A case of Angiomyofibroblastoma

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Abstract: Angiomyofibroblastoma of the vulva is a rare mesenchymal tumor. It is characterized by superficial and slow growth, low propensity for local recurrence and often misdiagnosed as a Bartholin's gland cyst and aggressive angiomyxoma. We report a case of angiomyofibroblastoma of the vulva in a 23-year-old female patient. She presented with two lobulating left vulvar masses for about 2 years. Initially the mass was small and reminiscent of a Bartholin's cyst but grew rapidly during the recent two months. Grossly the tumor consisted of large two lobulating masses with superficial ulceration.

Microscopically there were alternating hypercellular and hypocellular edematous zones, in which abundant capillary blood vessels were noted. Immunohistochemically the stromal cells showed positive reaction for vimentin, desmin, and smooth muscle actin.

Key Words: Angiomyofibroblastoma, Vulva

Angiomyofibroblastoma of the genital region is a relatively recently described tumor of the superficial soft tissues with a marked preference for female patients. Since symptoms of benign and malignant conditions in the vulvar area are similar, early diagnosis and treatment is important.

This report presents several interesting features of an angiomyofibroblastoma of the vulva with respect to the origin of the tumor and histologic and immunohistochemical findings.

Case report

A 23-year-old woman having a vulvar mass over a period of two years visited our clinic in February 2001. The mass was initially confused with a Bartholin's cyst but grew rapidly during the recent one month to have about 7 to 8 times the size of that at the initial diagnosis. The genital examination showed no other abnormal findings and the tumor located subcutaneously in her left labial region consisting of two lobulated masses. It was well defined, soft to cystic in consistency, mobile, nontender, and superficially ulcerated (Fig. 1). The patient had no remarkable previous medical record and no history of medication. General physical and abdominal examination revealed no abnormalities.
A vaginal examination showed normal findings without an adnexal mass. There was no inguinal lymphadenopathy. We did not check pelvic CT or MRI before removal of the mass.

We performed a simple excision of the mass (Fig. 2). The whole vulvar mass was excised and silastic drain was applied (Fig. 3).

Postoperative course of the patient was uncomplicated and there is currently no evidence of recurrence one year after surgery.

The tumor measured 16.0 × 15.5 × 7.0 cm in size and 425.0 g in weight. The cut surface showed gray-to-pinkish solid appearance with variable sized cystic spaces and also showed myxoid appearance simulating leiomyoma with myxoid degeneration (Fig. 4).

On microscopic examination the tumor showed alternating hypercellular and hypocellular edematous areas containing small vessels. The tumor cells were spindle-to-oval cells in shape with bland nuclei and showed a tendency to perivascular aggregation (Fig. 5, 6). The cells were immunoreactive to vimentin, desmin, and smooth muscle actin (Fig. 7).

**Discussion**

Angiomyofibroblastoma is a rare, recently
described, soft tissue tumor that occurs mainly, but
not exclusively, in the vulvar area.\textsuperscript{2} As in our case,
patients are usually premenopausal and present with
a vulvar mass initially misdiagnosed as a Bartholin’s
gland cyst. Angiomyofibroblastoma should be
differentiated from other neoplasms of the vulva in
which radical surgical treatment is indicated. Local
excision of the tumor is the treatment of choice.\textsuperscript{4}

Except for one case with malignant transformation
of angiomyofibroblastoma in an 80-year-old
woman, there are currently no published reports of
local recurrence or metastatic disease of
angiomyofibroblastoma in the literature.\textsuperscript{4}

