INTRODUCTION

Angiomyofibroblastoma (AMF) is a rare benign mesenchymal tumour of lower female genital tract. It was first described by Fletcher et al. in 1992 as a new entity that occurred exclusively in the vulva of young to middle-aged women. He described 10 cases of a previously unrecognized benign soft tissue tumour of the vulva that was often misdiagnosed as aggressive angiomyxoma. Clinically, most of the tumours present as slowly-growing painless masses, with low tendency for local recurrence and are often misdiagnosed as a Bartholin's gland cyst, hydrocele of the canal of Nuck, and aggressive angiomyxoma. Its texture is rubbery or smooth and looks oedematous, when cut. As a rule, it shows spindle-shaped cells with production of collagen fibres and alternates hypercellular areas, particularly around the vessels, and hypocellular areas. Sometimes mature adipocytes can be found in the tumoural tissue. Conventional morphologic analysis is paramount in the recognition of genital AMF. This unusual neoplasm should be distinguished from aggressive angiomyxoma and other myxoid malignant tumours with widespread metastatic potential.

Immunohistology may be helpful in a limited context in excluding other differential diagnoses.

CASE REPORT

A 30-year-old female presented in the outpatients department of GTTH, Lahore in July 2010 with a history of a swelling in the right labia for the past 6 months. The patient noticed a pea sized swelling in the right labia majora initially, which gradually increased till it reached the present size. The swelling was non-tender and non-irritating. Systemic history was not remarkable. The patient had regular menstrual cycles with moderate flow. She was gravida 4 and para 4 with no history of abortion. All children were delivered in hospital by the normal vaginal route. Past medical and family history were not significant. General physical and systemic examination were unremarkable.

Local examination revealed a 6 x 5 cms firm swelling on her labia. The lesion was covered by skin on all sides without any hypo/hyper-pigmented or ulcerated areas on the surface (Figure 1). The mass was non-tender, with normal overlying temperature. External genitalia including left labium major were normal looking.

The patient underwent complete surgical excision of the lesion. The sample was sent for histopathological examination. The specimen consisted of a 5 cm x 4.5 cm x 1 cm, soft tissue piece covered by skin on all sides. Multiple serial sectioning of the specimen revealed brown coloured fibro-fatty tissue. Multiple representative sections were taken and submitted for processing.

Histological examination revealed a skin covered tissue showing hyper/hypocellular areas containing spindle and plump stromal cells admixed with blood vessels.
The initial diagnosis of the lesion was that of a fibroepithelial polyp. However, the diagnosis of angiomyofibroblastoma was later made, considering the presence of plump spindle cells admixed with thick collagen strands (Figure 2) and the presence of blood vessels. Immunohistochemical study was not done. The resection proved to be curative with no recurrence till the last follow-up which was in February 2011.

**DISCUSSION**

AMF was initially thought a vulva-specific neoplasm, however, it can also occur in the vagina, perineum and inguinal areas. It is believed to be derived from mesenchymal cells in the subepithelial myxoid stromal zone that extends from the endocervix to the vulva. This hypothesis explains to some extent the propensity of this tumour to arise in the lower genital tract.

The most crucial issue was to determine whether this case should be assigned to AMF or to aggressive angiomyxoma (AAM). The latter is a histologically benign soft tissue tumour, associated with a high risk of local recurrence as well as with local infiltration that often results in entrapment of nerves and mucosal glands, thus making complete excision difficult. AMF is typically a well-demarcated tumour that ranges in size from 0.5 to 12 cm (mean 4.5 cm). The microscopic characteristics of this case were in agreement with those demonstrated by AMF rather than AAM, since the tumour was a well-circumscribed, relatively small-sized (6 cm), lesion with no infiltrating margins. With regard to biologic behaviour, all published reports suggest a benign clinical outcome in patients with AMF with no signs of local recurrence till the last follow-up.

The differentiation of 'angiomyofibroblastoma' based on histological properties is formulated on the two integral components of this tumour: stromal cells and the blood vessels. AMF is characteristically a well-circumscribed lesion composed of alternating hyper and hypo-cellular mass. Plump round to spindle-shaped cells either in cluster or present in a linear array around numerous delicate capillary-sized vessels within a variable edematous to collagenous matrix. Mitoses are not a prominent feature. The histopathology of this case revealed the characteristic findings.

Immunohistochemically, desmin expression was previously thought to be specific of AMF, this protein is no longer considered as a reliable marker for distinguishing the latter from AAM. However, it has been shown that AA shares immunophenotypic similarities with AMF. Hence, immunoreactivity does not appear to be helpful in distinguishing between these neoplasms.

The differential diagnosis of AMF also includes smooth muscle tumours, peripheral nerve sheath tumours, glomus tumour, chondroid syringoma, myxoid malignant fibrous histiocytoma, angiomyolipoma, spindle cell lipoma and myxoid liposarcoma. The distinction between AMF and these tumours has been described in detail elsewhere.

The recommended treatment is complete surgical excision of the mass with long-term follow-up examination, as local recurrence may occur many years after resection of the lesion. Rapid intraoperative pathologic diagnosis should be performed if possible, considering the possibility of diseases like AMF and aggressive angiomyxoma.

In summary, AMF is a rare benign mesenchymal tumour. Critical evaluation for AMF should be performed, mimicking that for AA, which requires the resection with wide tumour-free margins to prevent recurrence.

**REFERENCES**


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