Angiomyofibroblastoma of the vulva: report of a case

F. Montella, M. Giana, A. Vigone, D. Surico, N. Surico

Department of Obstetrics and Gynaecology, "Piemonte Orientale" University, Novara (Italy)

Summary

A case of solitary angiomyofibroblastoma of the vulva in a 16-year-old woman was examined by histology and immunohistochemistry. Microscopic examination of the tumor revealed typical features of a mesenchymal neoplasm, composed of bundle spindle cells with low cellular density, rich in collagen fibers and thin-walled blood vessels. Immunohistochemistry revealed immunoreactivity for progesterone receptor, CD34 , desmin and vimentin, tumor cells expressing positivity, but not for estrogen receptors.

The stains for the muscle-specific actin and S-100 were negative. These results were mostly consistent with those of previous reports and suggest that the tumors cells were derived from primitive mesenchymal cells which occur normally in this region and show the potential for diverse lines of myoid differentiation.

Key words: Angiomyofibroblastoma; Desmin; CD34; Vulva.

Introduction

Angiomyofibroblastoma is a rare benign solid tumor in the expanding spectrum of benign soft tissue tumors that have been discovered, and it should be considered in the differential diagnosis of vulvar lesions [1].

This rare neoplasm is a distinctive clinicopathologic entity. It is a well-circumscribed tumor (0.5-12 cm in size) that generally arises in the superficial soft tissue of the vulva and vagina. Angiomyofibroblastoma predominantly occurs in middle-aged women. Patients usually present with a vulvar mass persisting for variable periods of time (2 weeks to 13 years), frequently misdiagnosed as a Bartholin's gland cyst. Microscopically, the typical tumor is characterized by high cellularity, numerous blood vessels, and plump stromal cells. The lesion can be confused at pathologic analysis with aggressive angiomyxoma due its location and cellularity. Since angiomyofibroblastoma does not recur after complete excision, it should be differentiated from other neoplasms of the vulva where radical surgical treatment is indicated. To our knowledge, more than 60 cases of angiomyofibroblastoma have been reported in the English literature [1-5]. Recently, these tumors have been described as existing in male inguinal areas and even in the scrotum [2].

We report here an additional, oligocellular case of angiomyofibroblastoma of the vulva which was subjected to immunohistochemical study.

Case Report

A 16-year-old woman, gravida 0 para 0, was admitted to our hospital complaining of a four-month history of painless swelling of the right labia majora, rapidly increasing in size. The examination of the external genitalia revealed a mobile, rubbery mass (3 cm in diameter) below the right labia majora (Figure 1). The tumor was immediately removed by means of local excision. The patient was healthy, with no recurrence one year after surgery.

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Pathologic examination revealed a 4.5 x 2.7 cm, soft and solid lesion, and circumscribed by a thin, slightly striated fibrous capsule. The tumor, at the naked eye, showd a homogeneous to slightly fascicolated, light gray cut surface. No foci of hemorrhage or necrosis were detected. As shown in Figure 2, histologic examination showed prevalent hypocellular and rare hypercellular areas. The great majority of the tumor cells were spindle or oval shaped. Scant plasmocytoid cells were also observed.

Nuclear atypicality was minimal and mitotic figures were not detected. Tumor cells were arranged individually in edematous stroma with wavy collagen fibers. The tumor contained thin-walled, small-sized blood vessels and numerous interspersed mast cells and very few intralesional lobules of adipose tissue. There was no entrapment of mucosal glands or nerve bundles. Immunohistochemistry of the tumor cells revealed diffuse reactivity for CD34, vimentin and, less intensely, desmin. The stains for α-smooth muscle actin and S-100 were negative; unusually, the tumor cells expressed positivity for progesterone receptors, but not for estrogen receptors.

The pathological diagnosis was angiomyofibroblastoma of the vulva, with unusual predominance of hypocellular areas.

