarcity of the sample, the diagnosis was considered inconclusive. The direct exam of the intra-abdominal mass by video laparoscopy (February 19, 2020) showed it loosely adhered to the peritoneum (Figure 3A) and in close contact with the lower epigastric vessels. The resection of the posterior aponeurosis of the rectus abdominis muscle and the clipping of the lower epigastric vessels were performed after the opening of the peritoneum with safety margins (Figure 3, B and C). It was not possible to approach the peritoneum to cover the exposed musculature. The surgical specimen (Figure 3, D and E) was removed by enlarging the umbilical incision and the anatomopathological study characterized a poorly differentiated fusocellular malignancy with 10 mitoses/10 high-power fields (HPFs) (Figure 4). No angiolymphatic embolization or perineural invasion was detected. The IHC evaluation showed positivity for pan-cytokeratin (CK) AE1/A3, B-cell lymphoma 2 (Bcl-2), calretinin, cluster of differentiation (CD)56, CD99 (focal), Ki67 (in 1% of cells), and CD34 (in vessels); and negativity for CD34 and CK7 in tumor cells (Figure 4).

These data associated with the pattern of the HP picture allowed to establish the diagnosis of intra-abdominal SS. The hospital discharge was on the first postoperative week, and she is on outpatient follow-up. There was no hernia at the surgical site and the pain at the resection site was claimed only for a single month.

Figure 1 – (A and B) Images of abdominal CT with contrast showing a solid expansive image (arrows) with regular contours and slight post-contrast enhancement in the left pelvic cavity and iliac fossa. CT: Computed tomography.

Figure 2 – (A and B) Images of abdominal and pelvic MRI study revealing a heterogeneous solid mass in the anterior wall of the left iliac fossa (arrows), with intense post-contrast enhancement and displacement of intestinal loops. MRI: Magnetic resonance imaging.

Figure 3 – Video laparoscopic view of the intra-abdominal mass appearing loosely adhered to peritoneum (A); images of the abdominal cavity after resection of the posterior aponeurosis of the rectus abdominis muscle (B and C); and gross aspects of the removed surgical specimen (D and E).
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Figure 4 – (A–C) Photomicrography of the surgical tumor sample consistent with a poorly differentiated fusocellular tumor, which presented 10 mitoses/10 HPFs. HE: Hematoxylin–Eosin; HPFs: High-power fields.

Figure 5 – (A–F) Photomicrography of the IHC evaluation of the tumor sample with positivity for CD99 (A and B), Bcl-2 (C and D), calretinin (E), and CD56 (F). Bcl-2: B-cell lymphoma 2; CD: Cluster of differentiation; IHC: Immunohistochemical.

## Discussions

The SS corresponds up to approximately 10% of the soft tissue sarcomas, predominantly affect synovial structures in the extremities (up to 88%) of young adults, and more rarely derive from a sarcomatoid differentiation of peritoneal mesothelial cells simulating synovial tissue [1, 3–11]. Based on the pathological and IHC features there are three types of SS: (i) monophasic, with exclusively of spindle-shaped cells; (ii) biphasic, with epithelial and spindle cells; and (iii) poorly differentiated, with rhabdoid pattern [1–11]. Molecular biology resources can reveal specific gene rearrangement consistent with translocation of chromosomes X and 18, resulting in three alternative fusion products of the SS18 gene (SYT) with the SSX1, SSX2 or SSX4 gene [1–11]. Primary intra-abdominal SS is a very uncommon condition, which constitutes a challenging diagnosis due to non-specific symptoms. The differential diagnosis includes carcinosarcoma, chondrosarcoma, choriocarcinoma, desmoid tumor, endometriosis, deep-seated fibromatosis, fibrosarcoma, gastrointestinal stromal tumor (GIST), histiocytoma, leiomyosarcoma, mesothelioma, rhabdomyosarcoma, schwannoma, small round cell tumor, and the very rare solitary fibrous tumor (SFT) [1–3, 7, 9–12]. Immunohistochemistry can be confirmatory because peritoneal mesothelial cells show mesenchymal and epithelial features and express filaments typical of mesoderm (vimentin) and epithelium (CKs) [1–7, 9–11]. The worst outcomes, including recurrences and metastases of intra-abdominal SS, are associated with a diameter over 5 cm, young age, high mitotic count, retroperitoneal and pelvic sites, and vascular or neural invasion [1–3, 7–9]. Because the recurrence and metastasis are frequent, the first option of treatment is en bloc resection, ≥1 cm margins and a long follow-up [1–3, 5–11]. The management may involve pre- or post-operative radiotherapy and neoadjuvant or adjuvant chemotherapy; the more often utilized drugs include Doxorubicin, Ifosfamide plus Mesna, Pazopanib, and Trabectedin [1, 3, 5–10]. The control of the metastatic disease employs palliative chemotherapy and radiotherapy,
and lung is the most frequent site; less commonly the implants of SS are described involving lymph nodes, liver, and bones [3, 6–11].

