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Aggressive Angiomyxoma of Vulva

Pages with reference to book, From 123 To 125

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Aggressive angiomyxoma is an infrequent tumour of the vulva of which only single case and small series have been reported in the literature¹⁻⁴. The biological behaviour of this lesion is variable. In many patients, the tumour is benign, although an aggressive outcome with recurrences is common. Metastasis has not been described. We report three cases of aggressive angiomyxoma of the vulva that clinically presented as large bulky polypoidal masses.

Clinical and pathological Findings

Case 1

A 16-year old unmarried female presented with a history of vulval mass in the labial region which has steadily grown since last two years. Over a period of 5 months prior to her visit, she had fever off and on and was seen by a number of doctors and was prescribed a wide array of drugs including various antibiotics, antituberculous drugs and steroids. She also had an echocardiogram elsewhere which was interpreted as chronic pericarditis with mild effusion suggestive of tuberculosis. Apparently, her vulval mass was never examined. At the time of presentation, the mass showed extensive skin ulceration with purulent discharge arising from right labia majora (Figure 1).

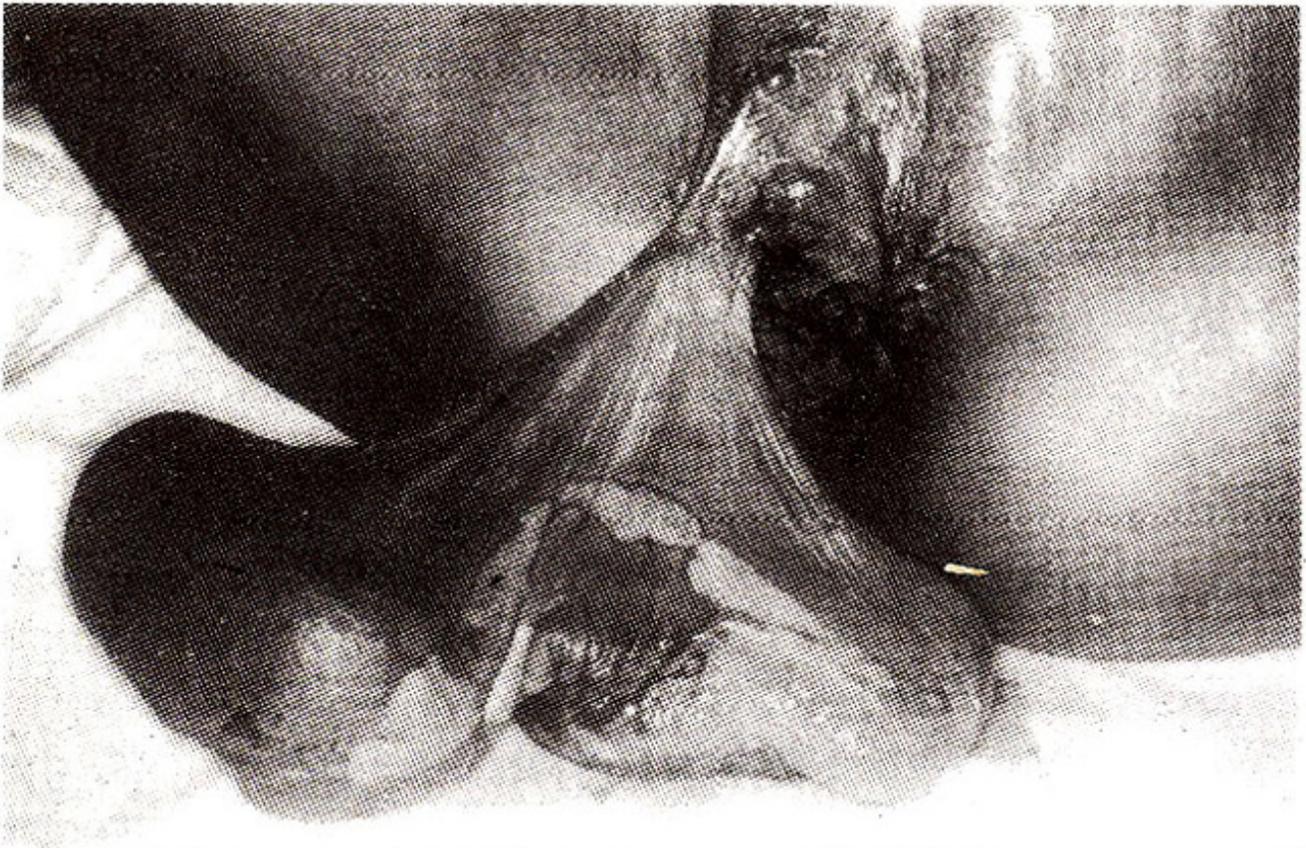


Figure 1 - Case 1. Large polypoid mass arising from labia majora.