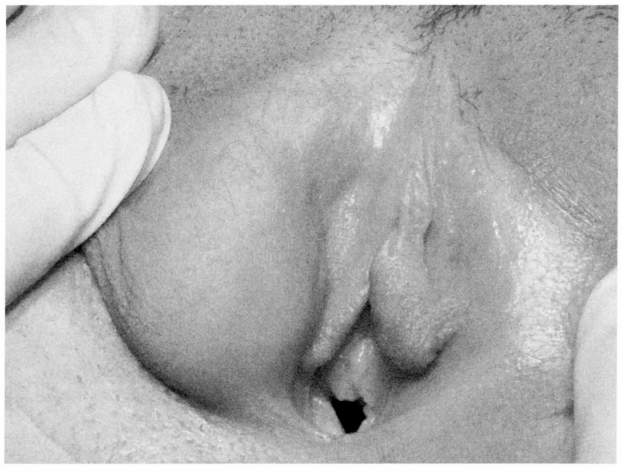

Figure 1. — Rubbery mass 3 cm in diameter below the right labia majora.

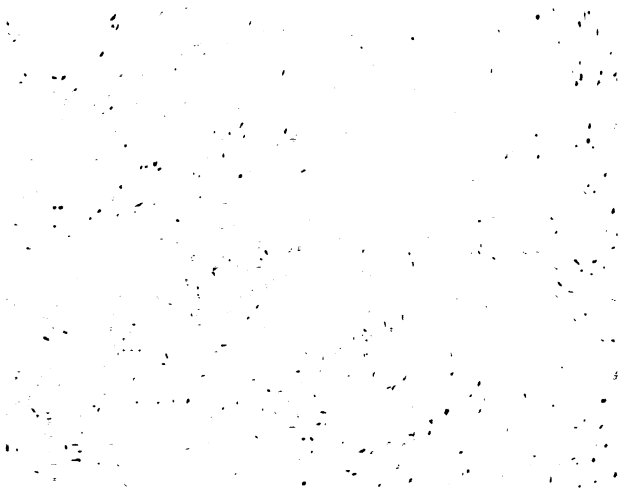

Figure 2. — Histologic examination shows prevalent hypocellular and rare hypercellular areas. The great majority of the tumor cells are spindle or oval shaped. Scant plasmocytoid cells can also be observed.

Discussion

Angiomyofibroblastoma is a well-circumscribed and apparently non recurring neoplasm that can be adequately treated by means of simple excision [1-3].

Since symptoms of benign and malignant conditions in the vulva area are frequently similar, early diagnosis and appropriate treatment are mandatory.

Histologically angiomyofibroblastoma can be confused with aggressive angiomyxoma, a distinctive locally infiltrative but non metastasizing neoplasm with a tendency to occur in the female pelvic and perineal region.

It is very important to distinguish these entities because of different surgical treatment and clinical survey they require.

The sharp circumscription of angiomyofibroblastoma contrasts with the infiltrative growth of aggressive angiomyxoma, which frequently entraps mucosal glands and nerves. Moreover, cellular density is normally higher in the former tumor than in the latter; in our predominantly hypocellular case, this was a doubtful point for pathological diagnosis. The vascular pattern presents as irregular, numerous, thin walled capillaries in angiomyofibroblastoma and as discrete, thin-to-thick walled, irregularly sized in aggressive angiomyxoma, in which perivascular actin-positive myoid bundles can also be observed. The paucity of stromal mucin in angiomyofibroblastoma also contrasts with the hyaluronic acid-rich stroma of aggressive angiomyxoma [5].

Aggressive angiomyxoma shows an immunophenotype that is vimentin-positive, MSA/α-SMA-variably positive, expecially in myoid bundles, and desmin-negative. This contrasts with the immunophenotype of the tumor cells in angiofibroblastoma [1], that is not so distinctive, but in most cases is desmin-, vimentin-, and laminin-positive, and α-SMA- negative. However, some are negative for desmin or positive for α-SMA.

These findings definitively support a myofibroblastic origin of angiomyofibroblastoma.

It is admittable that angiomyofibroblastoma in vulvar region derives from desmin-positive and α-SMA-negative primitive mesenchymal cells normally occurring in the subepithelial myxoid stromal zone, which extends from the endocervix to the vulva [5] and retains the potential for diverse lines of differentiation including myoid differentation [3].

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Address reprint requests to:
F. MONTELLA, M.D.
Dep. Obst./Gyn.
Maggiore della Carità Hospital
Corso Mazzini, 18
28100 Novara (Italy)