Pak J Med Sci April - June 2003 Vol. 19 No. 2 132-135

Case Report

ANGIOMYOFIBROBLASTOMA OF THE VULVOVAGINAL REGION

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ABSTRACT

Objectives: To demonstrate the clinico pathological profile of four cases of Vulvovaginal Angiomyofibroblastoma.

Setting: Department of Pathology, Basic Medical Sciences Institute and Department of Obstetric and Gynecology, Jinnah Postgraduate Medical Centre, Karachi, Pakistan.

Subject: Four female patients with vulvo vaginal masses were studied.

Results: The microscopic and Immunohistochemical examinations confirmed that these vulvovaginal masses were Angiomyofibroblastoma distinct from aggressive angiomyxoma and characterized by a more indolent course.

Conclusion: Angiomyofibroblastoma of vulvovaginal region is a distinct entity differing from aggressive angiomyxoma and must be considered in the differential diagnosis of the vulvovaginal masses.

KEY WORDS Angiomyofibroblastoma (AMF), vulvovaginal.

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INTRODUCTION

Angiomyofibroblastoma was first described by Fletcher in 1992 as a well circumscribed subcutaneous vulvovaginal tumor histomorphologically distinct from the aggressive angiomyxoma and characterized by a more indolent course. Since the initial report of 38 cases only seventeen additional examples have been described in the literature. We report four cases of Angiomyofibroblastoma.The clinical presentation of these patients is as a painless mass in vulvo vaginal region, the duration of symptoms ranging from few months to few years.

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* *Received for publication:* October 3, 2002
Revision received: October 16, 2002
Revision accepted: January 6, 2003

CASE REPORT

We report four cases of AMF seen at the Department of Pathology, Jinnah Post Graduate Medical Center in the last five years (1993-1998).These patients (average age 29 years) presented with painless masses in the vulvovaginal region. The duration of symptoms ranged from two to six months. The masses measured from 3 cms to 28 cms in maximum diameter (Fig. 1). Grossly they were soft, well elineated. Cut section revealed myxedematous, gelatinous surface (Fig. 2).

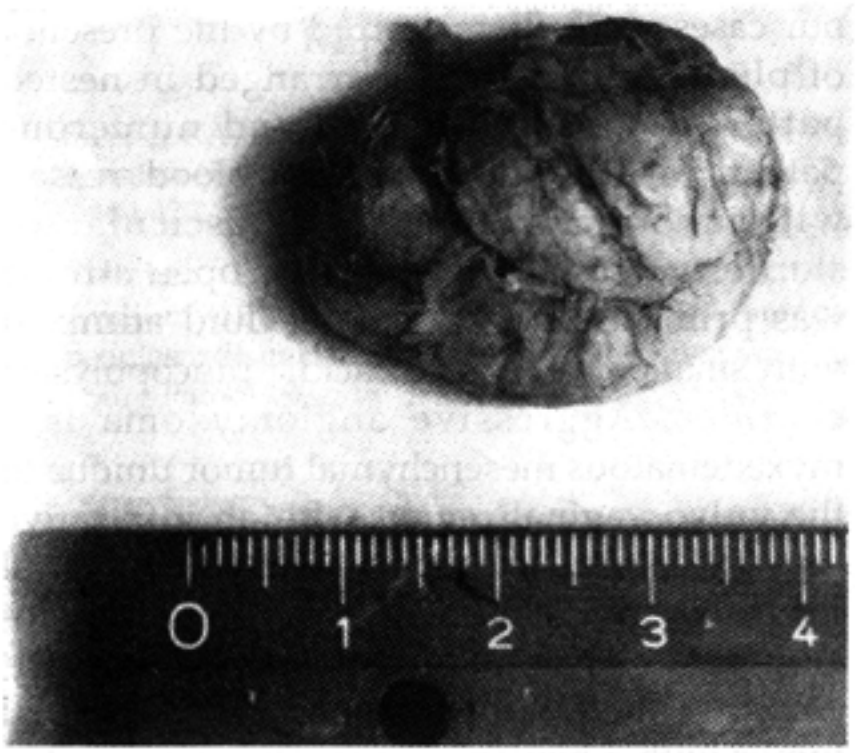

Figure 1: Gross appearance of the smallest vulval growth well-circumscribed measuring 3x3 cms.