Pelvic examination revealed a normal sized uterus and no adnexal mass was felt. Her haemoglobin was 7.6 g/dl and she was transfused four units of packed cells before the surgery for the excision of the mass. No recurrence was reported till the last follow-up one year after surgery. The surgical specimen weighed 860 g and measured 21x15x11 cm. The cut surface was gelatinous white and soft. Microscopically, the tumour consisted of myxoid tissue with enmeshed benign, uniform stellate cells which lacked atypia. The myxoid stroma was moderate to markedly vascular consisting of blood vessels of variable caliber (Figure 2).

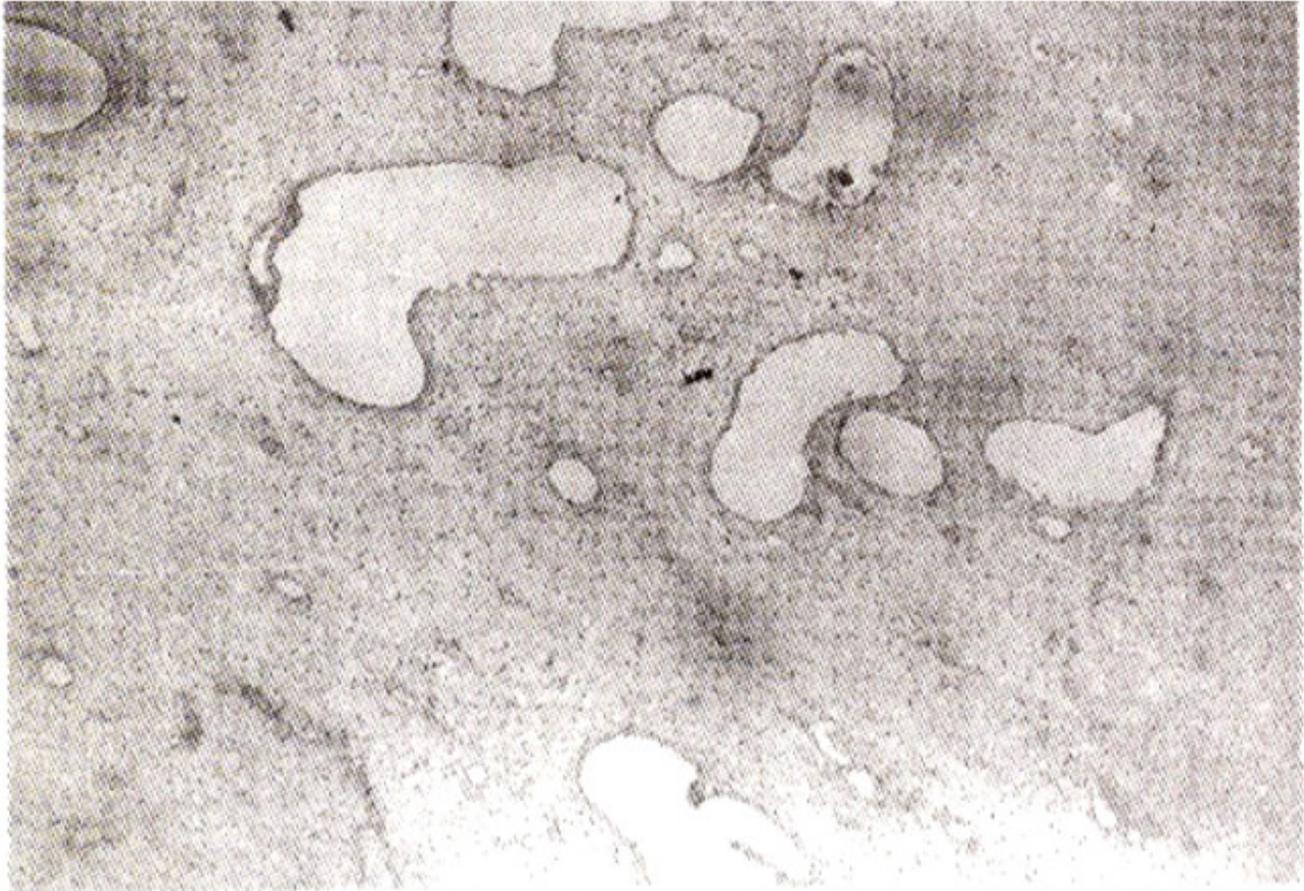


Figure 2 - Case 2. Thin walled vessels of various sizes separated by abundant myxoid stroma.

