Angiomyofibroblastoma of the vulva

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Summary

Background: Angiomyofibroblastoma (AMF) is a rare benign mesenchymal neoplasm that arises in the pelviperial region. *Case:* A patient presented with a painless mass in the right vulva. Under the preoperative diagnosis of Bartholin cyst, she underwent a simple tumor excision. Pathological examination revealed an AMF. Immunohistochemical examination showed that tumor cells were positive for estrogen receptor, progesterone receptor, vimentin, and CD34. She has been with no evidence of local recurrence for ten months after surgery. *Conclusion:* AMF of the vulva is a distinctive mesenchymal tumor that is curable with a simple excision.

Key words: Angiomyofibroblastoma; Immunohistochemistry; Vulva.

Introduction

Angiomyofibroblastoma (AMF) is a benign, well-circumscribed myofibroblastic neoplasm that usually arises in the pelvic and peritoneal regions, especially in the vulva, of the middle-aged women. This tumor is often misdiagnosed as a Bartholin gland cyst, lipoma, and aggressive angiomyxoma. Despite the difficulty in the precise preoperative diagnosis, it is clinically important to distinguish AMF and aggressive angiomyxoma with a high propensity for local infiltration.

Here, the authors present a case of AMF arising in the vulva with the findings of pathological and immunohistochmical analyses.

Case Report

A 42-year-old woman presented with a painless mass in the right vulva which she had noticed ten months ago. Physical examination revealed a ruby mass arising from the right labia majora with no tenderness, measuring six cm in the diameter. She was diagnosed as having a Bartholin cyst and underwent a simple excision. She has been with no evidence of recurrence for ten months since surgery.

Macroscopically, the excised tumor was well-circumscribed, elastic soft, and 63 x 43 x 19 mm in size. The cut surface was solid, yellowish-white in color, and homogeneous with no areas of hemorrhage or necrosis. Microscopic examination revealed AMF of the vulva. Tumor was well demarcated by a thin fibrous pseudocapsule and showed hypocellular areas with an abundant, loose edematous stroma in the periphery (Figure 1A), and collagenous, hypercellular areas in the central (Figure 1B). In the hypocelluar areas, thin-wall blood vessels were distributed in the reticular pattern, and thin wavy collagen fibers were scattered. Tumor cells were composed of primitive spindle-to-stellate cells with scanty cytoplasm and spindle-to-round cells with eosinophilic cytoplasm including multinucleated cells. Typically, tumor cells were concentrated around vessels (Figure 2A) and clustered with an epithelioid appearance (Figure 2B). Occasionally, mitoses were found (Figure 2C). Furthermore, mature adipocytes were focally seen (Figure 2D). No mucin in the edematous stroma was stained with Alcian blue at pH 2.5.

A panel of immunohistochemical analysis with estrogen receptor (ER), progesterone receptor (PR), vimentin, CD34 , α-smooth muscle actin (SMA), desmin, H-Caldesmon, S-100, and AE1/AE3 was performed using a streptavidin-biotin method. The majority of tumor cells were positive for ER（Figure 3A), PR (Figure 3B), and vimentin (Figure 3C), and spindle-to-stellate cells in the edematous stroma were weakly positive for CD34.

Discussion

AMF is an uncommon benign mesenchymal tumor that usually occurs in the female genital tract. By reviewing 71 cases of AMF, Sims *et al.* summarized that the mean age of AMF at presentation was 45 years and that the lesions were equally distributed between the left (52%) and right (48%), with the mean diameter being 5.9 cm [1]. The most common diagnosis is a Bartholin gland cyst (46%) or lipoma (15%) [1]. The differential diagnosis includes aggressive angiomyxoma, cellular angiofibroma, fibroepithelial stromal polyp, and epithelioid leiomyoma [2]. It is crucial to distinguish AMF from aggressive angiomyxoma because aggressive angiomyxoma has a marked tendency to local recurrence [3]. The treatment of choice of AMF is a simple total excision [1], while the wide surgical excision with tumor-free margin is the traditional treatment of choice in aggressive angiomyxoma [4].

