VULVAR ANGIOMYOFIBROBLASTOMA

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Angiomyofibroblastoma is a rare tumor of the female genital tract. This vulvar tumor was first described by Fletcher et al in 1992 [1] as a mesenchymal neoplasm that is characteristically composed of blood vessels and stromal cells. It predominantly occurs in middle-aged women [2]. These tumors are usually found in the female genital tract, including the vulva, urethra, vagina and fallopian tubes [3–5]. Men are affected less frequently than women. It may be cured by complete local excision. Its histologic features distinguish it from aggressive angiomyxoma and malignant myxoid carcinoma.

Case 1 describes a 54-year-old woman who presented with a painless mass in the anterior aspect of the right labia majus. The lesion was noted to had increased in size gradually over several years. Her medical history was significant for hypertension, which was controlled with antihypertensive medications taken regularly for several years. An examination of the external genitalia revealed a mobile, non-tender mass at the superior aspect of the labia majora. The mass was rubbery and well circumscribed (Figure), measuring about 3 × 3 × 3 cm. The tumor was excised and was freed from its interior attachment. The skin was closed with no difficulty, and there had been no sign of recurrence. The excised tumor was yellowish to whitish in color, globular in shape, devoid of capsule, elastic in consistency, measuring 3.4 × 3.6 cm, and weighed 34 g. Pathology revealed a relatively circumscribed tumor with alternating hypocellular and hypercellular areas, manifested as proliferation of bland-appearing spindles or round/oval stromal cells with a tendency to perivascular concentration and prominent, irregular-shaped capillary vessels. Mitotic figures were absent. Immunohistochemical study showed that the stromal cells stained positive for vimentin and desmin but negative for S-100 protein, smooth muscle actin, CD34 and cytokeratin. These results indicated angiomyofibroblastoma.

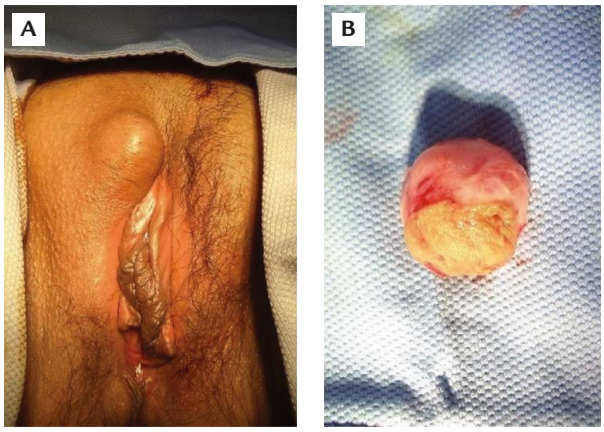

Figure. (A) The 3 × 3 × 3 cm tumor at right labia majora in Case 1. (B) The tumor after excision. Note the absence of capsule.

Case 2 describes a 50-year-old, gravida 5, para 3, abortus 2, who complained of a painless mass at the perineal area for 3 years. A pelvic examination disclosed a soft, 2.5-cm, non-tender swelling in the right labia majus, assumed to be lipoma. A local excision was performed for the lesion. The tumor was well delineated with minimal local infiltration. Her postoperative care was uneventful with no sign of recurrence after a year.

The tumor lesion was similar to that of the first case. It was whitish to yellowish in color, globular, and elastic. It measured 3.0 × 2.6 cm and weighed 30 g. It also shared several microscopic features with Case 1. The final diagnosis was angiomyofibroblastoma.

Angiomyofibroblastoma is a rare, benign, mesenchymal tumor of the vulva. So far, only 20 cases have been reported in the English literature [6]. The clinical features of this neoplasm are in the form of a vulvar mass that is most often misdiagnosed as a Bartholin gland or labial cyst, lipoma or hydrocele, etc. [7]. Microscopically, it consists of spindle cells with monomorphous and hyperchromatic nuclei and mesenchymal cells that surround small-sized blood vessels within a fibrous to myxoid mast cell containing stroma. Occasionally, mature adipose tissue is present in the lesion, and it is sometimes predominant.

The classification of vulvovaginal mesenchymal tumors and differential diagnosis of such lesions are crucial.

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Table. Variants of mesenchymal tumors

Benign tumor	Intermediate tumor	Malignant tumor
Angiomyofibroblastoma	Aggressive angiomyxoma	Myxoid carcinoma
Superficial angiomyxoma	Atypical lipomatous tumor	Myxoid embryonal rhabdomyosarcoma
Cellular angiofibroma	Pelvic fibromatosis	Myxoid liposarcoma
Fibroepithelial stromal polyp	Dermatofibrosarcoma protuberans	Myxoid leiomyosarcoma

The Table summarizes the variants of mesenchymal tumors, both benign and malignant types [2,8]. It is important to distinguish angiomyofibroblastoma from other mesenchymal tumors because of the differences in surgical treatment and clinical course. When intermediate or malignant tumors like angiomyxoma are diagnosed, more aggressive surgical approaches to treatment are required.

According to Horiguchi et al [9], a high mobility group protein I-C (HMGI-C) could be expressed in angiomyofibroblastoma. Meanwhile, basic fibroblast growth factor and stem cell factor may be related to vascularity and stromal–mast-cell infiltration. HMGI-C may be involved in the tumorigenesis of angiomyofibroblastoma [9].

Although rare, angiomyofibroblastoma should be considered during differential diagnosis of vulvar masses, especially in the vulvovaginal region. Differentiation from the more aggressive angiomyxoma is helpful for assessing recurrence risk.

The intermediate filaments, vimentin and desmin, have been recommended as useful markers to distinguish angiomyofibroblastoma from the aggressive angiomyxoma via immunohistochemistry [1]. A thorough evaluation of mesenchymal tumors such as angiomyofibroblastoma should be done whenever possible, because it may be mimicked by the aggressive angiomyxoma.

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