The most important differential diagnosis of
angiomyofibroblastoma is aggressive angiomyxoma
(Table 1). Angiomyofibroblastoma is distinguished

| Table 1. Comparison of angiomyofibroblastoma with aggressive angiomyxoma\textsuperscript{2} |
|-----------------|-----------------|-----------------|
| **Clinical features** | **Angiomyofibroblastoma** | **Aggressive angiomyxoma** |
| Age | 25-54 yrs (mean 36.3 yrs) | 18-63 yrs (mean 32.1 yrs) |
| Presentation | Vulvar mass | Vulvar/pelvic mass |
| Duration of symptoms | 10 weeks - 8 years | Usually a few months |
| Location of lesion | Vulva | Vulva, vagina, perineum or pelvic soft tissues |
| size of lesion | 0.5-12 cm (usually < 5 cm) | 3-60 cm (usually > 5Ø) |
| Behavior | No recurrence after simple excision | Local recurrence in >70% of cases, often within 2 years (but sometimes delayed) |
| **Pathology** | | |
| Borders | Well circumscribed | Infiltrative; at most partly circumscribed |
| Blood vessels | More numerous: mostly capillaries, with some thin-walled ectatic/cavernous veins | Small to medium-sized vessels, many of which are thick-walled or hyalinized |
| Stromal cells | More abundant and with perivascular accentuation; wavy spindle, plump spindle, oval, and multinucleated giant cells | Low cellularity; stellate or spindled stromal cells, with delicate cytoplasmic processes |
| Stroma | Edematous to collagenous; extravasation of erythrocytes uncommon | Myxoid to collagenous, often with extravasation of erythrocytes |
| Immunostaining | Positive | Positive |
| Vimentin | Negative | Variable; usually negative |
| MSA/ASMA | Positive | Negative |
| Desmin | | |

from aggressive angiomyxoma by its well
circumscribed border and higher cellularity, by the
frequent presence of plump stromal cells, and by a
lesser degree of stromal myxoid change.\textsuperscript{8} In contrast
to angiomyofibroblastoma aggressive angiomyxoma
is a deeply seated tumor with an infiltrative growth
pattern which frequently results in entrapment of
mucosal glands and nerves.\textsuperscript{8} In the present case
immunohistochemical staining showed the tumor
cells were positive for desmin, vimentin, and smooth
muscle actin.

The size of the angiomyofibroblastoma in the
vulvar area varies from 0.5cm to 12cm, however in
our case, the tumor measured 16cm in its greatest
dimension and thought to be the largest tumor ever
reported.\textsuperscript{9}

Angiomyofibroblastoma is relatively a newly
recognized tumor on the expanding spectrum of
benign soft tissue tumors and should be considered
in the differential diagnosis of vulvar lesions in
elderly postmenopausal patients. Differentiation of
the tumor from other diseases such as aggressive
angiomyxoma, where more aggressive surgical
approaches are required, is mandatory.\textsuperscript{9}
Reference


■ 국문 요약 ■

외음부의 혈관근섬유아세포종(angiomyofibroblastoma)는 비교적 드문 중간엽성 종양이다. 혈관근섬유아세포종은 표재성이며 느린 성장을 보이고 국소 재발이 적은 경향이 있지만 종종 바르톨린샘ynthia(Bartholin's gland cyst)와 친근성 혈관섬유종(AGGRESSIVE ANGIOMYXOMA)으로 오인하는 경우가 있어 감별을 요한다. 최근 저자들은 23세 여성의 외음부에서 발생한 혈관근섬유아세포종을 경험하였는데 환자는 약 2년간 두 개의 분엽을 가진 왼쪽 외음부 종괴를 호소하였다. 처음에는 바르톨린샘 양종으로 생각하였으나 최근 2개월간 빠르게 성장하였으며 옥인적으로 보았을 때 종양은 표재성 세포를 동반한 큰 두 개의 분엽을 이루고 있었다.

광학 현미경 소견상 종양내에는 풍부한 모세혈관과 함께 주위 세포밀도가 다르게 관찰되는 부위가 있었다. 면역조직화 학적 면에서 종양의 기질세포는 vimentin, desmin, smooth muscle actin에 양성을 보였다.

핵심단어 : 혈관근섬유아세포종, 외음부