In the present case, microscopic findings were in agreement with the reported histological features of AMF [5]. Histological findings of AMF are characterized by the presence of variable hypocellular and hypercellular areas, edematous stroma containing wavy collagen fibers, tumor cells varying in shape from spindle, stellate, plasmatoid to epithelioid appearance, isolated or grouped tumor cells in cords or nests typically around blood vessels, the presence of small- to medium-sized and dilated vessels, the mixture of lipomatous elements in a few cases, and minimal mitotic activity [1, 5-12].

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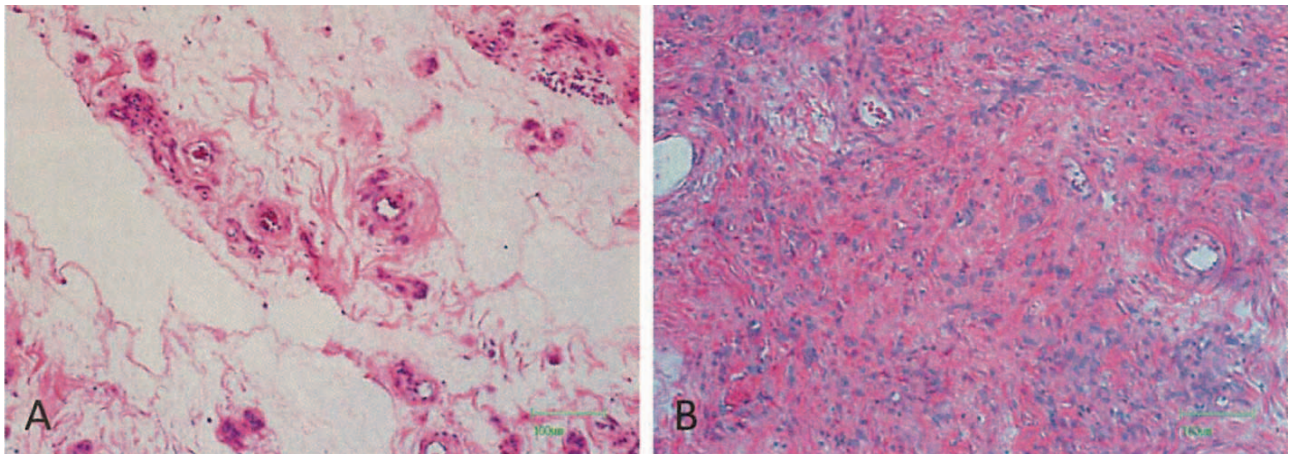

Figure 1. — Microscopic findings of AMF. (A) Hypocellular area in the edematous stroma. (B) Hypercellular area in a dense collagenous stroma (H&E stain, original magnification x10).

