



# Closed Perforation of Small Bowel Caused by Primary Intra-abdominal Synovial Sarcoma

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## Abstract

Synovial sarcoma is a rare and aggressive tumor, typically found in the extremities, with intra-abdominal cases being exceptionally uncommon. We present a 47-year-old male with abdominal pain and significant weight loss, whose imaging revealed a retroperitoneal mass originating from the duodenojejunal flexure. Computed tomography demonstrated a lobulated mass with heterogeneous enhancement, cystic and necrotic areas, and perilesional lymphadenopathy. Despite extensive imaging, definitive diagnosis required histopathological confirmation. Intraoperatively, a closed-loop perforation and vascular invasion rendered the tumor unresectable. Given the extreme rarity and diagnostic challenges of intra-abdominal synovial sarcoma, a multidisciplinary approach is essential. Comprehensive radiological evaluation, assessing mass morphology, vascular involvement, resectability, and metastatic potential, is crucial for accurate diagnosis and treatment planning. This case highlights the importance of advanced imaging in managing rare abdominal malignancies.

## Keywords

- ▶ intra-abdominal sarcomas
- ▶ soft-tissue mass
- ▶ synovial sarcoma

## Introduction

Synovial sarcoma (SS) is a rare and aggressive tumor.<sup>1</sup> In a study by Ferrari et al, 1,531 cases of SS were diagnosed, of which 68.7% were seen in the extremities, 15.7% in the trunk, 6.3% in the head and neck, 5.3% intrathoracic, 2.2% others, and 1.8% in the intra-abdominal region.<sup>2</sup>

Radiologically, SS typically appears as a heterogeneous soft-tissue mass containing variable components such as cystic, necrotic, or hemorrhagic areas on ultrasonography (US). Computed tomography (CT) and magnetic resonance imaging (MRI) play a critical role in tumor characterization and staging by providing detailed assessment of the internal

architecture of the mass and its relationship to the adjacent anatomical and vascular structures.<sup>3</sup>

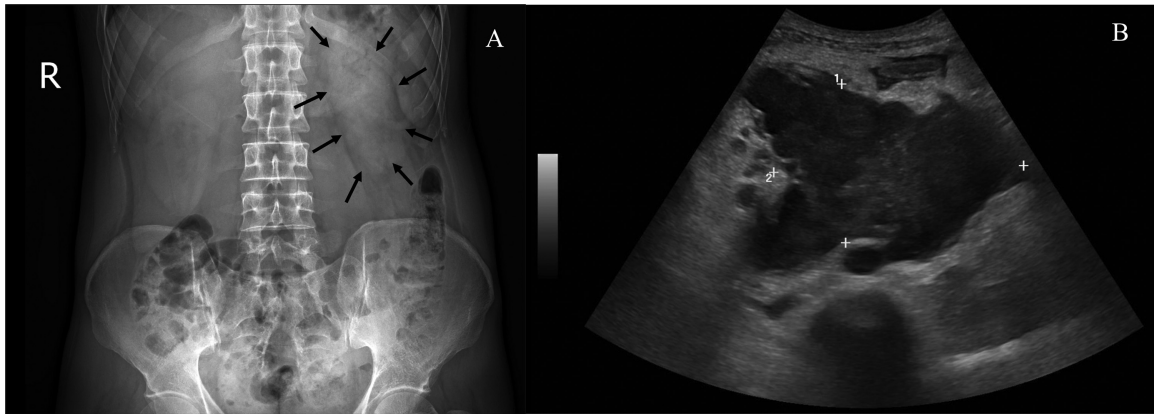
## Case Report

A 47-year-old male with a history of smoking presented with abdominal pain, gas, and significant weight loss. Laboratory tests revealed a hemoglobin level of 9.6 g/dL, with no other significant pathological finding in other laboratory tests. Radiography, US, and CT images showed a mass originating from the level of the duodenojejunal flexure to the proximal jejunal loops, approximately 8 × 7.5 cm in size with an exophytic extension to the mesentery (▶ **Figs. 1** and **2**).