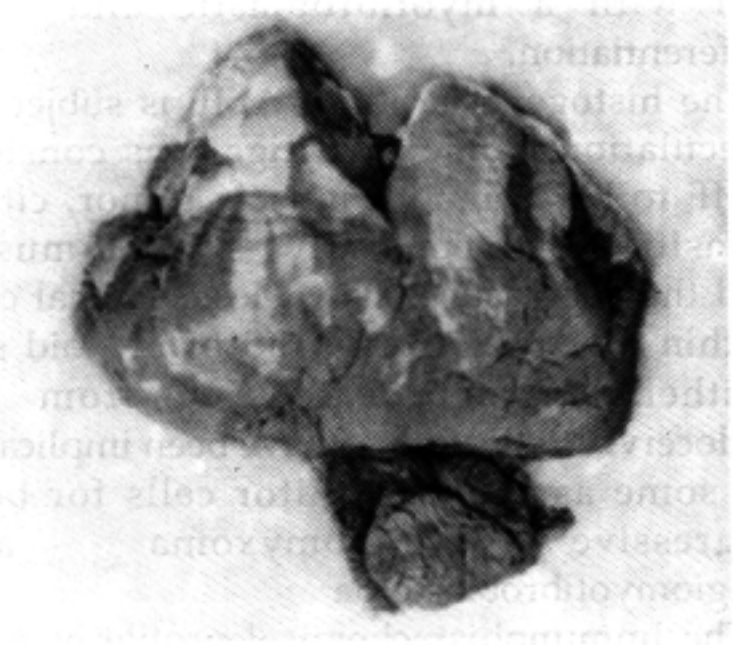

Figure 2: Gross appearance of the largest vulval growth covered by partly ulcerated skin.

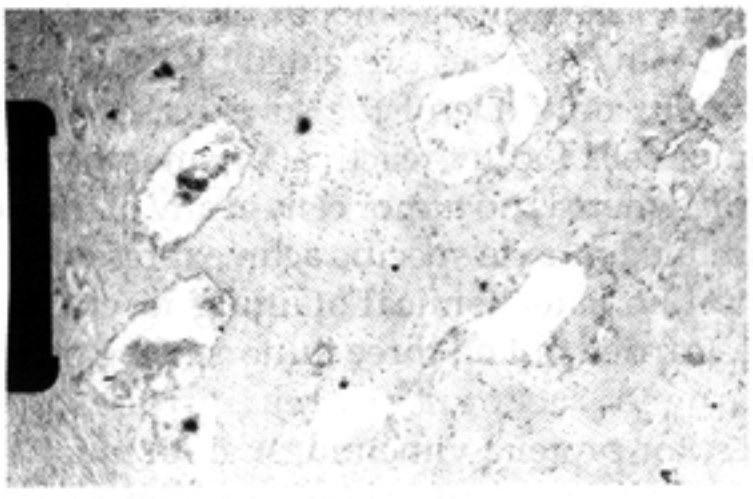

Figure 3: Microphotograph of the tumor showing spindled cells set in a myxedematous, loosely collalgenous stroma. (H&E x 40)

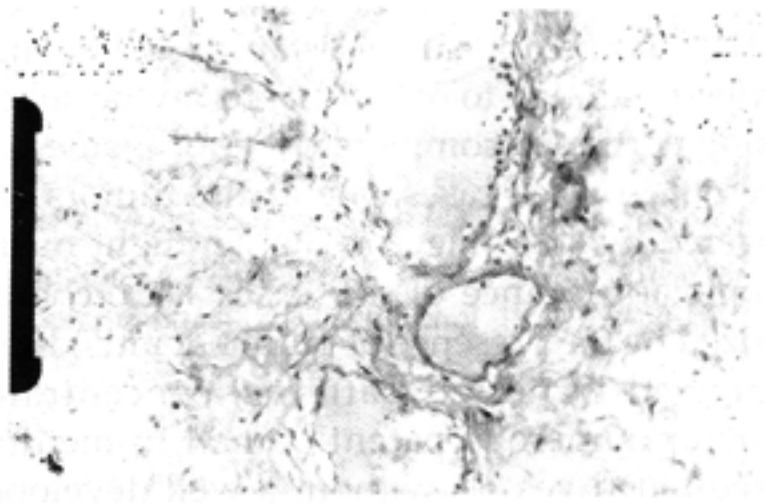

Figure 4: Microphotograph of the tumor showing small to medium sized vessels surrounded by spindled cells set in a myxedematous, loosely collagenous stroma.

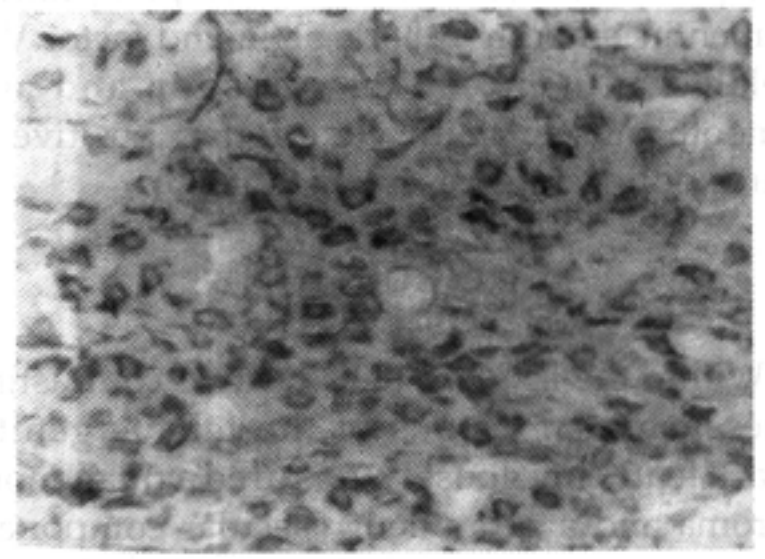

Figure 5: Microphotograph of the tumor showing positivity for Desmin(Desx 20)

