

외음부 혈관근섬유모세포종의 지방종 아형

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Lipomatous variant of angiomyofibroblastoma on the vulva: A case report

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Lipomatous variant of angiomyofibroblastoma is a rare tumor of the vulva. Most of cases are usually premenopausal and present with a slowly growing mass without pain. A 46-year-old woman presented with a swelling of the left labium major. The lesion was thought to be a Bartholin's gland cyst or a lipoma preoperatively and a local excision was performed. The tumor was well demarcated and rubbery. On microscopic examination, the alternating hypercellular and edematous hypocellular component with abundant vessels was noted. A large amount of adipose tissue was scattered in the lesion. Stromal cells were positive for vimentin, estrogen receptor, and progesterone receptor and negative for desmin antibody immunostaining. The local excision was enough for complete treatment. No recurrence was noted for 7 months after surgery.

Key Words: Vulva, Angiomyofibroblastoma, Lipomatous variant

Angiomyofibroblastoma (AMFB) is a rare, benign tumor of the vulva and shows a distinctive, well-circumscribed myofibroblastic feature. The tumor was firstly reported by Fletcher et al.¹ in 1992. The lesion is mainly composed of conspicuous blood vessels and stromal cells. Interestingly, intralesional adipose tissue in AMFB was reported in only six cases (i.e. lipomatous variant).²

We present herein the case of lipomatous variant of

AMFB in 46-year-old woman complaining of a left vulva mass.

Case Report

A 46-year-old woman, gravida 1 para 1, presented with a 2-year history of a swelling of the left vulva. The lesion had slowly increased in size. On gross examination the mass was easily movable, not adhered to adjacent tissue, and the size was 4×3×2 cm (Fig. 1A). On computed tomography (CT), highly enhanced mass was noted on the left vulva (Fig. 1B). On ultrasonogram, 4.2 cm sized subserosal myoma of the uterus and 2.2 cm sized right ovary cyst were noted. An

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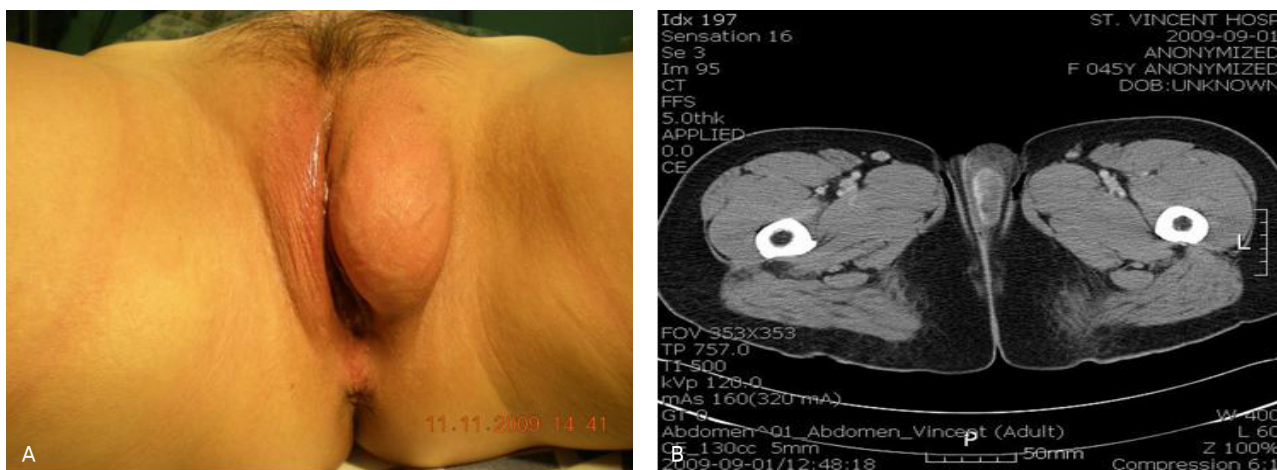


Fig. 1. Gross appearance of the left vulva and image of computer tomography. (A) The mass was soft and movable. (B) The mass with high enhancement on the left vulva did not infiltrate the adjacent tissue.

intrauterine device was located in the uterus. No enlarged lymph node was palpated on the bilateral inguinal area. Abnormal result was not detected on laboratory test.

During the operation, there was no infiltration of tumor into deeper soft tissue and the tumor had a distinct border from the adjacent tissue. No recurrence was detected for 7 months after surgery.

1. Gross findings

The tumor was 4 cm in maximal diameter, well circumscribed, and found in the subcutis. The surgical specimen was a soft, elastic consistency without encapsulation. Neither necrosis nor hemorrhage was found. The surface had a homogeneous, glistening appearance with yellow to pinkish color (Fig. 2).

