A Rare Case of Extragenital Müllerian Adenosarcoma

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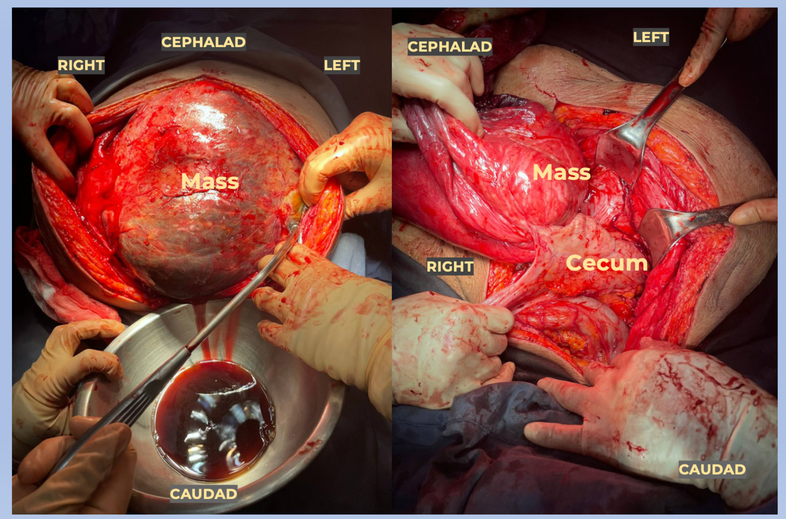
CASE REPORT:

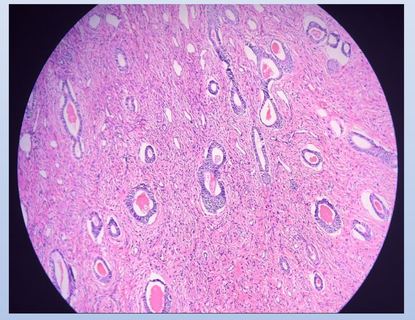
A 51-year-old gravida 5 para 5 (5005) presented with an increasing abdominal girth and a palpable abdominal mass. She was initially diagnosed with ovarian new growth and underwent exploratory laparotomy. Intraoperatively, the uterus, fallopian tubes, and ovaries were grossly normal and a large mass was seen attached to the cecum where the appendix should be. Referral to surgery service was done. Right hemicolectomy and ileostomy were performed.

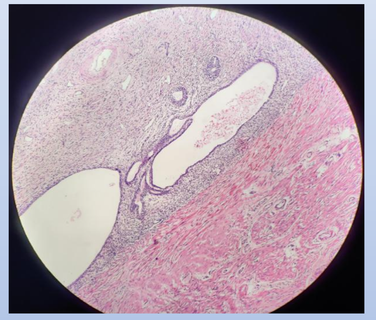


Histopathology report showed Mullerian adenosarcoma involving the appendix and cecum.

DISCUSSION:

Mullerian adenosarcoma (MAS) of the female genital tract is a rare tumor of low malignant potential consisting of benign epithelial and malignant mesenchymal components. It typically involves the uterus but extremely rarely it may arise in extragenital sites such as the colon. It is the rarest form of all uterine sarcomas. As of 2018, only 32 cases of extragenital Mullerian adenosarcoma have been reported in literature. Only 3 cases developed from the colon. How did it arise in this location? It can be associated with or may have developed from underlying endometriosis. Histologically, endometriotic sites were seen at the junction between mass and the cecum. This case satisfies Sampson's criteria for malignant transformation of endometriosis. The diagnosis of adenosarcoma is dependent mainly on histologic features and no immunohistochemical markers are pathognomonic. Immunohistochemistry studies have limited application to the diagnosis of adenosarcoma but may be necessary for differentiating those with sarcomatous overgrowth and may provide biologic insights into the possible efficacy of hormonal therapy. Her ER/PR assays were positive. Microscopic examination showed evidence of endometriosis with no evidence of sarcomatous overgrowth, features that are favorable prognostic factors associated with higher disease-free survival. Post-operatively, the plan of management was hormonal therapy.





CONCLUSION:

The rarity of this case imposes a diagnostic challenge on the clinician. No guidelines have been created on the management of extragenital MAS due to limited data and the variability of its presenting features, but complete surgical resection represents the best course of action. Hormonal therapy should be considered ER/PR positive MAS without sarcomatous overgrowth.

REFERENCES:

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2. *WHO Classification of Tumours Editorial Board. Female Genital Tumors: WHO Classification of Tumours (5th ed). World Health Organization 2020*