



DEPARTMENT OF PATHOLOGY

Case of the Week

Gynecologic Pathology: Aggressive Angiomyxoma

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History

The patient is a middle-aged female with remote history of myomectomy who presents with a pelvic mass. Gross examination revealed a lobular mass with rubbery consistency measuring 11.5 cm in greatest dimension. The cut surface was yellow to tan with areas of focal hemorrhage.

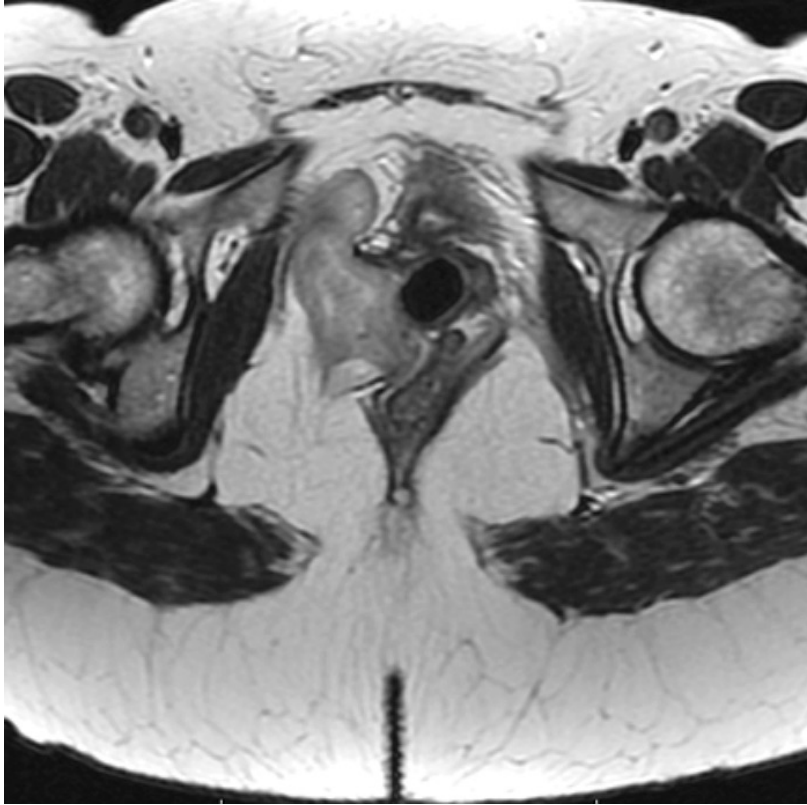


Figure 1: MRI shows a mixed density lesion in the right lateral vulvovaginal area.

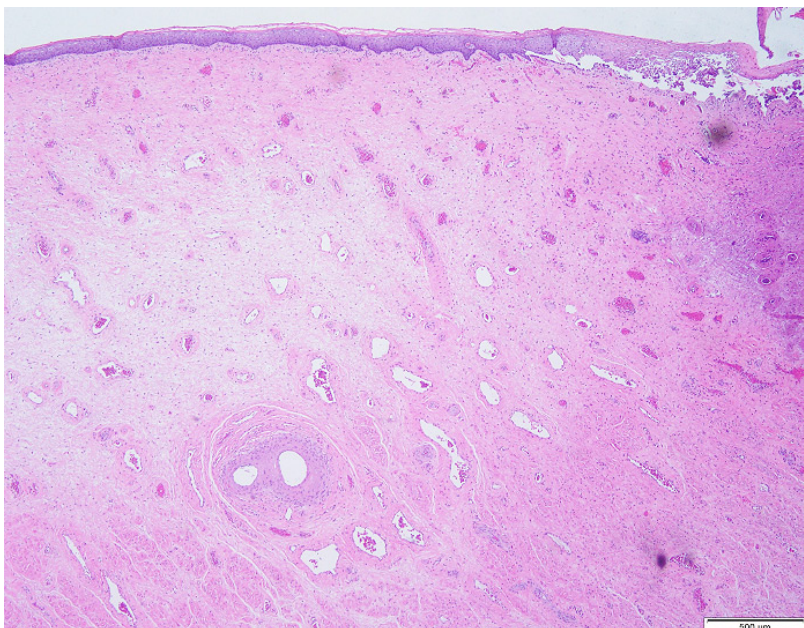


Figure 2: Section of cervix showing a hypocellular spindle cell proliferation with myxoedematous stroma and numerous scattered vessels. (H&E, 40X magnification)

