

# Aggressive angiomyxoma of the vulva: A case report

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Aggressive angiomyxoma (AA) is an unusual mesenchymal tumor. AA occurs most commonly in women of reproductive age and is located in the perineal or pelvic region. This is a distinct soft tissue tumor that has a prominent myxoid matrix and numerous thin-walled blood vessels and may have an aggressive local recurrence. The tumors have the characteristics of large size (usually greater than 10 cm) and slow growth, and are not painful. The standard treatment for AA is total excision and close follow-up. We announce a case of a 35 year-old female presenting with a pedunculated AA on the right labium majora that has not relapsed for seven years.

**Keywords:** Myxoma; Mesenchymal tumor; Vulva

## Introduction

Aggressive angiomyxoma (AA) is a rare, locally aggressive myxoid mesenchymal neoplasm arising in the pelvis and perineal regions. AA was first described in 1983 by Steeper and Rosai [1]. They presented AA as a vulvar polyp clinically and diagnosed it histologically. It usually occurs in women, especially middle-aged; 95% of total cases are found in females [2]. The term "aggressive" denotes its propensity for local aggression and recurrence after excision. However, AA tends to grow slowly with a low tendency to metastasize. Up to now, there has been no complete consensus on its pathogenesis.

## Case report

A 35-year-old female presented with a slow-growing mass on the right labium majora. She visited the clinic because she felt uncomfortable as the size of mass had grown. The patient was referred to gynecological services for further evaluation with considerations of a fibroepithelial polyp, a vulvar fibroma or a giant acrochordon. Local examination showed a well-circumscribed pedunculated polypoidal mass measuring about 10 × 7 cm. The mass was non-tender, soft and spongy in consistency. The inguinal lymph nodes were not enlarged. At first, we supposed the mass to be vulvar hemangioma and performed a pelvis magnetic resonance imaging (MRI). The pelvis MRI revealed findings that were compatible with those of AA in the subcutaneous layer of the right labium majora. A

well-defined mass-like lesion of about 10 × 7 cm was seen in the subcutaneous layer of the right labium majora. The mass showed bright signal intensity on T2WI and demonstrated delayed enhancement after contrast administration (Fig. 1A). The patient underwent local excision of the tumor with ligation of the stalk. The tumor mass was about 10 × 7 cm and it was soft and pink-tan colored (Fig. 1B). Microscopically, the tumor was composed of spindle and stellate-shaped cells embedded in a loose myxoid matrix. These cells showed low to moderated cellularity and had eosinophilic cytoplasm with no significant nuclear pleomorphism and mitosis. Variable-sized thin-walled capillaries and thick-walled vascular channels were haphazardly arranged in the stroma. Some of these vessels showed perivascular hyalinization in the vascular walls (Fig. 2A). Immunohistochemical staining of the tumor was positive for estrogen receptor (ER), progesterone receptor (PR), vimentin