The 64-year-old woman herein described had a poorly differentiated and fusocellular primary intra-abdominal SS of peritoneal origin, with 10 mitoses/10 HPFs, and the absence of local invasion or distant implants. An unspecified chronic pain in the left iliac fossa was the unique symptom of the unsuspected malignant tumor. Worthy of note, her clinical course could be mistaken as an eventual tardive complication of laparoscopic oophorectomy, cholecystectomy, and hysterectomy performed in the previous three decades. A long undetected evolution favored the tumor growing up to more than 6 cm of diameter that may predispose to a poor prognosis. The neoplasm was locally restricted, although measuring over 5 cm and with a mitotic activity index of 10 mitoses/10 HPFs. She underwent an open surgical procedure after the results of a preoperative routine including abdominal CT and MRI, in addition to the laparoscopic evaluation with biopsy of the tumor. An intra-abdominal SS vimentin (V9), Ki67 (MIB-1), pan-CK AE1/AE3, calretinin, and CD56 immunopositivity was confirmed. Her immediate postoperative course was uneventful and after the hospital discharge, she is on outpatient control.

In this scenario, we have reviewed the case studies published in 2015 by Val-Bernal et al. about SFTs or hemangiopericytomas of the mesentery, an exceedingly rare condition that may pose diagnostic pitfalls related to intra-abdominal SS [12]. A 61-year-old man with an incidental diagnosis was asymptomatic, with previous malignant melanoma in the inferior limb and the sentinel node free of tumor. His abdominal CT of control detected a 3 cm solid heterogeneous formation in the jejunal mesentery, which was removed by laparoscopy. The IHC study showed: diffuse positivity for CD34, CD99, and calponin; focal positivity for Bcl-2 and epithelial membrane antigen (EMA), and Ki67 labeled 4% of cells; and negativity for CD117, DOG1, CD31, CD56, D2-40, human melanoma black 45 (HMB45), alpha-smooth muscle actin (α-SMA), desmin, pan-CK AE1/AE3, neuron specific enolase (NSE), collagen IV, S100 protein, and Melan A. A 32-year-old man with longstanding abdominal pain, distention and hematuria had a solid mass of 13 cm detected by tomographic images of the mesosigmoid space. The IHC study revealed: positivity for CD34, CD99, and Bcl-2 and focal positivity for calponin and alpha-smooth muscle actin (α-SMA); and negativity for CD117, DOG1, CD31, pan-CK AE1/AE3 and EMA [12]. Worthy of note were the reviewed data from the 13 previously published cases of SFT in the whole world. Moreover, the authors commented on the main differences between the SFT and the monophasic variant of SS (MSS) with hemangiopericytoma-like areas. They emphasized the lack of tumor cells with expression of CD34 in MSS [12]. If considering their useful practical comments and the exclusive positivity of CD34 in the vessels of the tumor one could speculate the hypothesis of primary intra-abdominal MSS in the case study herein described.

Concerns about the present study include the lack of evaluation to detect specific gene rearrangements. Almost all SS contain the translocation t(X;18)(p11.2;q11.2) involving the SYT gene on chromosome 18 and the SSX gene on the X chromosome (SSX1, SSX2, or SSX4) [1–11]. So, the analysis by reverse transcription–polymerase chain reaction (RT–PCR) or fluorescence in situ hybridization (FISH) can be useful for a more targeted effective management, with a better outcome [1, 8]. However, high-cost tools may not be easily accessible to everyone in the low- and medium-income countries; and besides, the scarce number of reported SS makes it difficult to establish consensus protocols for treatment.

Conclusions

The SS most commonly occurs in the extremities, in special the knee, and rarely originates within the abdomen and pelvis. This tumor predominates in middle age and often becomes voluminous, with difficulty excision and local recurrences. Imaging studies are useful tools for diagnostic hypothesis, for the staging, and post-treatment control. The SS diagnosis confirmation depends on the consistent IHC findings. The best option for local management of this malignancy is radical surgical excision, with a minimum follow-up period of 10 years due to the possibility of late recurrence and metastasis. The intra-abdominal SS herein described in a 64-year-old woman was successfully treated by open surgical procedure. Despite the inherent weaknesses of a single case study, this might contribute to better knowledge about this rare malignant condition.

Conflict of interests

The authors had full freedom of manuscript preparation and there was no potential conflict of interests.

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