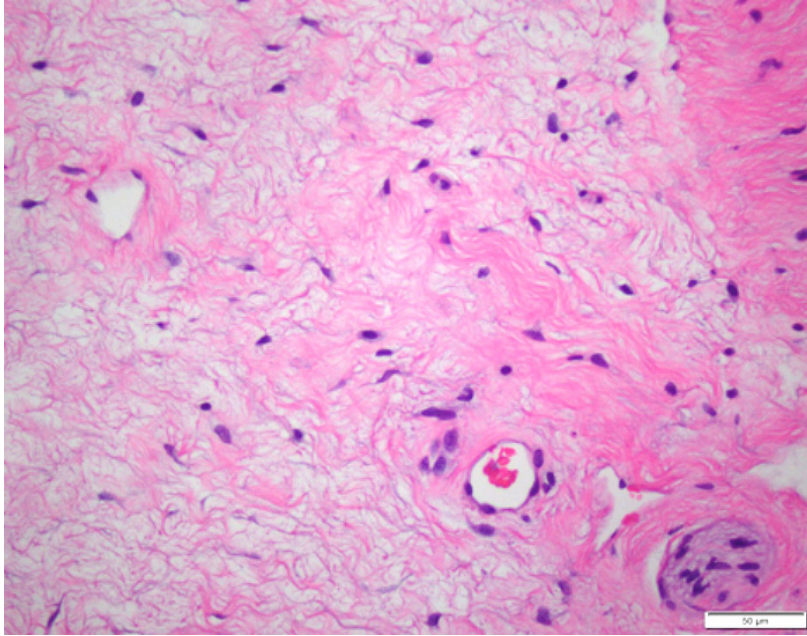


Figure 3: The tumor is composed of uniform oval and stellate-shaped cells in a background of wispy collagen fibers. No mitotic activity is observed. The tumor cells were seen surrounding vessels with foci of nerve entrapment (H&E, 400X magnification)

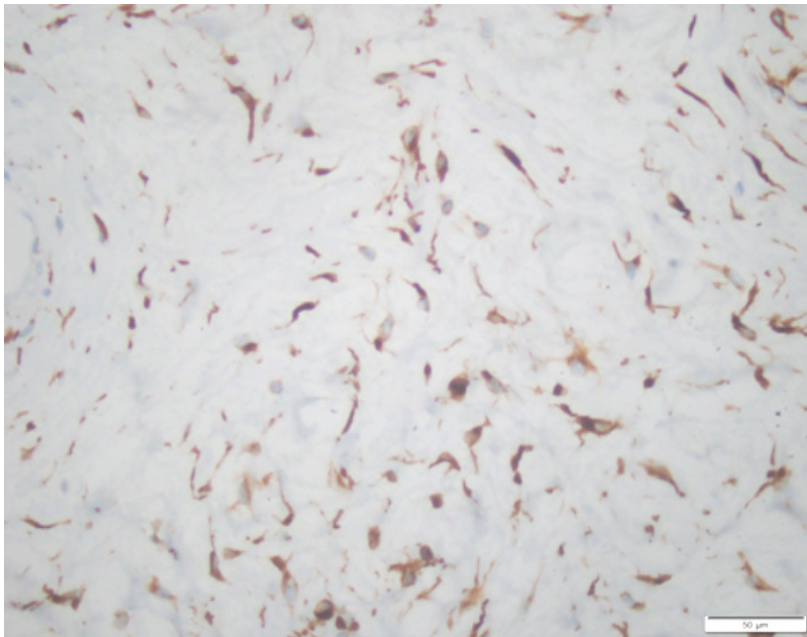


Figure 4: The tumor cells demonstrate immunohistochemically reactivity for desmin (IHC, 400X magnification)