2. Microscopic findings

The specimen was embedded in paraffin after formalin fixation and slide sections were stained with hematoxylin and eosin. The prominent vascular pattern was noted in the mixture of hypercellular and hypocellular component. The tumor cells were aggrega-



Fig. 2. The surgical specimen of the vulva. The size of mass was 4×3×2 cm sized and showed soft, gelatinous feature.

ted in cords and nests predominantly around vessels in the edematous hypocellular matrix. The small to medium sized vessel had a dilated and thin wall. The tumor cells, round to spindle shaped, had eosinophilic cytoplasm and ovoid nuclei with uneven chromatin arrangement. No significant atypism was noted on nucleus. A lot of adipocytes with plump nucleus were scattered in the matrix (Fig. 3).

In immunohistochemistry staining, the stromal cells showed strong positivity for vimentin, estrogen receptor, and progesterone receptor. The tumor had no reactivity for desmin (Fig. 4).

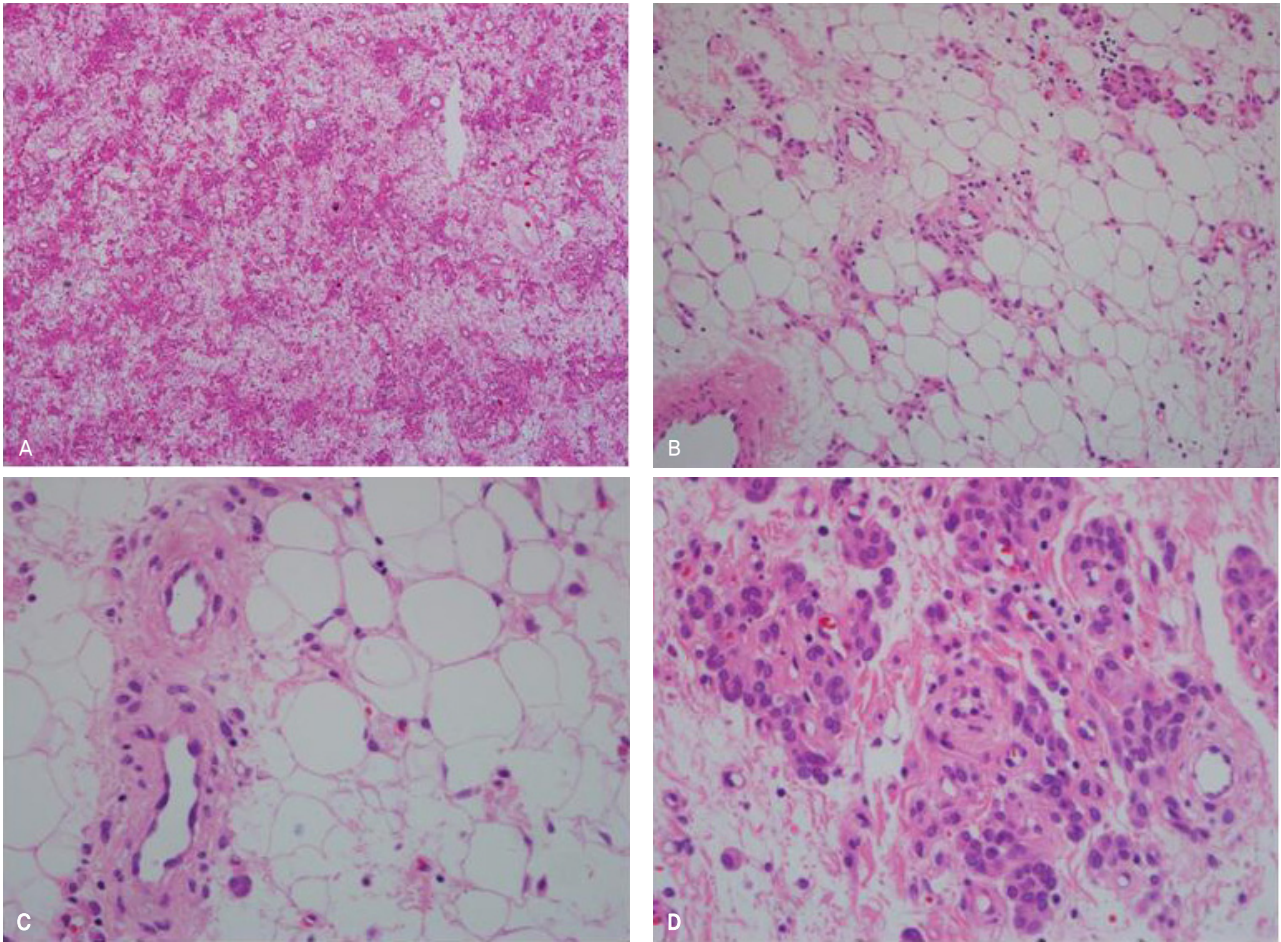


Fig. 3. Histologic findings of the vulva mass. (A) The lesion of the vulva had irregular mixture of hypercellular and hypocellular area with prominent blood vessels (H&E stain, $\times 40$). (B) Majority of cells had oval or spindle appearance with an eosinophilic cytoplasm. A lot of adipocytes were scattered. Vessels were encircled with round tumor cells and located in a loose edematous matrix (H&E stain, $\times 200$). (C) Small to medium sized vessels and adipocytes were noted (H&E stain, $\times 400$). (D) Tumor cells were plasmacytoid or epithelioid and multinucleated cells were noted (H&E stain, $\times 400$).

Discussion

AMFB occurs frequently in young-to-middle-aged premenopausal women.^{3,4} The tumor arises in perineal subcutaneous tissue and is found predominantly in the vulva.^{5,6} Most of the cases exhibit painless mass of the vulva and slowly growing feature. The typical characteristics of AMFB are as follows: 1) irregular mixture of hypercellularity and hypocellularity area; 2) absence of stromal mucin.^{1,7}

In the present case, adipose tissue was noted among

the hypercellular and hypocellular area. The morphologic spectrum of AMFB is expanded to the lipomatous variant and the perivascular stem cell can be an origin of myofibroblastic and fatty differentiation.^{2,4,8} Only one case of lipomatous variant of AMFB was reported in men.⁹ A location and a size of the tumor of our case showed similar patterns, compared with 6 cases of the lipomatous variant in the literature.² Differential diagnosis of lipomatous variant of AMFB includes Bartholin's gland cyst, liposarcoma, phleomorphic sarcoma, spindle cell lipoma, aggressive angiomyxoma in vulva lesion.^{2,10}

