High-grade serous carcinoma presenting as an isolated splenic mass: A Case Report

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The authors have no conflicts of interest that relate to the content of this abstract.

Introduction

- Splenic involvement by high-grade serous carcinoma usually occurs in the setting of widely disseminated disease from a known Mullerian primary, or as a recurrence years after primary surgery or treatment.[1]
- We present a case of high-grade serous carcinoma presenting as a splenic mass in the absence of concurrent or previous gynecologic malignancy. Given that splenic pluripotent stem cells are known to have low oncogenic potential,[2] and that the spleen is an intraperitoneal organ, we propose that this case represents a primary peritoneal high-grade serous carcinoma presenting as a mass lesion in the spleen.

Case presentation

- The patient is a 76-year-old woman with no significant medical history who presented with left upper quadrant pain for a few months. A PET/CT of the abdomen and pelvis done in June 2023 showed an irregular, heterogeneous 5.5 cm mass in the spleen, abutting the pancreatic tail with an SUV of 12.2 **(Fig. 1)**. The differential diagnosis included splenic angiosarcoma, lymphoma and a pancreatic mucinous cystic neoplasm involving the spleen.
- An endoscopic FNA of the pancreatic tail lesion revealed a carcinoma with micropapillary features. The tumor was strongly and diffusely positive for CK7, p16, WT-1, and PAX-8. P53 showed abnormal expression (null pattern). CK20 was focally positive while villin and TTF-1 were negative. These findings supported a diagnosis of high-grade serous carcinoma **(Fig. 2)**.
- The patient underwent bilateral salpingo-oophorectomy and histologic examination revealed only a serous tubal intraepithelial lesion (STIL). She started treatment with carboplatin and paclitaxel, resulting in interval decrease of the splenic mass to approximately 3.3 cm. Subsequently, she underwent a distal pancreatectomy and splenectomy, which showed high-grade serous carcinoma involving the spleen and pancreas and metastatic carcinoma to two peripancreatic lymph nodes **(Fig. 3, 4 and 5)**. The patient is currently on maintenance therapy with bevacizumab. Molecular studies revealed a *p53* and *CDK12* pathogenic variants.

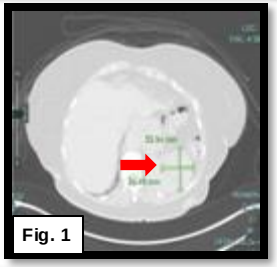
Fig. 1

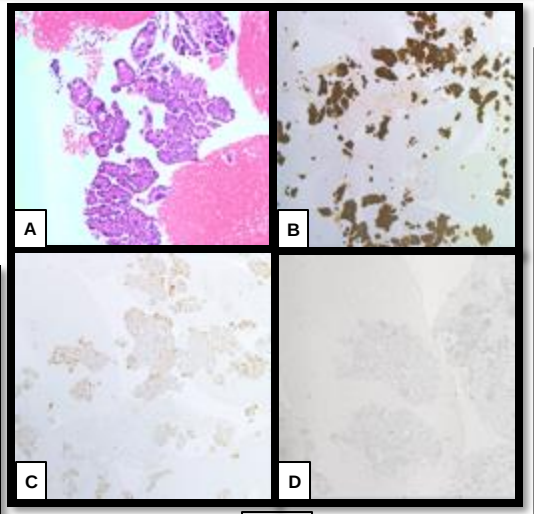

Fig. 2


Fig. 3

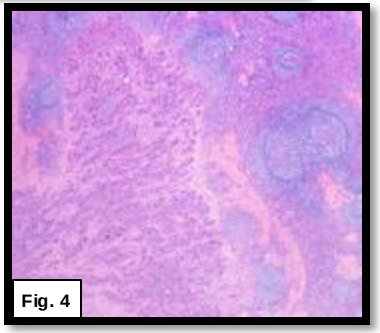
Fig. 4

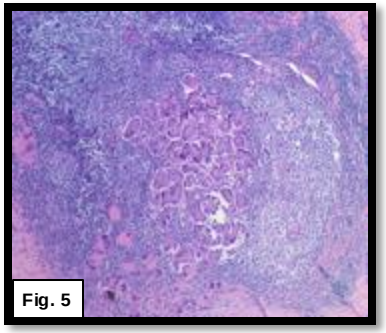
Fig. 5

Fig. 1. PET/CT showing an irregular and heterogenous mass measuring approximately 5.5 cm (arrow) in the lateral spleen involving the pancreatic tail. **Fig. 2: A**. Biopsy revealed a carcinoma with micropapillary features (H&E, 20x). **B**. Strong and diffuse positivity for p16. **C**. PAX-8 positive tumor nuclei. **D**. Null pattern of expression for p53. **Fig. 3.** Distal pancreatectomy and splenectomy specimen with 2.1 cm irregular mass (arrow) centered in the spleen and involving the pancreas. **Fig. 4.** Spleen with high-grade serous carcinoma (H&E, 5x). **Fig. 5.** Lymph node with metastatic high-grade serous carcinoma (H&E, 5x).

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Discussion

- Both primary splenic neoplasms and metastases to the spleen are uncommon. Primary splenic neoplasms are usually lymphomas, angiosarcomas, hamartomas, or hemangiomas.[3] Even though the spleen is the most vascularized organ, different anatomical characteristics like the sharp angle of the splenic artery, the contractile nature of the spleen and the lack of afferent lymphatics make it difficult for tumor emboli to enter and lodge in the spleen.[4] In the case of high-grade serous carcinoma, splenic involvement usually occurs in the setting of widely disseminated disease from a known Mullerian primary or as a recurrence years later after primary surgery or treatment.[1]
- In the present case, the patient had no history of cancer, and there was no evidence of concurrent gynecologic malignancy after an extensive workup. The pathogenesis of isolated splenic high-grade serous carcinoma is still unclear. We considered the possibility of a true primary splenic high-grade serous carcinoma; however, splenic pluripotent stem cells are known to have low oncogenic potential.[2] Therefore, we favor that this case most likely represents a primary peritoneal high-grade serous carcinoma arising in the splenic fossa and presenting as a mass lesion in the spleen.

Conclusions

- Most cases of high-grade serous carcinoma involving the spleen occur in a widely disseminated gynecologic primary setting, at initial presentation, or as a recurrence.
- The pathogenesis of isolated splenic high-grade serous carcinoma is still unclear. Possible mechanisms include involvement by a primary peritoneal high-grade serous carcinoma arising in the splenic fossa or malignant transformation from pluripotent stem cells.
- High-grade serous carcinoma can present as an isolated mass in the spleen; however, metastatic carcinoma from a Mullerian primary should be excluded before considering the tumor primary to the spleen.