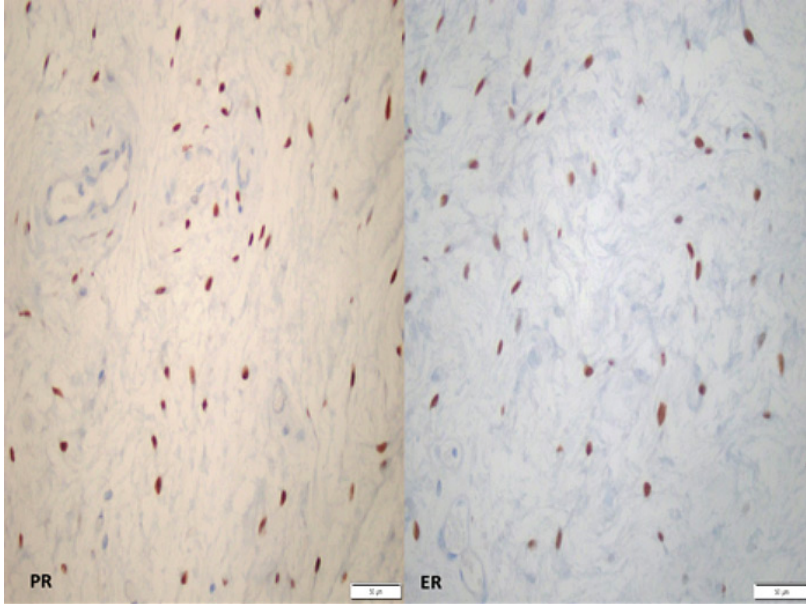


Figure 5: The tumor cells show strong nuclear positivity for both PR and ER (IHC, 400X magnification)

Diagnosis

Aggressive Angiomyxoma

Discussion

Aggressive angiomyxoma (AAM) is an uncommon mesenchymal tumor of the deep pelvicoperineal region that typically presents in the 4th-5th decade of life. Patients may complain of a pressure-like sensation although the mass itself is usually painless. The treatment is excision with wide margins due to the propensity for local recurrence.

Steeper and Rosai were the first to describe AAM as a spindle/stellate lesion with myxoid stroma, prominent thick-walled blood vessels, and pushing borders.¹ The cells are typically bland with coarse chromatin and little to no mitotic activity. Other key features include wispy collagen fibrils around vessels and entrapment of nerves, the latter of which was seen in this case.

Interestingly, the myxoid component is thought to represent edema rather than true “myxomatous” stroma and does contain abundant mucosubstances.² The spindle cells share ultrastructural features with myofibroblasts and demonstrate immunohistochemical reactivity for SMA, Desmin and Vimentin; additionally, they are strongly reactive for ER and PR.²⁻⁴

The differential diagnosis includes both benign and malignant mesenchymal tumors. AAM has some overlapping histologic features with superficial angiomyxoma and angiomyofibroblastoma.⁵ Superficial angiomyxoma tends to be located in the dermis/subcutis area and usually lacks the larger hyalinized vessels. Additionally, it has abundant hyaluronic acid

(Alcian blue positive) while being negative for ER and PR. Angiomyofibroblastoma is typically a sharply demarcated lesion that features epitheloid cells clustered around vessels. The overall lack of nuclear atypia and uniform appearance of the cells distinguish it from myxoid sarcoma.

AAM has been reported to have a characteristic cytogenetic abnormality in the HMGA2 that may be helpful in differentiating it from other pelvis/perineal mesenchymal tumors. HMGA2 gene rearrangement has been reported to occur in 33% of AAM and is not present in angiomyofibroblastoma or cellular angiofibroma.⁵

References

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