The majority of vessels had dilated lumina with small foci of fresh hemorrhages in the myxoid stroma. Acute and chronic inflammation was seen only in areas immediately adjacent to skin ulceration. No mitosis or nuclear atypia was noted in stellate stromal cells. Immunohistochemical studies revealed these stromal cells positive for vimentin but negative for actin and desmin.

Case 2

A 36-year old married female who presented with a history of progressively increasing swelling in the left labial region for the last four years. She experienced discomfort on sitting and mild dyspareunia. Examination revealed a soft mass arising from the left labia majora. There was no discharge, oozing or ulceration. Bimanual examination and ultrasound revealed a normal pelvis. The tumour was excised under general anaesthesia. Postoperative recovery was uneventful. No recurrence was noted for a year, but then she was lost to follow-up. The surgical specimen measured 13x10x8.5 cm. The overlying skin was intact and unremarkable. Cut surface was light grey white, gelatinous with small focal haemorrhages. Histological features were almost identical to that of case 1, however, the vascular walls tended to be thicker and there were foci of perivascular lymphocytic aggregates. Focal areas of fibrosis were also present. On immunohistochemistry, the stromal cells were positive for vimentin but were

negative for actin and desmin.

Case 3

A 35-year old married female gave a 10-year history of a slowly growing mass in right labia majora. The mass was excised under general anaesthesia. The post-operative period was uneventful. The mass measured 10.5x5.5x4.5 cm. No overlying skin was present in the specimen. The cut surface was soft and gelatinous with many mucoid and cystic areas. Histological findings were similar to the above two cases. In addition, however, the vascularity tended to be angiomatous. Small foci of capillary hemangioma like areas were separated by vascular myxoid areas (Figure 3).

