

Background

A female in her 40s presented with 3 weeks of abdominal pain, abdominal bloating, constipation and weight gain of 15 pounds in one month. CT showed a 16 cm complex right ovarian mass highly concerning for ovarian malignancy, large volume ascites, omental caking and nodularity consistent with metastatic disease. Lab work showed elevated CA125 and low hemoglobin. She received a paracentesis the next day removing 8 L of serosanguinous fluid which was sent for CBC and cytology. CBC showed decreased hemoglobin overnight which was favored to be due to tumor hemorrhage. Cytology results were adenocarcinoma, but could not be further classified at the time from the received material due to low number of malignant cells on the cell block. The initial plan was for neoadjuvant chemotherapy. However, she had an increasingly distended abdomen as well as anemia with need for repetitive transfusion and almost immediate loss. Due to this concerning blood loss, she was consented for exploratory laparotomy, resection of mass and possible debulking.

Methods

During the surgery, 9L of hemoperitoneum was evacuated and the following findings were noted: A 16cm firm mass replaced the right ovary, omental caking (apparent source of hemorrhage), miliary hepatic surface involvement, miliary disease throughout the cul-de-sac and at the small bowel mesentery, several implants of tumor on the small bowel serosa, and a rind of nodular tumor at the left abdominopelvic peritoneum which extended down into the left pericolic gutter with extension onto the sigmoid colon serosa. The right ovary, right fallopian tube, omentum and a peritoneal biopsy were removed and sent to surgical pathology.

References

1. Provenza, Claudia MD^{*}; Young, Robert H. MD, FRCPATH[†]; Prat, Jaime MD, FRCPATH^{*}. Anaplastic Carcinoma in Mucinous Ovarian Tumors: A Clinicopathologic Study of 34 Cases Emphasizing the Crucial Impact of Stage on Prognosis, Their Histologic Spectrum, and Overlap With Sarcomalike Mural Nodules. *The American Journal of Surgical Pathology* 32(3):p 383-389, March 2008. | DOI: 10.1097/PAS.0b013e3181451b93
2. PDQ Adult Treatment Editorial Board. Ovarian Borderline Tumors Treatment (PDQ®): Health Professional Version. 2025 Feb 12. In: PDQ Cancer Information Summaries [Internet]. Bethesda (MD): National Cancer Institute (US); 2002-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK66031>
3. What is Angiosarcoma. Angiosarcoma Awareness. Published 2026. <https://www.cureasc.org/what-is-angiosarcoma/#:~:text=The%205%2Dyear%20survival%20rate%20for,the%20body%20is%20about%2016%25>

Results

Received in the gross room was an intact 18.5 x 16 x 12.5 cm right ovarian mass with areas of tumor and adhesions on the external surface (Figure 1). The cut surface was a multiloculated mucinous cyst with diffuse papillary excrescences, tan-red to tan-white solid areas and multifocal areas of hemorrhage and necrosis (Figure 2). The attached fallopian tube appeared to be grossly involved by tumor. The separately received omentum measured 37 x 21 x 3 cm in aggregate and had a tan-yellow to tan-red indurated hemorrhagic cut surface.

The diagnosis of the ovarian mass was epithelioid angiosarcoma arising from a mural nodule within mucinous carcinoma, involving ovarian parenchyma and surface. The mass was staged as pT3c and FIGO stage IIIC. The mucinous component of the mass showed a variety of morphologies including benign, borderline and carcinoma. The carcinoma showed mostly expansile growth pattern with some areas showing infiltration. The fallopian tube serosal surface, omentum and peritoneal biopsy were positive for angiosarcoma. Molecular testing was performed with both the angiosarcoma and carcinoma components demonstrating a *TP53* mutation and only the angiosarcoma component demonstrating a *KRAS* mutation. IHC results were as follows: ERG, which stains for vascular differentiation, was positive in the angiosarcoma portion of the mass (Figure 4), and HER2 was positive in the epithelial component but negative in the angiosarcoma component (Figure 5).



Figure 1. Intact ovary with extensive angiosarcoma tumor involvement of the external surface.