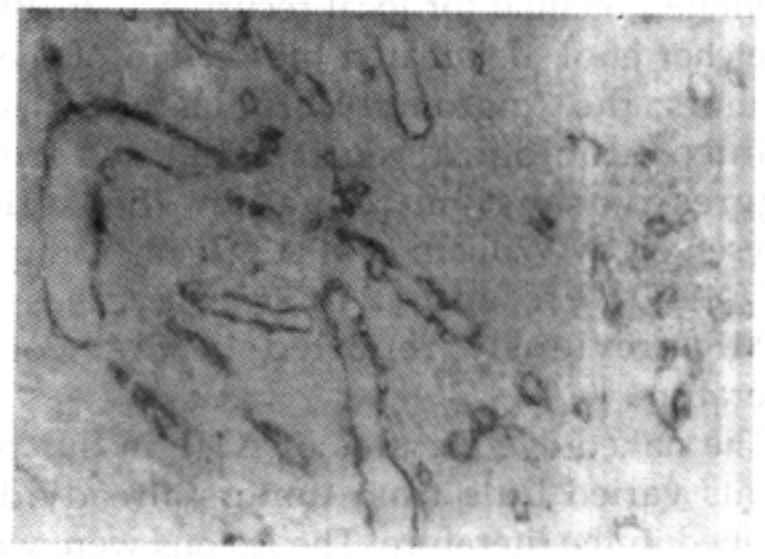

Figure 6: Microphotograph of the tumor showing positivity for Alpha smooth-muscle antigen (A.S.m.A.x10).

MICROSCOPICAL EXAMINATION

The tumors appeared as fairly well circumscribed rounded to ovoid masses in the subcutis. A partial or complete pseudocapsule was present in three cases.One of the tumors was covered by skin (Fig. 3). The key light microscopic appearance of our cases were characterized by the presence of plump spindled cells arranged in nested patterns, concentrated around numerous patent , small to medium sized blood vessels without a well developed fascicular or storiform growth pattern., set in a myxedematous, loosely collagenous stroma (Fig. 4).

Immunohistochemical staining was carried out on three tumors using desmin, estrogen receptor protein, progesterone receptor protien, vimentin, smooth -muscle actin and factor VIII related antigen markers. Desmin and anti smooth muscle actin were found positive in three out of four cases (Fig. 5 & 6).

DISCUSSION

Within the last two decades two contrasting mesenchymal lesions of the female genital tract featuring loosely textured collagenous stroma and a prominent vascular component have emerged in the literature. First Steeper and Rosai[1] described the aggressive angiomyxoma as a myxedematous spindle cell tumor of low to moderate cellularity having definite potential for local recurrence. In 1992 Fletcher et al[2] delineated the second of these tumors; the Angiomyofibroblastoma, as a clinicopathological entity distinct from aggressive angiomyxoma. In the series presented by William et al[3] 1997, 17 examples of AMF were added to the 38 cases of AMF previously reported in the world literature.

The naked-eye features of AMF in our patients varied little from tumors already described in the literature. The lesions were well circumscribed with a mean size of 13 cms and displayed a gelatinous or myxedematous cut surface. The light microscopic appearance of our cases were characterized by the presence of plump spindled cells arranged in nested patterns, concentrated around numerous patent, small to medium sized blood vessels without a well developed fascicular or storiform growth pattern. The open stroma was primarily due to edema fluid admixed with smaller amounts of acidic mucopolysaccharides. Aggressive angiomyxoma is a myxedematous mesenchymal tumor unique to the vulvo vaginal region with an infiltrative growth pattern and has a potential for local recurrence while Angiomyofibroblastoma including its lipomatous variant, is a well-circumscribed, subcutaneous vulvo vaginal tumor with a benign course. The origin of the tumor is proposed to be a perivascular stem cell with a myofibroblastic and fatty differentiation.

The histogenesis of the AMF is subject to speculation. Some investigations consider AMF to be a smooth muscle tumor, citing transition between native smooth muscle and the neoplastic cells.[4] Mesenchymal cells within the specialized band of myxoid sub epithelial stroma extending from the endocervix to the vulva have been implicated by some as the progenitor cells for both aggressive angiomyxoma and Angiomyofibroblastoma[5].

The Immunohistochemical profile of AMF as summarized from the literature includes the nearly uniform expression of vimentin and desmin and variable expression of muscle-specific actin, alpha-smooth muscle actin and CD 34. In the series presented by William et al in 1997[3], only one of the seven cases showed tumor cells focally reactive for smooth muscle actin, whereas no tumor cells in any of the cases exhibited muscle specific actin expression. The Immunohistochemical staining in our cases was carried out on three tumors using desmin, estrogen receptor protein, progesterone receptor protein, vimentin, smooth muscle actin and factor VIII related antigen markers. Desmin and anti smooth muscle actin were found positive in three out of four cases (Fig. 5,6).

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