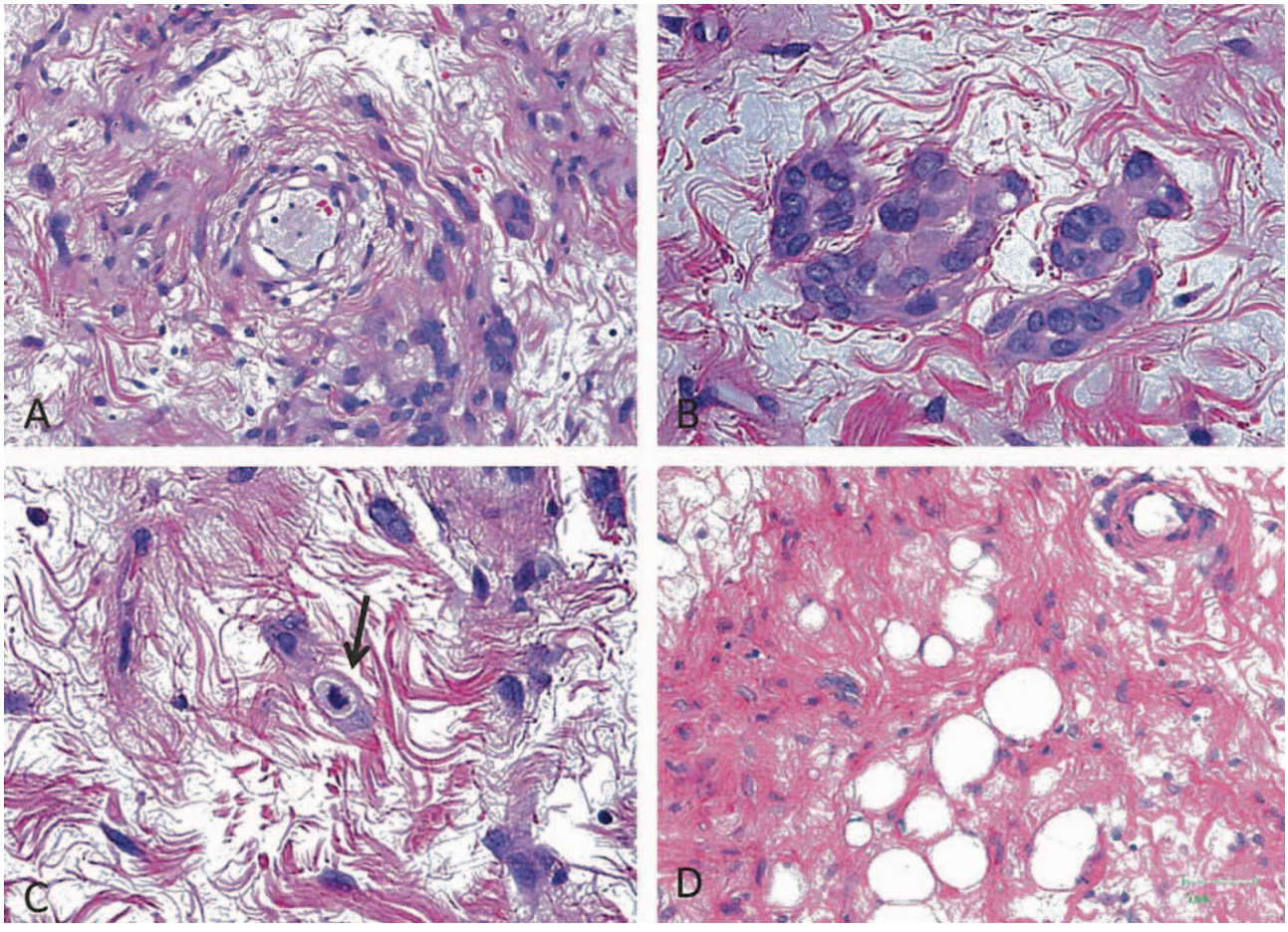

Figure 2. — Microscopic findings of AMF. (A) The concentration of the tumor cells was seen around a degenerated vein. (B) The tumor cells were clustered. (C) Mitosis was seen (arrow). (D) Mature adipocytes were focally contained in the tumor (H&E stain, original magnification (A)x20, (B)x40, (C)x40, (D)x20).

AMF mimics aggressive angiomyxoma due to the presence of spindle-shaped stromal cells, myxoid stroma, and abundant blood vessels [2]. However, Fletcher *et al.* described that AMF can be distinguished from aggressive angiomyxoma by its circumscribed borders, much higher cellularity, more numerous blood vessels, frequent presence of plump stromal cells, minimal stromal mucin, and rarity of erythrocyte extravasation [5].

