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Case Report

Angiomyofibroblastoma of the Vulva

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Angiomyofibroblastoma is a benign well-circumscribed tumor characterized by alternating hypocellular and hypercellular areas with abundant thin-walled blood vessels. The tumor cells are bland and spindle-shaped or epitheloid and tend to concentrate around the vessels or cluster in small nests. Herein, we report a case of angiomyofibroblastoma of the left labia major in a 50-year-old female. The tumor measured 18×16×11 cm and appeared as an ulcerated rubbery vulvar mass with rapid enlargement during one month. Ultrasonography demonstrated a soft tissue tumor with homogeneous echo and normal vascularity. Histologically, cells were positive for vimentine, desmin, and estrogen and progesterone receptors but negative for cytokeratin; all in favor of the diagnosis of angiomyofibroblastoma.

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Introduction

Angiomyofibroblastoma (AMFB) is a rare mesenchymal tumor predominantly occurring in the genital region of middle-aged women, especially in the superficial area of the vulva.[1] Clinically, most of the tumors 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.[2–5] Histologically, the tumors are well-circumscribed and` characterized by alternating hypo- and hypercellular areas with abundant thin-walled blood vessels. The tumor cells are bland and spindle-shaped or epitheloid, and tend to concentrate around the vessels or cluster in small nests.[4,6] Histopathologic differential diagnoses of the tumor include aggressive angiomyxoma, myxoma, mixoid lipoma, mixoid liposarcoma , and mixoid neural tumors.[4,7–10] Immunoreactivity for both desmin and vimentin is detected in almost all tumor cells, which also reveal estrogen and progesterone receptors, but staining for cytokeratin is negative.[11–13] Here, a case of AMFB is presented.

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Case Report

A 50-year-old female was referred to the Gynecologic Clinic of Yahya-Nezhad Hospital in Babol, Mazandaran Province because of a foul-smelling large painless ulcerated vulvar mass. The patient gave a history of small nodular masses in her vulvar region starting six years before, which had grown rapidly during the last month, and was now ulcerated with malodorous discharge. On physical examination, a huge pedunculated mass, measuring almost 20×15×10 cm with two ulcerated areas and purulent discharge was seen in the left labia major (Figure 1). On palpation, its consistency was not firm. Ultrasonography revealed a soft tissue tumor with homogeneous echo and normal vascularity. The patient underwent local excision of the tumor. The postoperative course was uneventful. No recurrence was seen during the eight-month follow-up period.

The resected tumor had a bag-like brownish soft appearance measuring 18×16×11 cm. The cut section surface was homogeneous and myxoid-like with focal areas of different consistency. Microscopically, the mass consisted of

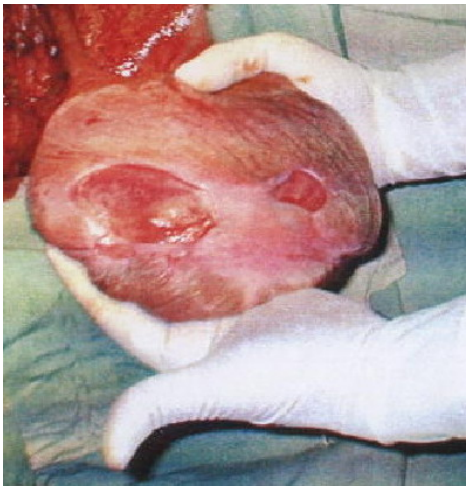

Figure 1. Angiomyofibroblastoma of the vulva.

fibroconnective tissue with abundant vessels of various wall thicknesses, no capsule or ulceration, and a few parts covered by stratified squamous epithelium. In cytology, the spindle-shaped cells showed moderate pleomorphism. No mitotic or atypical cells were seen and the stroma was edematous. All microscope fields were uniformly hypocellular. In immunohistochemistry, tumor cells were strongly positive for vimentin, desmin, and estrogen and progesterone receptors; it was, however, negative for cytokeratin; all in favor of the diagnosis of AMFB.

Discussion

We report a 50-year-old female with AMFB who presented with a huge and rapidly-growing mass in her left labia major. AMFB is a rare, distinctively benign mesenchymal tumor, which occurs mainly in the vulvar region of premenopausal women.[1] AMFB in women was first reported in 1992.[14] Furthermore, two cases of AMFB with perineal location in males were reported.[6,15]

Nielsen et al in 1996 analyzed 12 patients with AMFB in vulvar and vaginal regions. The tumors' mean size, in their largest diameter, was 4.7 cm (range: 0.9 – 11).[7] To the best of our knowledge, our patient had the largest tumor size reported to date.

It is important to suspect the diagnosis and to distinguish AMFB from aggressive angiomyxoma.[4,7,9] In histopathology, AMFB has a thin pseudocapsule, with typical features of a mesenchymal neoplasm, composed of bundle spindle cells with low cellular density, rich in collagen fibers, and thin-walled blood vessels.[4,6,9,14] Immunohistochemistry can also be helpful. Both estrogen and progesterone receptors are diffusely expressed in tumoral cells, suggestive of the sex-steroid-dependency of this tumor.

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.[1,2,9,16] Rapid intraoperative pathologic diagnosis should be performed if possible, considering the possibility of diseases like AMFB and aggressive angiomyxoma.[8,17] When aggressive angiomyxoma is suspected, the peripheral tissues should also be resected to prevent recurrence.[1,12,18]

To the best of our knowledge, this is the first case reported from Iran.

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