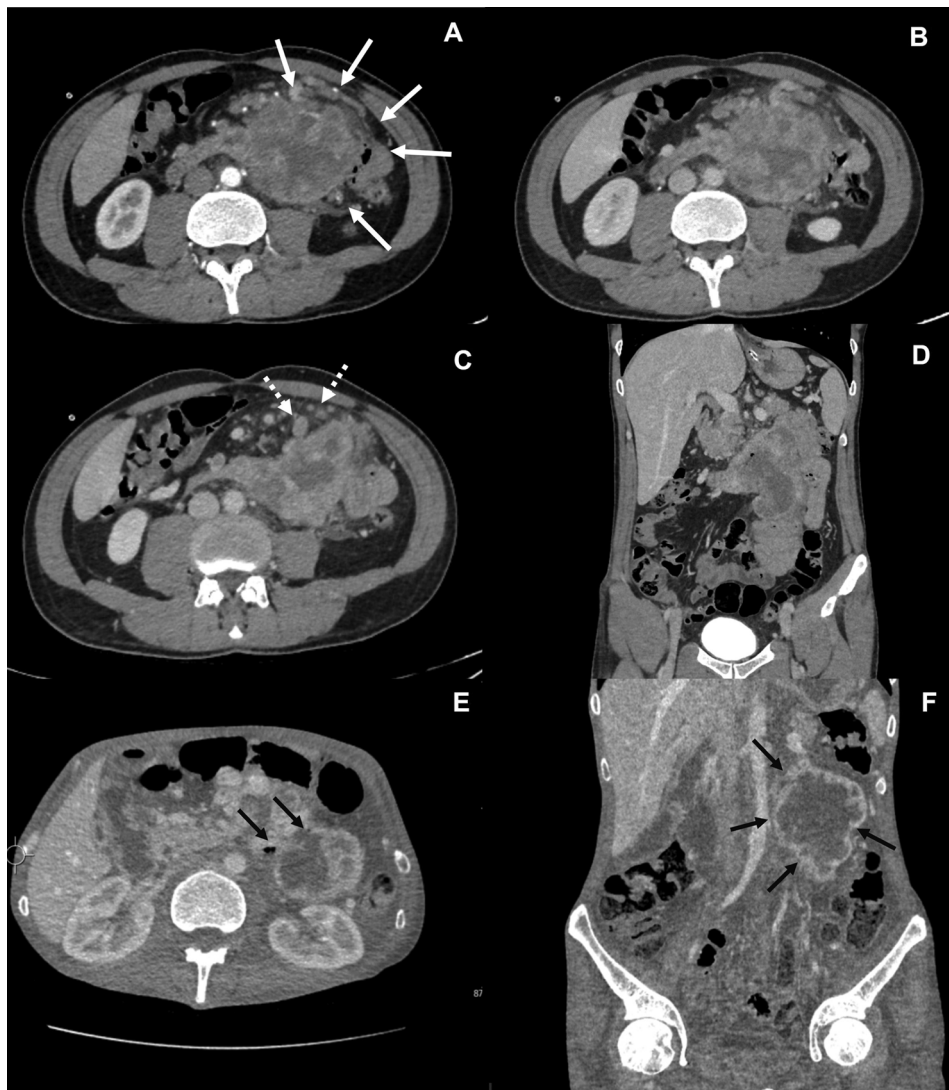
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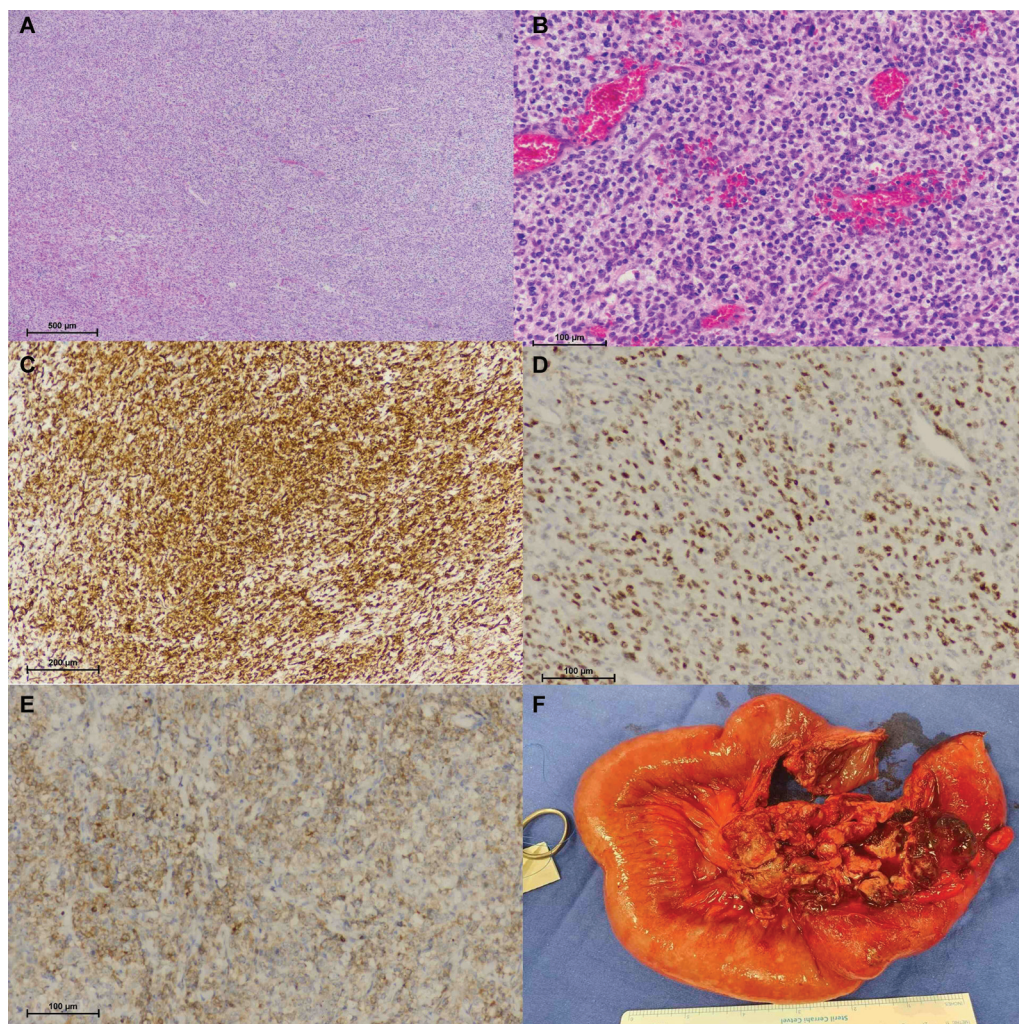
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**Fig. 1** Radiological images of the tumor (X-ray and ultrasonography). (A) Abdominal X-ray. An opacity with indistinct borders indicated by arrows is noticeable in the upper left quadrant of the abdomen (arrows). (B) Ultrasonography imaging revealed a lobulated contoured hypochoic heterogeneous solid appearance (“+” signs indicate that there are two dimensions of the lesion).