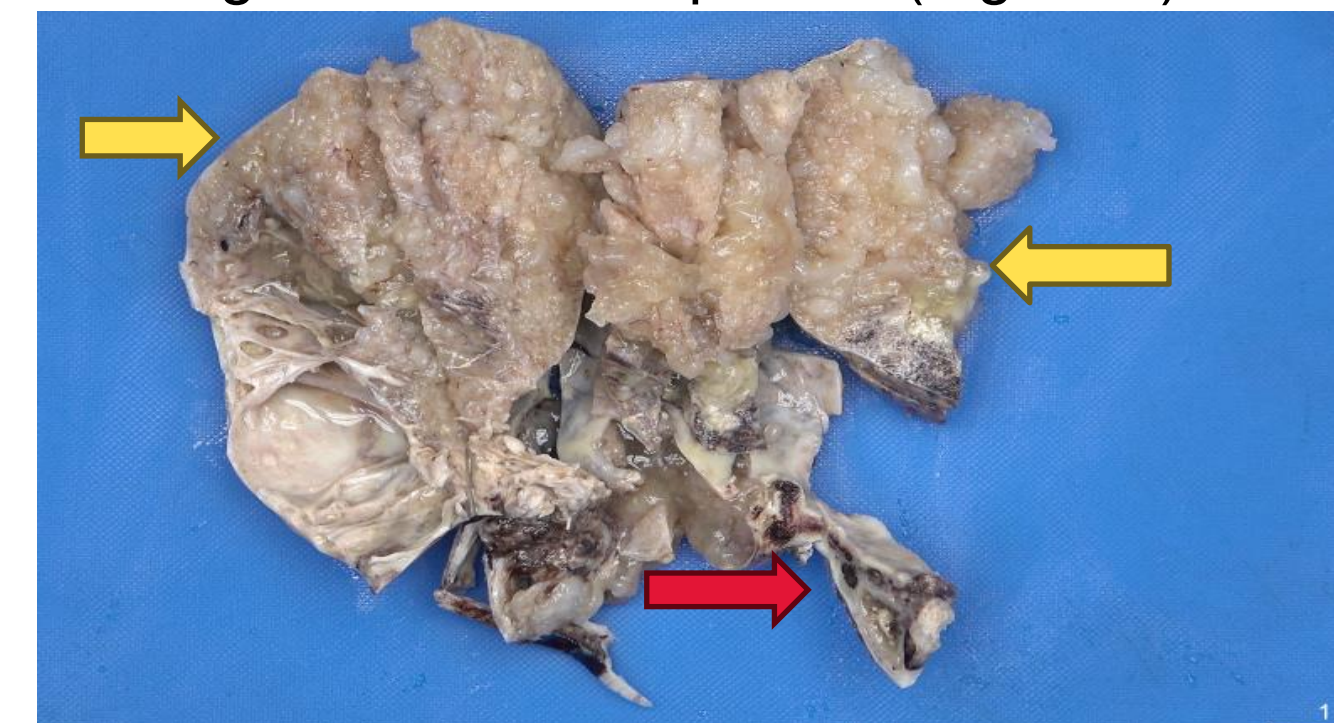


Figure 2. Cut surface of the ovary demonstrating mucinous cystic (top of specimen, yellow arrows) and tan-red mural nodule (red arrow) components

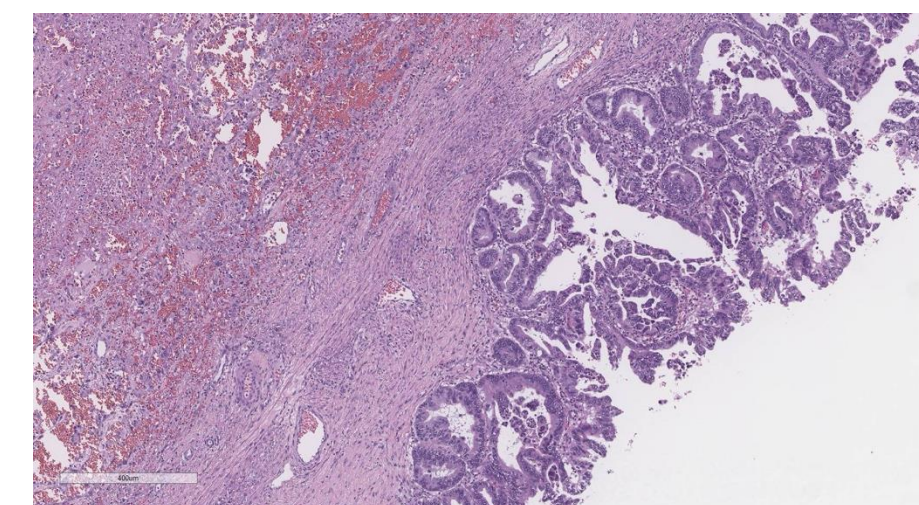


Figure 3. H&E, 6.5x magnification. The image shows the dual components of the neoplasm, with the mucinous adenocarcinoma component on the right side and the epithelioid angiosarcoma on the left side.