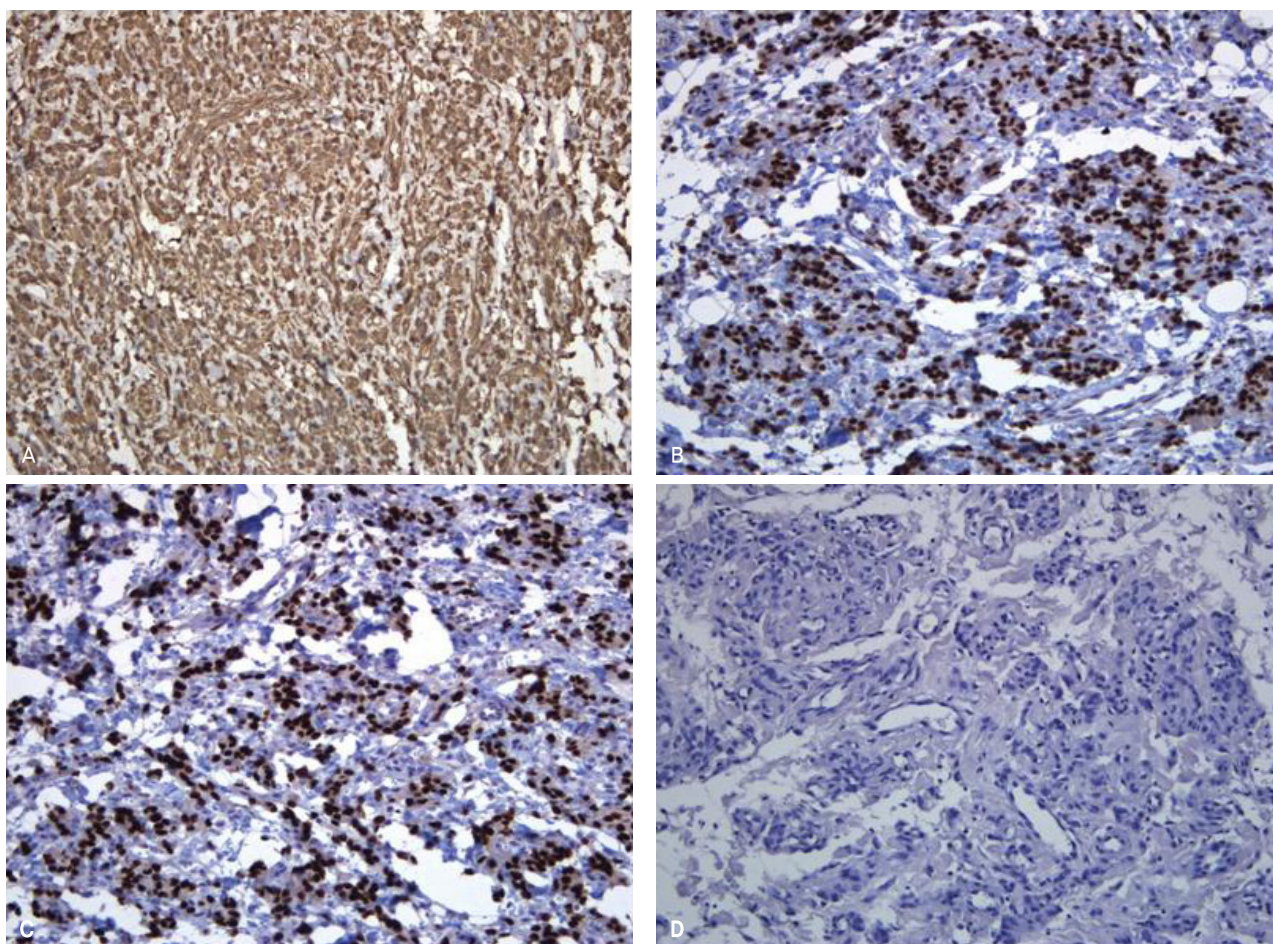


Fig. 4. Immunohistochemistry findings. (A) positive for vimentin, (B) positive for estrogen receptor, (C) positive for progesterone receptor, (D) negative for desmin.

Because a benign condition of the vulva is very similar to that of malignancy, the primary diagnosis is very significant. Importantly, differential diagnosis is needed to rule out the aggressive angiomyxoma (AAM) in clinical situation. AAM is likely to recur locally in case of incomplete resection of the tumor.¹¹ AMFB including lipomatous variant is frequently found at the superficial area of the vulva, while AAM infiltrates into the deeper tissue and has a greater tendency to recur.¹¹ AMFB is well-circumscribed and not encapsulated and is generally smaller than 5 cm.⁷ AAM is larger than 5 cm and poorly-circumscribed.¹² The stromal cells of AMFB are plump and oval shape, by comparison with that of AAM, which are spindle and

stellate-shape.^{1,11} Once in a while, AMFB has a plasmacytoid appearance which explains a round nucleus in the plentiful cytoplasm.^{4,6}

According to the result of immunohistochemical staining, all cases of AMFB were positive for vimentin, 92% were positive for desmin.¹⁰ It is reported that myofibroblast can be characterized by the composition of intermediate filaments protein such as vimentin and desmin.