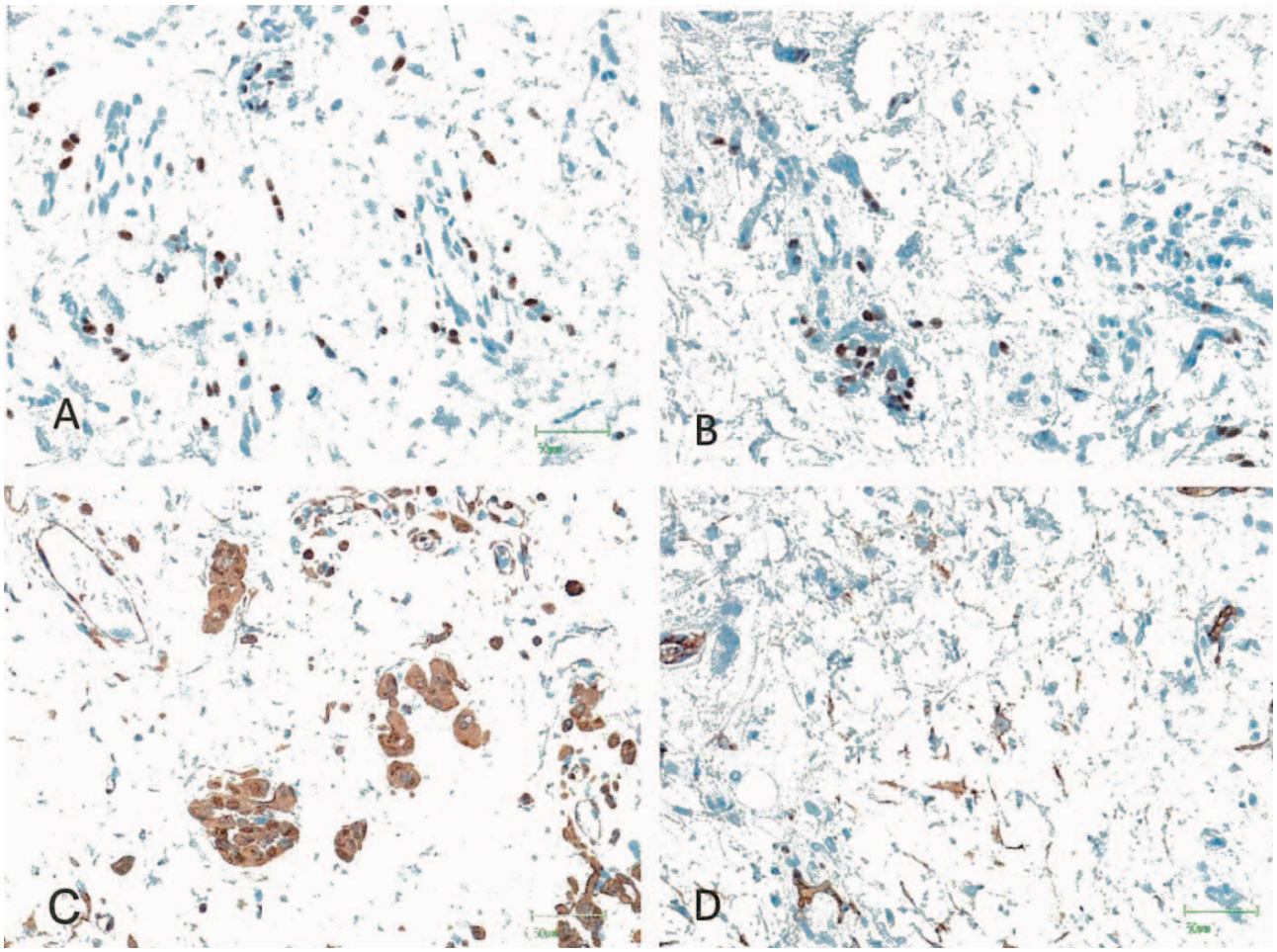

Figure 3. — Immunohistochemical staining of AMF. The tumor cells are positive for (A) ER, (B) PR, and (C) vimentin, and small spindle-to-stellate cells and endothelial cells are positive for (D) CD34 (original magnification x20).

In the present case, immunohistochemical analysis showed that tumor cells were positive for ER, PR, vimentin, and CD34. Previous reports have demonstrated that AMF is strongly positive for vimentin and desmin, and usually expresses ER and PR, but is negative for cytokeratin, α-SMA, and S-100 protein [5-8, 11-14]. In contrast, aggressive angiomyxoma was shown to be positive for vimentin [15,16], α-SMA [15, 16], ER [16], and PR [16], but negative for S-100 and desmin [15, 16]. Nagai *et al.* speculated that AMF and aggressive angiomyxoma are derived from the stromal stem cell with a capacity for hormone-inducing myofibroblastic differentiation [7]. Unlike microscopic evaluation, the differential diagnosis between AMF and aggressive angiomyxoma with immunohistochemistry seems to be difficult because of the overlapping of immunostaining pattern.

Collectively, AMF is a rare mesenchymal tumor that occurs mainly in the vulva. Histological evaluation is important to distinguish AMF from aggressive angiomyxoma with a propensity for local recurrence.

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