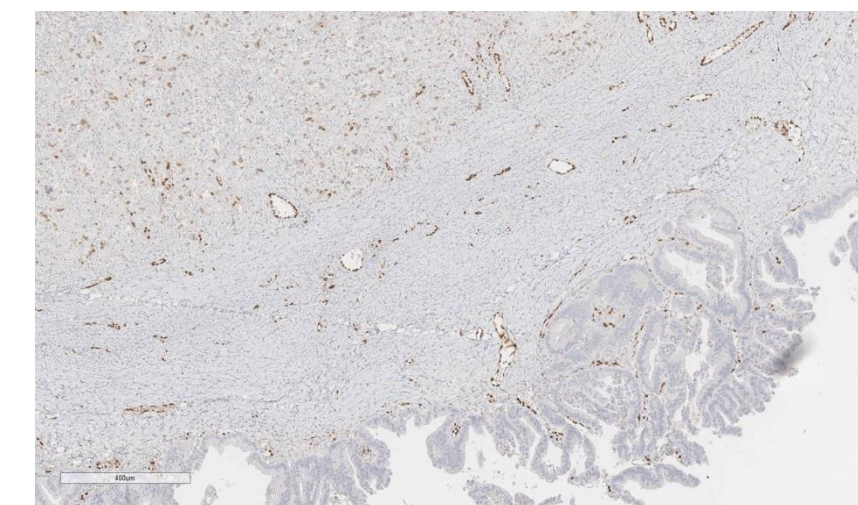


Figure 4. ERG IHC, 6.5x magnification. An ERG stain shows weak to moderate nuclear expression in the angiosarcoma component and is negative in the mucinous carcinoma. Background benign vessels show strong nuclear staining.

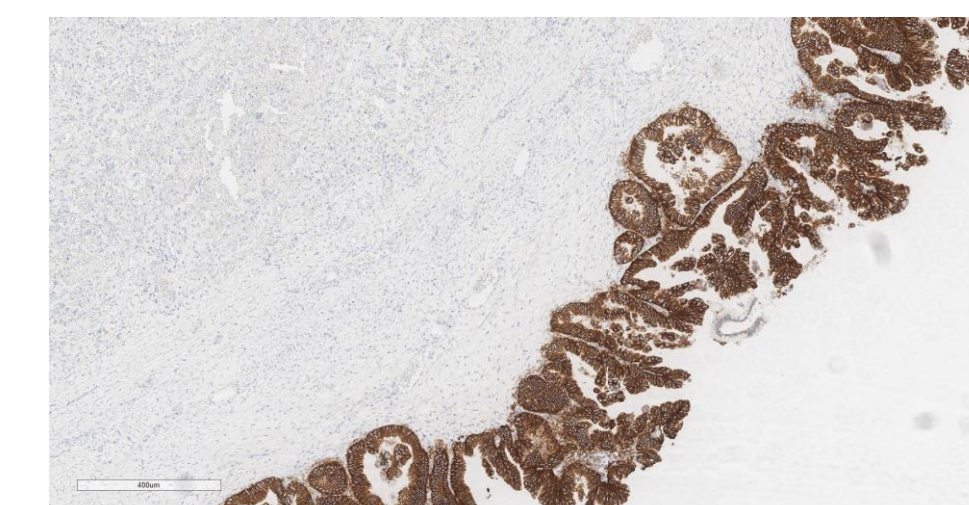


Figure 5. HER2 IHC, 6.5x magnification. A HER2 stain shows strong diffuse membranous staining of nearly all mucinous carcinoma cells and is negative in the angiosarcoma cells.

Conclusion

In rare cases, mucinous tumors of the ovary can be associated with a mural nodule which are classified as anaplastic carcinoma, sarcoma-like mural nodules (SLMN), or true sarcoma.¹ This mass has two distinct and separate components- mucinous carcinoma and epithelioid angiosarcoma. This is different from a carcinosarcoma, which is a biphasic neoplasm where the epithelial and sarcoma components are distinctive, but are intimately admixed.¹ An unusual finding in this case is the sarcoma infiltrated the ovarian tumor and extended beyond the ovary to the fallopian tube surface, peritoneum and omentum. More often, the mural nodules will stay confined within the tumor wall.¹ In a study of multiple mucinous neoplasms with anaplastic carcinoma nodules, the FIGO stage was the important prognostic factor.² Mucinous carcinoma with an infiltrative invasion is more aggressive and often associated with a higher FIGO stage and worse prognosis than one with expansile invasion.² The life expectancy for late-stage disease is 12-30 months and 5-year survival rate of 11-35%.¹ Angiosarcomas are aggressive and have a poor prognosis with a median survival rate of 15-24 months and 5-year survival rate averaging about 30%.³ If the tumor has spread to distant parts of the body, the a 5-year survival rate drops to about 16%.³ Treatment for mucinous carcinoma and angiosarcoma generally includes chemotherapy after surgery.^{2,3} The molecular and IHC results can guide the clinician to more targeted treatment. Mucinous neoplasms are sampled extensively to determine benign, borderline or carcinoma. Differentiating between these is important for both prognosis and treatment.² In cases where the tumor is confined to the ovary, surface involvement is important for staging and may also influence treatment. CAP recommends submitting 2 sections per centimeter of the greatest dimension of the mass in mucinous ovary tumors. Sample areas with thickened walls looking for mural nodules.