

Aggressive angiomyxoma of the vulva - Case report

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INTRODUCTION

- Aggressive angiomyxoma (AAM) is a **rare vulvovaginal, perineal or pelvic mesenchymal neoplasm** that occurs almost **exclusively in women of reproductive age**. The term aggressive emphasizes the often infiltrative nature of the tumor.
- Patients often present with **nonspecific symptoms** which are frequently **misdiagnosed** with more common entities, such as a Bartholin cyst, lipoma, or hernia.
- The tumor cells are characteristically **positive for estrogen and progesterone receptors**, suggesting a hormonal role in the development of the tumor.

FIG. 1a



FIG. 1b



FIG. 1c



FIG. 1a, b and c - Vulvar tumor excision

FIG. 2a

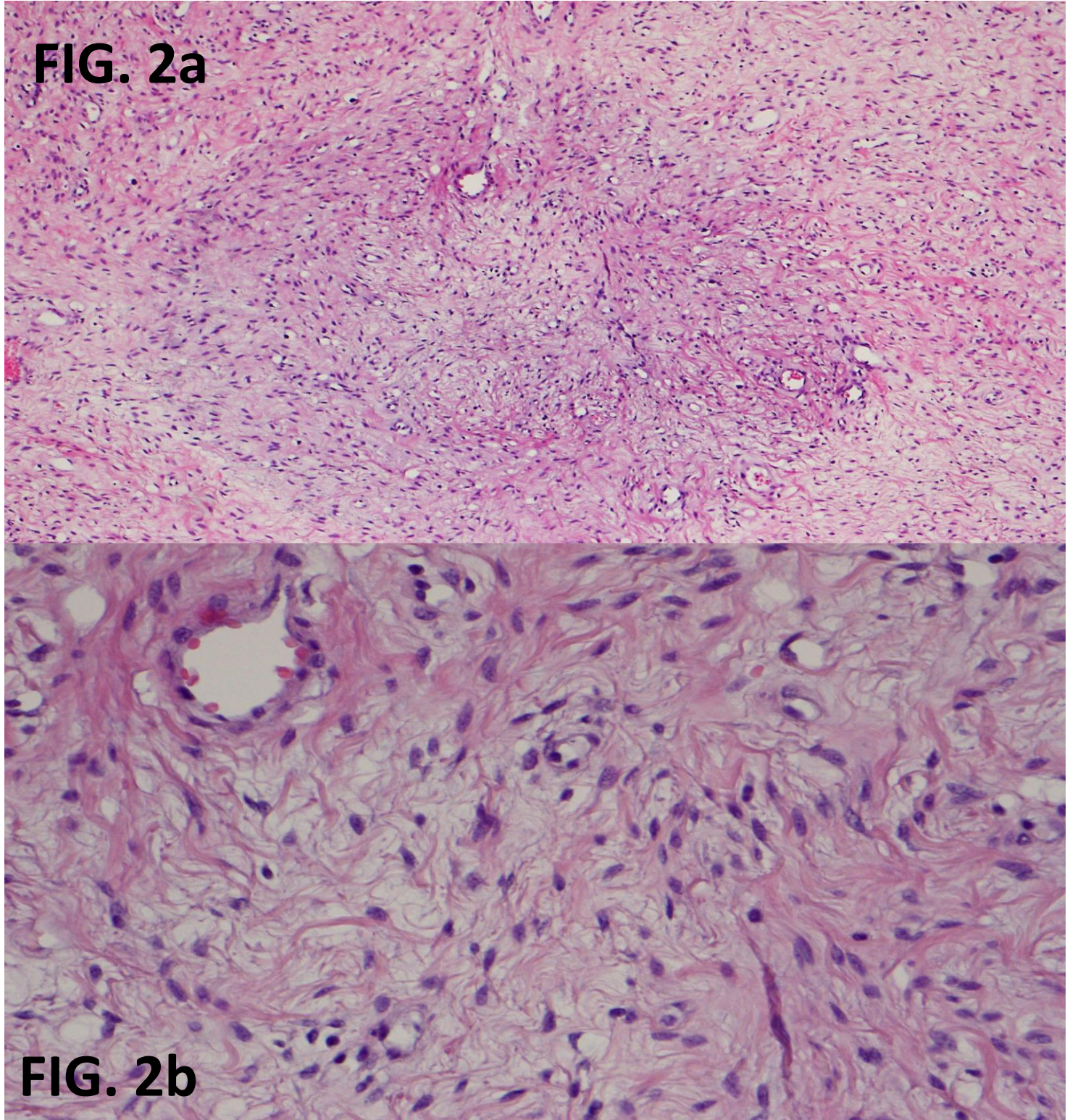


FIG. 2b

FIG. 2a and b – Histopathology: aggressive angiomyxoma on hematoxylin eosin

CLINICAL CASE

- We report a case of an aggressive angiomyxoma of vulva in a 33-years-old women, with progressive growth for six years, with rapid growth during pregnancy.
- On physical examination, a **tumor of 7 x 5cm was found, located on the right labia majora**, soft tissue dependent. Ultrasound-guided needle biopsy was performed, but the diagnosis was inconclusive.
- Wide **resection** of the tumor was performed at 19 weeks pregnant (FIG. 1).
- Histopathology (FIG. 2) reported an **aggressive angiomyxoma of the vulva**, with **positive estrogen and progesterone receptors** and **with tumor in resection margins (R1)**.
- In the oncologic multidisciplinary team meeting, the decision of **surveillance** was taken due to being pregnant.
- Adjuvant therapy was not performed and there is no recurrence until now (3 years later), with surveillance with MRI.

DISCUSSION

- **Surgery** remains the **main treatment** option. A retrospective study with the largest number of cases (106 cases) pointed out that there was **no significant difference in the recurrence-free rate** between patients with negative and positive margins within 10 years after surgery.
- As **late recurrences are known**, all patients need **long-term follow-up**. Clinical examination may be insufficient to detect recurrence. **MRI** may detect recurrence earlier, but there are no guidelines concerning their frequency.
- Adjuvant radiotherapy and hormonal therapy are not fully stablished. Metastases are exceedingly **rare**, and overall, the **prognosis is good**.