

Introduction

Pancreatic Leiomyosarcoma

- Very rare; most common mesenchymal tumour of pancreas yet <1% of primary pancreatic malignancies; peak incidence in 5th decade ¹⁻³
- Likely develops from smooth muscle cells in blood vessels or ducts ²
- Presents with abdominal pain, weight loss, abdominal mass on imaging ^{2,3}
- 5-year mortality is 77.8%; distant metastasis (present in 37.5% of cases) is the strongest prognostic factor, conferring 5x higher mortality rate ³
- Treatment for localized tumour is radical surgical resection; management becomes complex in metastatic disease (chemotherapy, palliative care) ^{3,4}

Chest Wall Leiomyosarcoma

- Rare; 1-4% of soft tissue sarcomas originating from the chest wall ⁵
- Usually present as palpable, painful, rapidly growing masses ⁶

Methods: Clinical History

- Patient presented to family physician with abdominal pain; family history for father (unknown cancer), sister and mother (breast cancer)
- Ultrasound identified pancreatic mass, confirmed on PET; core biopsy diagnosed pancreatic leiomyosarcoma, conventional type, grade 2
- Treated with 7 cycles of neoadjuvant chemotherapy as per the STRASS-2 (neoadjuvant chemotherapy vs surgery alone for high-risk retroperitoneal sarcoma) protocol ⁷
- Followed by distal pancreatectomy, (**Fig. 1**) retroperitoneal lymph node dissection and gastrojejunostomy
- Follow-up exam revealed palpable, mobile, superficial chest wall mass
- Ultrasound-guided biopsy confirmed leiomyosarcoma with spindle cell abnormality; mass was subsequently excised

