Primary Gastric Synovial Sarcoma: A Case Report

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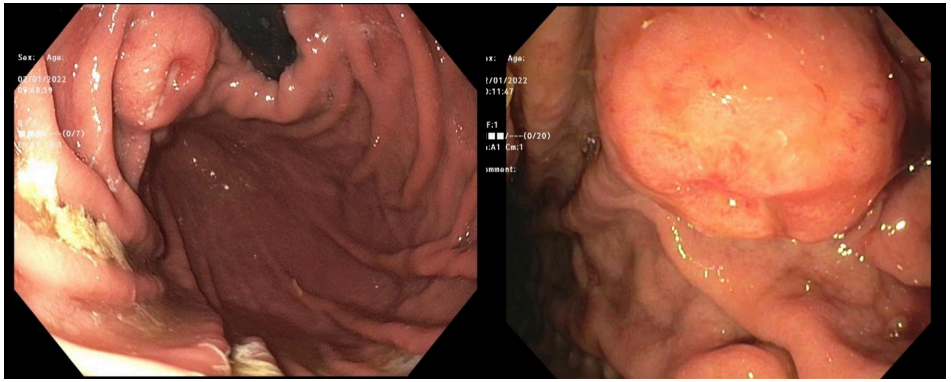
Introduction

Synovial sarcoma is an uncommon mesenchymal tumor, typically involving the extremities of young adults. This case outlines an especially rare occurrence of a primary synovial sarcoma occurring in the stomach of a 37-year-old female who ultimately required esophagogastrectomy.

Methods and Materials

- 37-year-old female presented with epigastric pain intermittently for more than a year. Her pain persisted and worsened.
- Initial workup including an EGD, CT of the abdomen/pelvis, and HIDA scan was negative.
- Several months later, an EGD was repeated due to ongoing symptoms. Her repeat EGD revealed a small ulcerated mass in the cardia, a c-KIT negative stromal tumor on histology.
- An EUS showed a 15mm, submucosal, ulcerated gastric mass, 1 cm from the GE junction.
- A primary synovial sarcoma with immunohistochemical stains and FISH technology for *SS18* translocation was identified.
- PET scanning showed no distant metastasis.
- The patient underwent a laparoscopic esophagogastrectomy with Roux-en-Y esophago-jejunal anastomosis.
- Final pathology confirmed a spindle cell sarcoma measuring 1.8 cm.No further adjuvant radiation or chemotherapy was indicated.

Results



Images 1 and 2. EGD images of ulcerated mass in the cardia of the stomach, approximately 1cm from the GE junction.

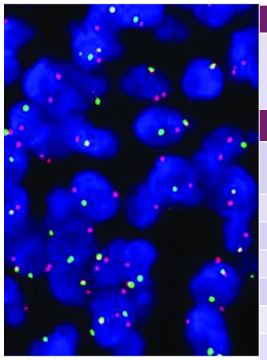


Image 3. Molecular study using fluorescence in situ hybridization (FISH) break apart probe confirmed a diagnosis of synovial sarcoma detecting the SS18 gene arrangement

FISH	Result	Interpretation
SS18 (SYT) Translocation	# of cells analyzed: 65 ○ **cells pos 55(84.6%)** ○ cells neg 10(15.4%)	Synovial sarcoma
Immunohistochemistry	**Result**	**Heading**
S100	Neg	Neural neoplasm (Neurofibroma/Leiomyoma)
SMA/Desmin	Neg	Smooth muscle neoplasm
CD34	Neg	Vasculature
CD68	Neg	Histiocytes
EMA/TLE1	Patchy/Positive	Highlighted malignant cells
C-kit and DOG1	Neg	GIST
Pankeratin (within lamina propria)	Neg	Atypical neoplastic proliferation of spindle cells

Table 1. Results of FISH study, and immunohistochemical testing demonstrating pertinent negative differentials

Discussion

- Synovial sarcomas account for approximately 10% of soft tissue tumors.
- Cell origin of these tumors are unknown, despite derivation of the name from the histological resemblance to synovial cells.
- These tumors are a result of chromosomal translocation t(X;18)p(11;qe1) which fuses the *SS18* gene from chromosome 18 to the *SSX* gene on the X chromosome resulting in either a biphasic tumor (*SS18-SSX1*) or a monophasic tumor (*SS18-SSX2*).

- Synovial sarcomas found in the gastrointestinal tract are exceptionally rare.
- As of 2021, 45 cases have been found in the stomach.
- Mortality of localized primary synovial sarcomas is 75% at 5 years and 34% at 10 years in a recent large case series.
- Poor prognostic factors include a tumor diameter >5 cm and microscopically positive margins.

Treatment & Prognosis

- Resection is the primary option depending on size and location of the tumor.
- This patient underwent an esophago-gastrectomy with clear margins.
- Rate of local recurrence is also high (30-50%) further supporting an esophagogastrectomy.
- Radiation is sometimes used as adjuvant or neoadjuvant therapy, and chemotherapy is typically reserved for metastatic disease.

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