**Fig. 2** Preoperative arterial-venous phase CT images (A–D) and postoperative 4-month follow-up contrast-enhanced CT images (E, F). (A) Arterial phase and (B) portal phase contrast-enhanced CT images: A mass,  $\sim 8 \times 7.5$  cm in size, extends from the level of the duodenojejunal flexure to the proximal jejunal loops, involving a long segment. It exhibits exophytic extension into the adjacent mesentery (arrows). (C) Axial and (D) coronal images of the mass with multiple lymphadenopathies in the nearby mesentery (dashed arrows). (E) Axial and (F) coronal images of the mass lesion containing cystic-necrotic areas (arrows). Postoperative imaging conducted in the fourth month showed an increase in tumor size and internal necrosis. CT, computed tomography.



**Fig. 3** Histologic features of the tumor. Sections from the mass located in the serosa of the small intestine revealed a tumor proliferation rich in vascular structures, surrounded by a relatively well-defined fibrous capsule. (A) Tumoral proliferation rich in vascular structures, with narrow cytoplasm and oval nuclei (4 ×, hematoxylin and eosin). (B) Tumor cells with spindle-shaped nuclei and marked pleomorphism (20 ×, hematoxylin and eosin). (C) Immunohistochemical positivity for vimentin (10 ×). (D) Immunohistochemical nuclear positivity for TLE-1 (20 ×). (E) Immunohistochemical positivity for Bcl-2 (20 ×). (F) Macroscopic image.