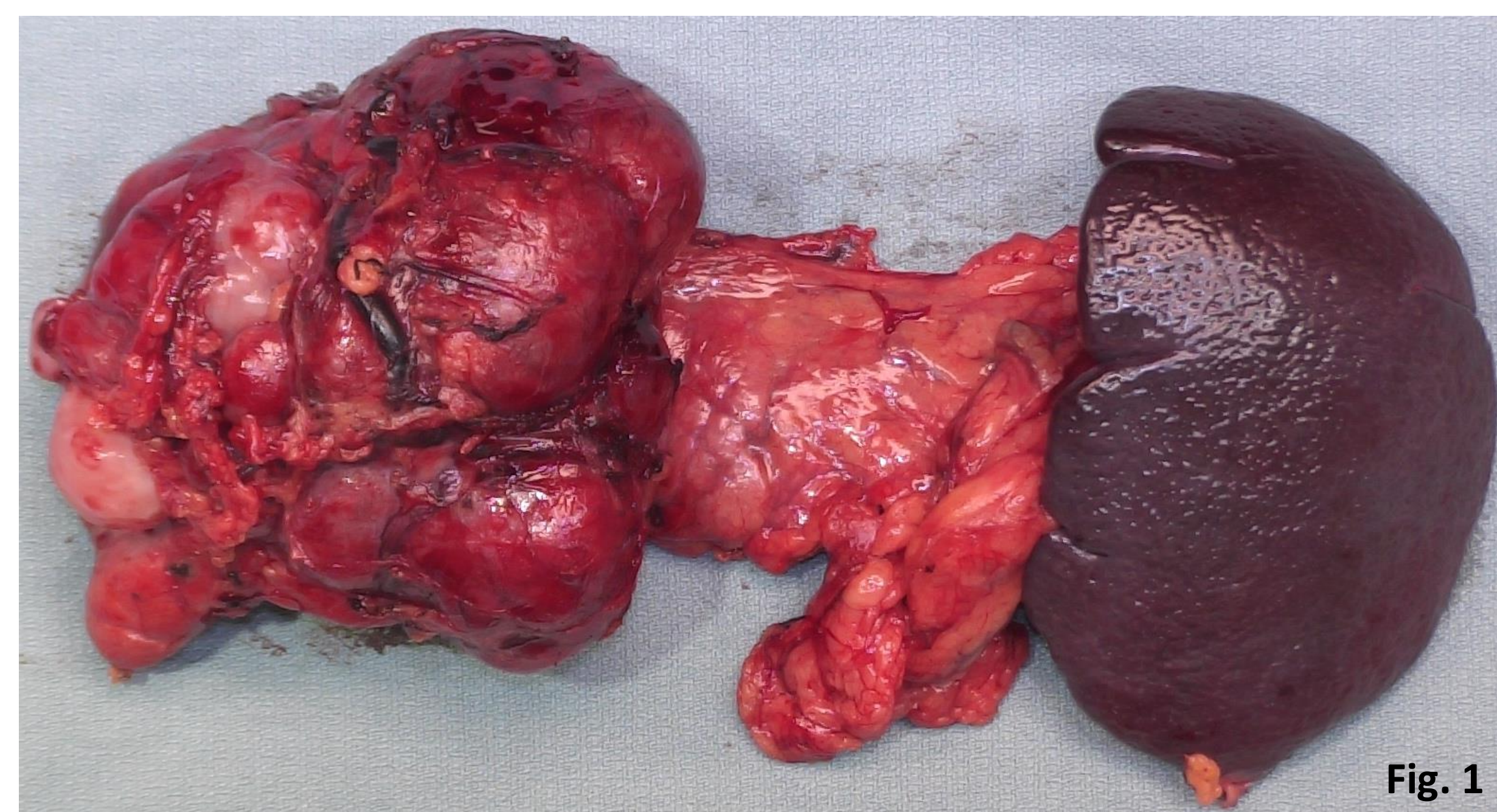


Figure 1. Specimen of distal pancreatectomy, demonstrating pancreatic mass (left)

Methods: Gross Examination

- Pathologists' Assistant serially sectioned and photographed both specimens (**Fig. 2, Fig. 3**).

Distal Pancreatectomy Specimen

- Tan, fleshy, hemorrhagic, lobulated mass (9.2 x 8.6 x 7.1 cm) arising from anterior pancreatic surface
- Mass is solid with areas of necrosis (25%) and does not *grossly* invade pancreatic parenchyma

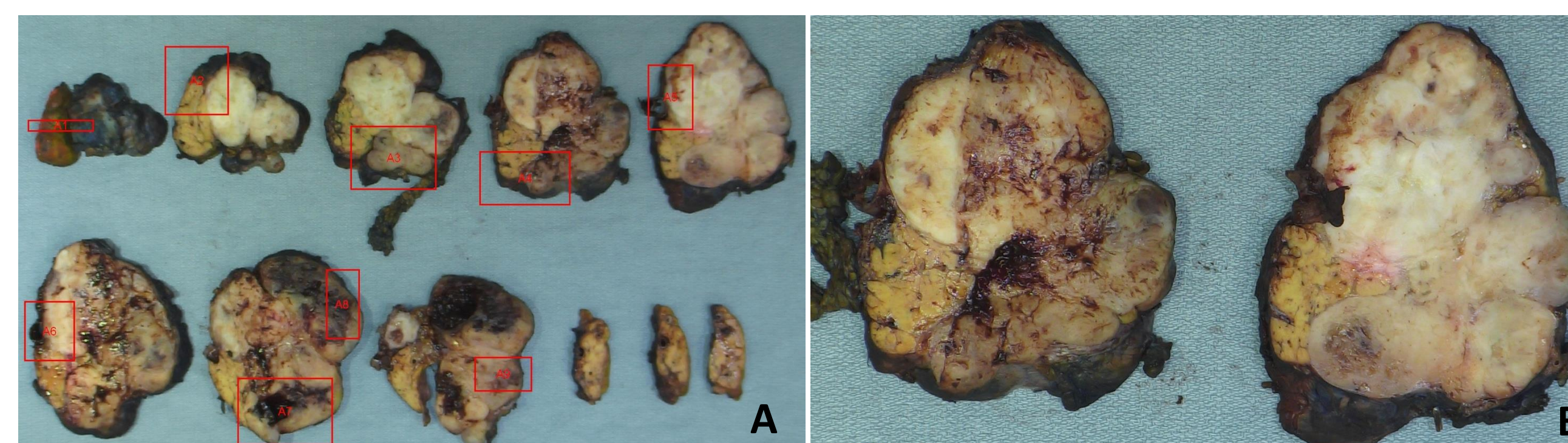


Figure 2. Serial sections of involved portion of the distal pancreatectomy specimen. A. Demonstrates the tan, fleshy, hemorrhagic, lobulated cut surface of the mass within the pancreatic parenchyma and the mapping of the sections submitted for histologic assessment. B. Enlarged sections of the mass.

