Angiomyxoma of Vulva – A Rare Undifferentiated Soft Tissue Tumor

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ABSTRACT
Angiomyxoma is a rare, locally invasive mesenchymal tumor occurring in women of reproductive age, which carries a high risk for local relapse. It can be mistaken both clinically and on microscopy for several other conditions; it should be included as a differential diagnosis for any vaginal mass. We report a case of 36 year old woman who was clinically diagnosed as Bartholin’s cyst/ Lipoma, intraoperatively found to have soft tissue mass underwent complete surgical excision and reported as Angiomyxoma.

KEY WORDS: Angiomyxoma, Mesenchymal tumors, Undifferentiated tumors.

Introduction
A 36 year old female patient presented with painless swelling in the left labial region since 6 months duration, not associated with fever, trauma. On local examination, 6x4 cm swelling was present over left labium majus, Skin was normal, Soft in consistency, Non transilluminant, No cough impulse, irreducible. Vaginal and rectal examinations: Normal, Clinical diagnosis is unknown. Bartholin’s cyst not known. Diffuse Lipoma in left labia majora.

Pre-op image: Left vulval swelling
Investigations:
Ultrasound over the genital region: Cystic lesion with internal echoes on left labium majus with no demonstrable intraperitoneal communication. Bartholin’s cyst not known.

FNAC: Smear showed only anucleate squames; suggested biopsy.

Surgery: Planned for excision biopsy. Intraoperatively, a solid fleshy mass of about 8x4 cm occupying lateral part of labium majus extending into ischiorectal fossa. Mass was excised too. Specimen marked with proper orientation and sent for HPE.
Intra op image: soft fleshy mass with deep extension. Post op recovery uneventful.

Histopathological Examination: Reported as Hypocellular elements and myxoid stroma with many blood vessels. Impression: Angiomyxoma of vulva, margins free of tumor.

HPE shows myxoid stroma with blood vessel.

Post OP MRI: Post op edematous changes with minimal fibrosis in lateral part of labium majus and ischiorectal fossa; No residual mass.

Patient is on regular follow up for the past one year. No evidence of any recurrence.

Discussion

The first description about Aggressive angiomyxoma was given by Steeper and Rosai in 1983, as a slow growing painless benign tumor but has infiltrative potential into skeletal muscle and fat. The WHO classified Angiomyxoma under “Tumors of uncertain differentiation” [2]. It is most commonly found in women of reproductive age group. The female: male ratio is 6:1. It is more common in perineum, vulva and vagina. It has high risk for local recurrence but with a low tendency to metastasize. Hence it requires long term follow up.

There is no complete consensus regarding the tumor pathogenesis. In chromosome 12, a gene in the region 12q13-15 called high-mobility group protein isoform I-C (HMGIC), which encode proteins involved in the transcriptional regulation, appears to have a role in the pathogenesis of this tumor [3]. Immuno histochemically, most Angiomyxoma express different combinations of estrogen and progesterone receptors, vimentin, desmin, smooth-muscle actin, muscle-specific action, CD34, and CD44, but all are invariably negative for S-100, CEA, and keratin [4-6].

Misdiagnosis occurs in more than 80% of cases [6], primarily due to the rarity of this tumor, and its vast differential diagnosis. Clinically, the differential diagnosis of a perineal mass in females include, Bartholin cyst, Gartner's duct cyst, vaginal cyst/polyp, vulvar abscess/neoplasm/cyst, vulvar edema, lipoma, canal of Nuck hernia, pelvic floor hernia, and vaginal prolapse [6-8,10].

Histologically angiomyxoma is a mesenchymal tumor, composed of fibroblasts within a strong myxoid background.

Vascular proliferation is also prominent, and virtually no mitoses are present. Histopathology mimics of Angiomyxoma: angiomyofibroblastoma, fibroma, myxofibrosarcoma, myxoid leiomyoma, lymphangioma, neurofibroma, malignant mesenchymoma, malignant fibrous histiocytoma, myxolipoma, sclerosinghemangioma, botryoid pseudosarcoma, myxoid smooth muscle and nerve sheath tumors, mixed mesodermal tumor, leiomyosarcoma, and embryonal rhabdomyosarcomas [7,10].

History taking and physical examination further guide the clinical workup. Imaging is ultimately recommended to identify the extent of pelvic involvement of perineal masses as
their sizes are often underestimated by clinical examination. Sonography shows a mass that is hypoechoic or appears frankly cystic. On CT, the tumor has a well-defined margin and attenuation less than that of muscle. On T2-weighted MR imaging, the tumor has high signal intensity [11].

The hormone dependency of this tumor suggests that gonadotropin releasing hormone agonists, may be of value in management of aggressive angiomyxoma, which are not amenable to surgical excision. GnRH agonists may also be used to reduce the size, so that more conservative surgery can be performed[5,9].

Surgery by margin free wide excision was the mainstay of treatment. Prior to surgery misdiagnosis of Angiomyxoma was common due to rarity of the tumor. Preoperative angiographic embolization, preoperative external beam irradiation and intraoperative electron beam radiotherapy were used to decrease the chances of local recurrence [12,13]. Detection of inappropriate HMGI-C expression using the immunoperoxidase technique with anti HMGI-C antibody may potentially be a useful marker for microscopic residual disease [3].

In our patient, physical examination revealed a well-circumscribed soft mass in the left labia majora and diagnosed as Vulval Lipoma or Bartholin’s cyst. Intraoperatively, we found a fleshy mass and diagnosed as a soft tissue tumor. Hence we did Wide local excision. It was encapsulated by a fibrofatty layer. Furthermore, the excised lesion had negative margins on pathologic evaluation. Since the patient is not affordable to do IHC and On H&E staining, the tissue resembled a typical aggressive angiomyxoma, obviating the need for any further immunohistochemical analysis. We did MR Imaging post operatively to assess the residual lesion and MRI revealed no residual lesion. Angiomyxoma is notorious for local recurrence in approximately 70% of the cases after a period of 2 years postoperatively[12].

Han-Guertsetal. [13] proposed the following guidelines in treating Aggressive Angiomyxoma:

1. Complete excision of the lesion when possible and avoiding mutilating surgery,
2. Adjunct therapy, when partial resection performed is acceptable using arterial embolization and/or hormonal treatment.
3. Radiotherapy is reserved to cases that are resistant to embolization and/or hormonal therapy and still symptomatic.

Multimodal therapy is advised when complete resection is not possible.

The prognosis is very good. Only 2 cases with metastatic disease followed by death have been reported. Recurrences are common, though, reported to be between 9 and 72%. There are no guidelines in the postoperative management of vulvar Angiomyxoma. However due to the high recurrence rate and potential morbidity associated with undiagnosed recurrences, many studies have recommended periodic evaluations with physical examination and MR imaging up to 15 years after treatment.

**Conclusion**

Although a rare disease to diagnosis, Aggressive Angiomyxoma can present with unusual features. Detailed radiological examination is helpful in detecting the problem, but histology is the gold standard for diagnosis. Wide excision is curative and prognosis of such tumors is good. Long-term follow-up is necessary and MRI is the preferred method for detecting recurrences.
References