Gastrointestinal stromal tumor, adenocarcinoma, sarcoma, and lymphoma were considered for differential diagnosis of the mass.

Surgical exploration revealed a mass originating from the fourth part of the duodenum. It had closed perforation resulting in the infiltration of the small intestinal mesentery and aorta. Therefore, it was nonresectable due to the extensive vascular invasion. Duodenojejunostomy and gastrojejunostomy were performed, and the mass was partially resected and sent for histopathological evaluation (→Fig. 3). Even in the absence of closed perforation, surgical resection was not considered feasible due to the tumor's invasion of the abdominal aorta. Based on the immunohistochemical and molecular findings, the case was evaluated in favor of SS and chemotherapy was planned (→Table 1). Due to the risk of potential strictures and ileus, radiation therapy was not recommended at this stage. At the postoperative fourth month follow-up, an increase in size was observed in both the solid component

and the internal necrosis, suggesting progressive disease (→Fig. 2E, F). The patient died in the fifth postoperative month.

## Discussion

A search of the PubMed database between 2000 and 2024 identified 55 cases of intra-abdominal SS. Only five of them were of duodenal origin. Our case of SS of duodenal origin, which is rare in terms of both location and incidence, is the only known case in the literature that causes closed perforation. SS appears as hypoechoic and heterogeneous solid mass. US-guided biopsy of the lesion is feasible and frequently utilized.<sup>3</sup> Intrabdominal SS typically appears on CT as lobulated, heterogeneously enhancing, solid masses. Cystic, necrotic, hemorrhagic areas, and calcifications can be observed. The lesions often demonstrate prominent vascularity following contrast administration.<sup>3</sup> MRI frequently reveals a multilobulated and infiltrative architecture, supporting the

**Table 1** Treatment Protocol

Order of administration	Dose	Method of administration	Day 1	Day 2	Day 3
1. Doxorubicin <sup>a</sup>	60 mg/m <sup>2</sup>	150 cc 5% dextrose or normal saline to be infused IV over 15 minutes	+		
2. Ifosfamide	2,500 mg/m <sup>2</sup>	1,000 cc 5% dextrose to be infused IV over 4–6 hours	+	+	+
	500 mg/m <sup>2</sup>	15 minutes before ifosfamide	+	+	+
3. MESNA	1,000 mg/m <sup>2</sup>	With ifosfamide	+	+	+
	1,000 mg/m <sup>2</sup>	After ifosfamide in 1,000 cc 5% dextrose over 12 hours	+	+	+

<sup>a</sup>If Adriamycin is given at 75 mg/m<sup>2</sup>, G-CSF support should be provided.

Note: The “+” symbol in the table indicates the days on which the medication or treatment is administered. For example, a “+” under “Day 1” means that the corresponding drug or treatment is given on that day. If there are “+” marks for multiple days, it signifies that the drug or treatment will be administered on those specific days, as outlined in the table.

locally aggressive nature of the tumor. SS show iso- or slight hyperintensity relative to muscle on T1-weighted images<sup>4</sup> and a characteristic “triple sign” (hypo-, iso-, and hyperintense areas) relative to fat tissue on T2-weighted images.<sup>5</sup> The solid parts show rapid and intense enhancement on dynamic contrast-enhanced MRI.

On MR spectroscopy, an elevated choline peak may be detected. MR angiography provides a three-dimensional evaluation of vascular structures, particularly in preoperative surgical planning.<sup>5–7</sup>

Intra-abdominal sarcomas often metastasize to the liver and lungs, especially the patients with tumors larger than 5 cm have a higher risk for lung metastasis. Therefore, total body imaging is required.<sup>7</sup> The primary treatment for SS typically involves wide local excision with safe margins, often accompanied by radiotherapy and chemotherapy.<sup>5</sup>

#### Ethical Approval

None available.

#### Patient's Consent

The patient was fully aware of the publication and gave written informed consent as per ethical principles outlined in the Declaration of Helsinki.

#### Conflict of Interest

None declared.

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