Chest Wall Excision Specimen

- Circumscribed, firm, pale-tan nodule (1.5 x 1.3 x 1.0 cm) within subcutaneous adipose tissue

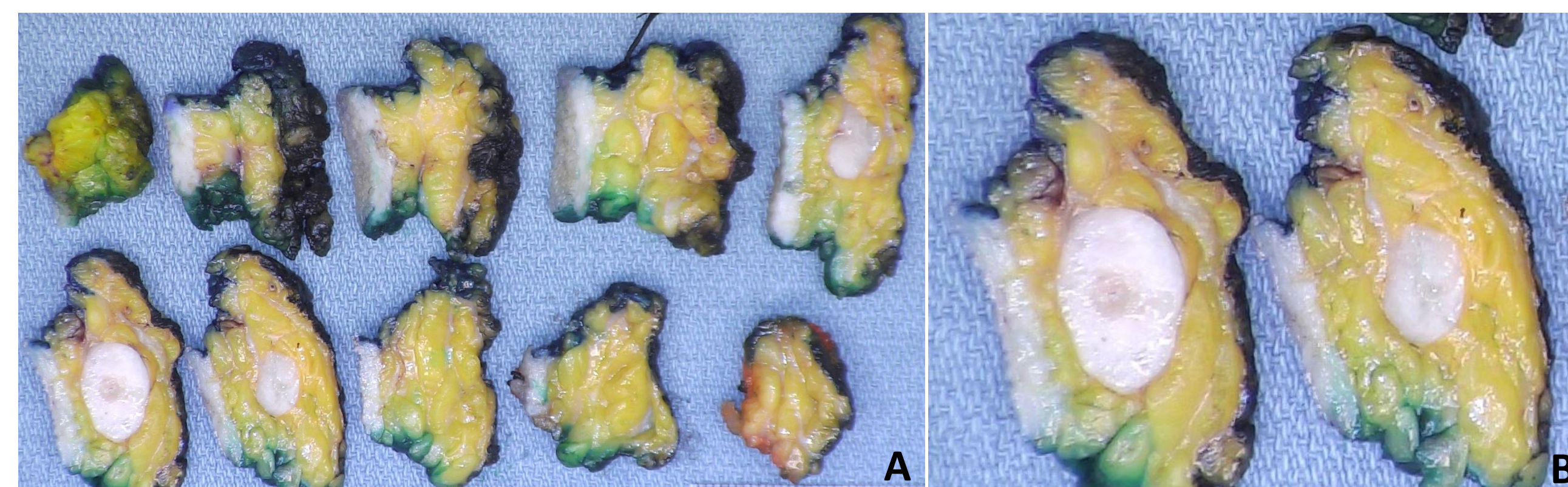


Figure 3. Serial sections of specimen: skin ellipse with underlying chest wall subcutaneous tissue. A. Demonstrates the firm, pale-tan cut surface of the nodule within the subcutaneous adipose tissue. B. Enlarged sections of the nodule.

Case Study: Results

Histology: distal pancreatectomy specimen

- Leiomyosarcoma microscopically involving pancreatic parenchyma (**Fig 4.**)
- Unidentifiable histologic grade due to post neo-adjuvant therapy, ~20% treatment effect
- Necrosis (<5%), hyalinization, hemorrhage, cystic change present
- No lymphatic/vascular invasion identified, negative lymph nodes and resection margins
- IHC staining negative for ER, PR, WT1, MDM2 (previous biopsy diffusely positive for actin, desmin, calretinin, negative for S100)

Histology: chest wall excision specimen

- Leiomyosarcoma with mitotic rate of 10/10 high-power field
- No lymphatic or vascular invasion identified
- Negative resection margins
- IHC positive for actin, desmin

Case Study: Results (cont.)