¹³ The strong positivity for vimentin and desmin explains a myofibroblastic origin of AFMB.¹⁰ Also, lipomatous variant of AFMB shows positivity for vimentin, ER and PR in immunohistochemistry.² In our case, immunohistochemistry showed vimentin-positive and desmin-negative.

To our knowledge, only eight cases of AMFB have been reported in Korean literature.¹⁴ And one recurrence was described.¹⁵ It is known that the prognosis is good and local excision with clear margin is enough for the treatment.¹⁶ Our patient has no recurrence 7 months after surgery.

In conclusion, lipomatous variant of AMFB should be considered in a differential diagnosis of vulva disease and immunohistochemistry can be helpful for understanding of the disease. We herein present AMFB with adipose tissue occurring in premenopausal woman.

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= 국문초록 =

혈관근섬유모세포종의 지방종 아형은 외음부에 발생하는 드문 종양으로 대부분 폐경 전 여성에서 통증 없이 서서히 자라는 종괴 양상을 보인다. 본 증례에서 46세 여성은 좌측 대음순 종창으로 불편감을 호소하였다. 수술 전 임상적 진단은 바톨린 낭종 혹은 지방종으로 의심되어 단순 절제술을 시행하였다. 육안적 소견으로는 경계가 명확하고 부드러운 표면을 가진 종괴였으며, 현미경적 소견으로는 기질세포가 많은 부분과 적은 부분이 혼재된 가운데 혈관이 발달하였다. 혈관 주위에 간질세포들이 밀집해 배열을 이루고 있다. 많은 지방 조직이 현미경 소견에서 관찰되었다. 본 증례에서 시행한 면역 염색 검사에서 vimentin, estrogen 수용체, progesterone 수용체에 양성 반응을 보였으나, desmin 면역 염색에는 음성 소견을 보였다. 본 저자들은 단순절제술을 시행하였고, 수술 후 7개월 동안 재발 소견이 없어 이를 문헌고찰과 함께 보고하는 바이다.

중심단어: 외음부, 혈관근섬유모세포종, 지방종 아형