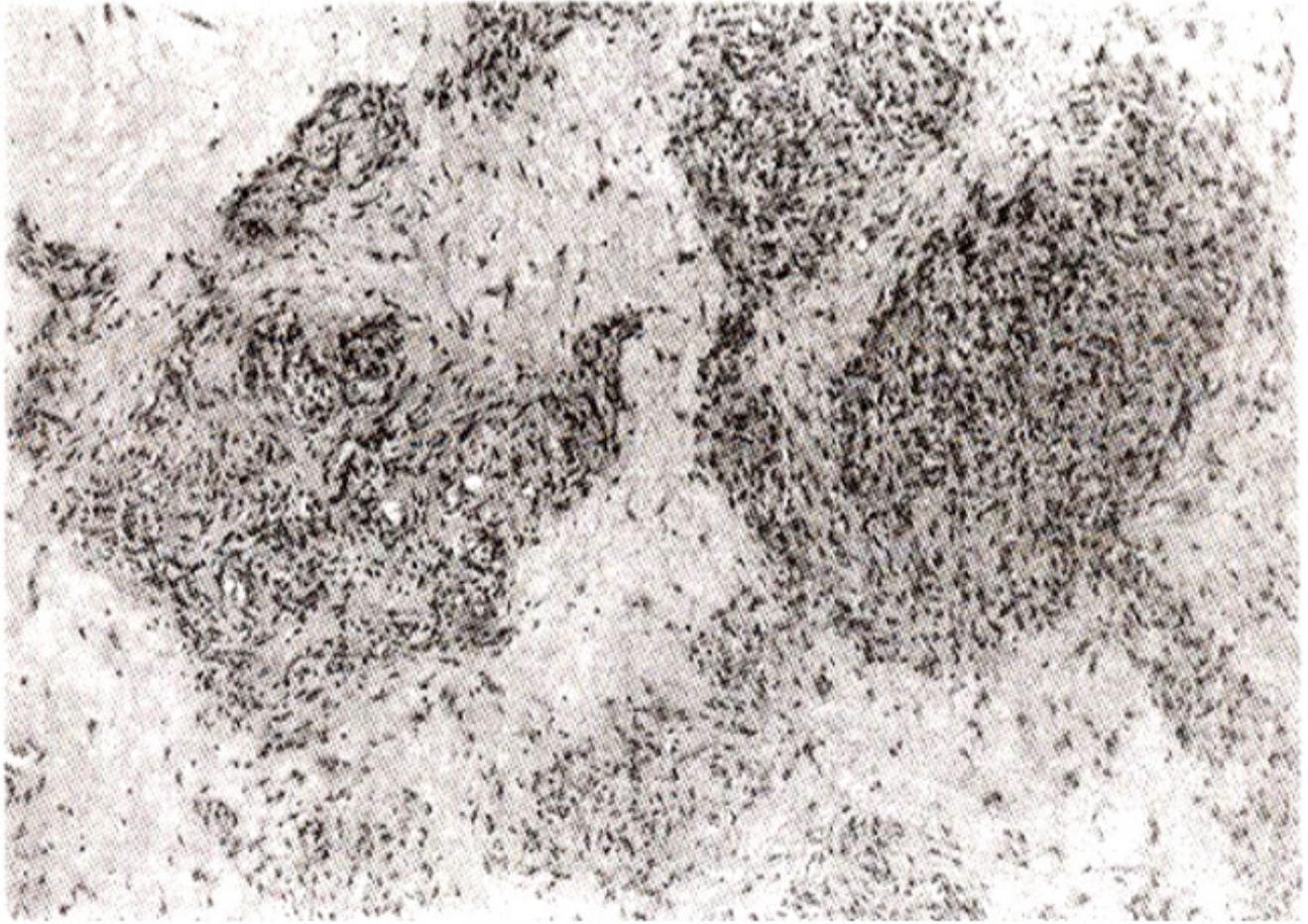


Figure 3 - Case 3. Capillary hemangioma like areas separated by myxoid stroma.

No atypia or mitosis were noted in the stellate stromal cells.

Discussion

Aggressive angiomyxoma was the term coined by Steeper and Rosai for a distinctive type of gynaecologic myxoid and vascular neoplasm¹. The term 'aggressive' denoted its propensity to recur if not adequately excised. Reliable estimates of recurrence rates are not known. To date, no metastatic lesion has been documented. Lesions with perhaps similar pathology had been identified before. Virchow described a labial lesion as 'myxoma' which later on recurred². Various lesions were thought as 'fibromas' and 'liposarcomas' with myxoid change attributed to degeneration³. Aggressive angiomyxoma diagnosed as such, is an exceedingly rare neoplasm and this is perhaps largely due to non-recognition. Our three cases measured more than 10 cm in greatest dimension. The size range in

Steeper's¹ series was 3-60 cm. One of our patients (case 1) was only 16 years old while in Steeper's series, the age range was 21-38 years. Aggressive angiomyxoma has also been reported in pelvic soft parts⁴, retroperitoneum⁴, umbilical cord⁵, levator hernia⁶ and scrotum⁷. However, its predominant location is in vulva and perineum and this is probably due to relatively myxoid and vascular stroma normally present at these sites. The neoplasm lacks encapsulation and may infiltrate the neighbouring areas causing inadequate excision and tendency to recur. The neoplasm has two basic histologic features, the myxoid stroma with benign stromal cells lacking mitosis and nuclear atypia. The more characteristic feature is its vascularity. The vessels are part of the tumour, moderately abundant, usually present as dilated capillaries, but lacks the anastomosing pattern. Thickening of the vascular smooth muscles can also occur. In one of our cases (case 3), capillary hemangioma like areas were widely separated by vascular myxoid stroma. The stromal cells were thought of fibroblastic or myofibroblastic on the basis of immune histochemical and ultrastructural investigations⁸. Our immunohistochemical findings are in accord with other investigators. Entrapped glandular elements⁹ were not seen in our cases. The differential diagnosis of aggressive angiomyxoma includes other myxoid neoplasms such as myxomas, myxoid malignant fibrous histiocytoma, embryonal rhabdomyosarcoma (sarcoma botryoides), myxoid liposarcomas, nerve sheath myxomas and leiomyomas with myxoid change, especially during pregnancy. However, in most cases, the diagnosis should not be difficult as long as one is aware of the existence of this entity and carefully excludes other diagnostic possibilities. Immunoperoxidase studies may be helpful in difficult cases. Smaller tumours are apt to be confused with benign vulval fibroepithelial polyps which paradoxically tend to have atypical stromal cells and will lack the peculiar vascularity of aggressive angiomyxoma.

Acknowledgement

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