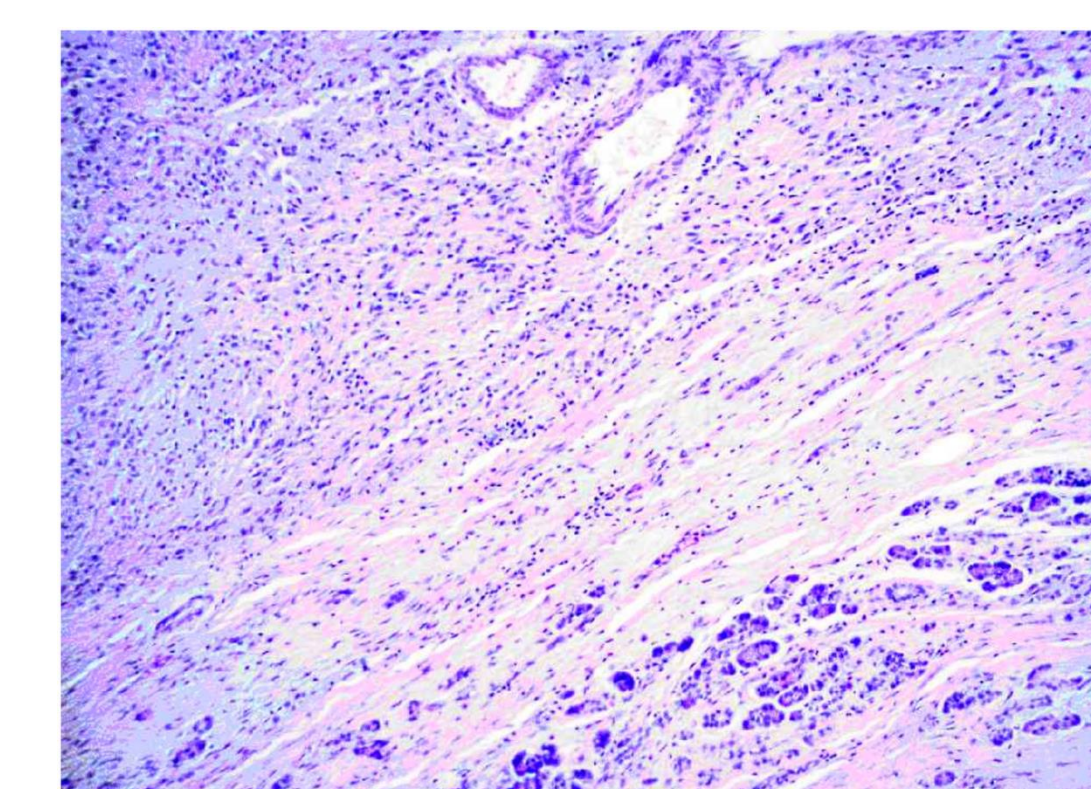


Figure 4. Representative histology of pancreatic leiomyosarcoma showing the interface between tumour (left) and compressed adjacent pancreatic parenchyma with associated fibrosis (H&E, x100). Image reproduced from Devaux PG, Aranha GV, Yong S. Leiomyosarcoma of the pancreas. HPB 2001;3(2):175-177. ⁸

Diagnosis

- Primary leiomyosarcoma involving pancreatic parenchyma
 - Tumor did not invade peritumoral adipose tissue and was staged as **ypT2 pN0** per the 8th edition of the American Joint Committee on Cancer (AJCC) ⁹
- Primary leiomyosarcoma involving subcutaneous adipose tissue

Discussion & Grossing Considerations

- Inconclusive whether the chest wall leiomyosarcoma represents a second primary or metastasis from the pancreatic leiomyosarcoma
- Genetic testing for Li Fraumeni Syndrome currently underway due to suspicion of two concurrent but independent primary leiomyosarcomas and family history of cancer
 - Positive result would point towards two independent primaries
- Pathologists' Assistant has critical role in thorough gross examination including careful dissection and tumour mapping (**Fig 2.**), which informs diagnosis and staging
- Differences identified and documented by Pathologists' Assistant (anatomical compartment, location (superficial vs. deep), gross appearance) aided in prompting sarcoma tumour board review
- Accurate sampling by Pathologists' Assistant important to capture areas of necrosis, viable tumor, and treatment effect
- pT staging for soft tissue tumors differs based on tumour site
 - Abdominal visceral organs (pancreatic mass) dependent on invasion, trunk and extremities (chest wall mass) dependent on size
 - Pathologists' Assistant must understand pTNM staging and which system applies to each tumour to direct gross dissection
 - Pathologists' Assistant assessment of pancreatic tumor's relationship to adjacent organs was therefore foundational to accurate staging

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