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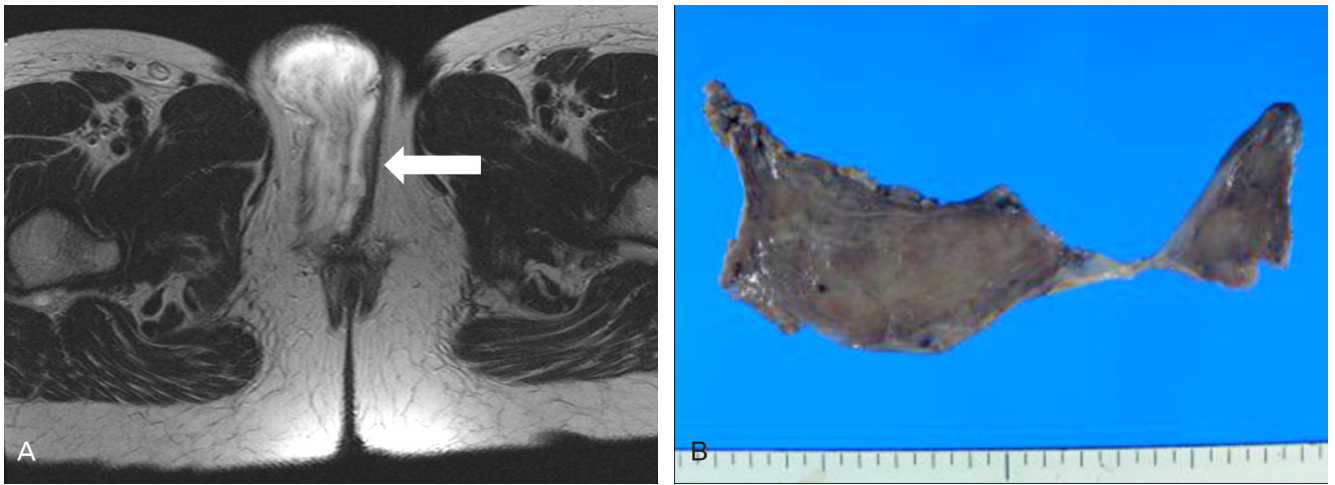
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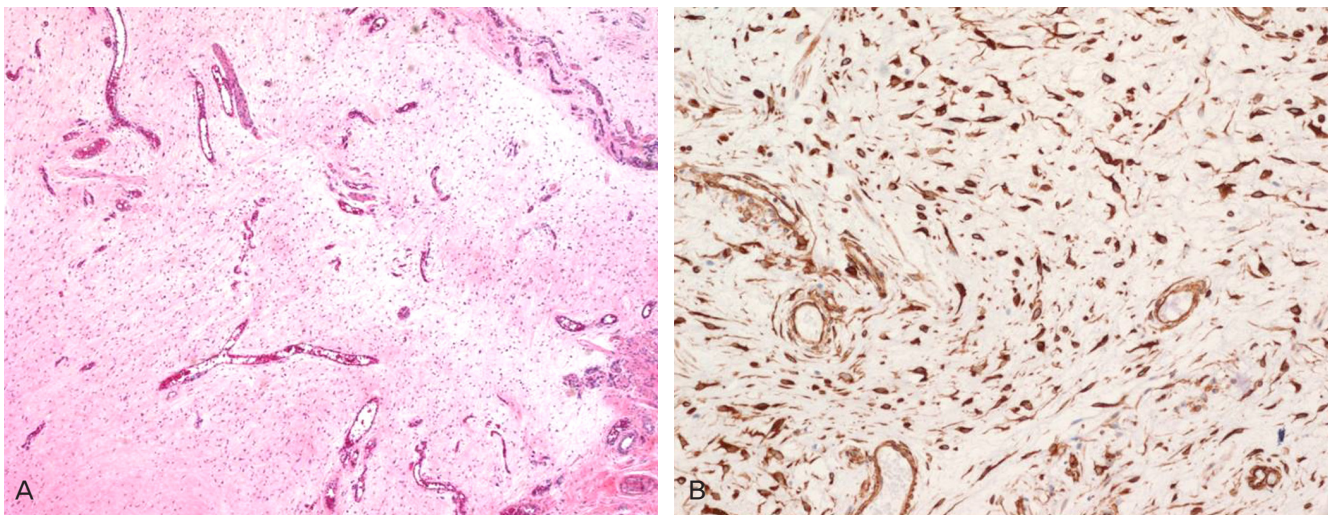
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**Fig. 1.** (A) A well-defined mass-like lesion about 10×7 cm in size is seen in the subcutaneous layer of right labium majora. It shows bright signal intensity on T2WI, and demonstrates delayed enhancement after contrast administration. (B) After surgical excision, the tumor mass was found to be a piece of soft, pink tan tissue, measuring 10×7 cm.



**Fig. 2.** (A) Tumor composed of spindle- and stellate-shaped cells in a myxoid matrix (H&E, ×50). (B) Immunohistochemical staining of the tumor was positive for vimentin (vimentin, ×200).

(Fig. 2B) and negative for S-100. Based on these pathologic features, aggressive angiomyxoma was diagnosed. She was continuously followed-up in our hospital for 7 years after resection, with no clinical or radiologic evidence of recurrence.

## Discussion

AA is a hormonally responsive tumor; it is positive for ER and/or PR and is believed to arise from specialized mesenchymal cells of the pelvic or perineal region. Sometimes suggest that

AA arises from multipotent perivascular progenitor cells because it often displays variable myofibroblastic and fibroblastic features [3]. Microscopically, the tumor consists of spindle and stellate-shaped cells in a myxoid matrix expressing vimentin, desmin, and smooth muscle antigen (SMA) but the cells are negative for S-100 [4]. Our case also had pathologic features of an AA. Immunohistochemical staining of the tumor was positive for ER, PR, and vimentin and negative for S-100.

The differential diagnosis of aggressive angiomyxoma included myxomas, angiomyofibroblastoma, fibroepithelial stromal polyp, superficial angiomyxoma, myxoid neurofi-

broma, myxoid liposarcoma, and myxofibrosarcoma. AA has thick-walled vessels, which are more uncommon than the thin-walled vessels seen in angiomyofibroblastoma. Angiomyofibroblastoma is well circumscribed (this characteristic can also be seen on MRI). Thus, on computed tomography (CT) scans, AAs have a well-defined margin with attenuation less than that of muscles. On MRI, these tumors show high signal intensity on T2-weighted images. The attenuation on CT and high signal intensity on MRI are likely to be related to the loose myxoid matrix and high water content of angiomyxoma [5]. Our patient was received a pelvis MRI. The pelvis MRI showed high signal intensity on T2-weighted images that were compatible with those of AA in the subcutaneous layer of the right labium majora, with no other abnormality.

Surgical excision was the treatment of choice. However, the tumor carries a high risk of local infiltration and recurrence after complete excision, with reports of around 50% to 70% of patients experiencing a relapse after surgical resection [6]. Recurrences generally occur in the first 5 years after primary surgery, and about 70% occur in the first 3 years, but late recurrences up to 14 years have been reported [7]. Five cases of AA of the vulva have been reported in the Korean literature; four cases were local recurrence of AA [8-11] and one case first occurred in the adolescent period. The case was followed up for one year without recurrence [12].

Various methods have been used to treat recurrences, and many options have been successful. Hormonal treatment with tamoxifen, raloxifene or gonadotropin-releasing hormone analogues has been shown to reduce tumor size. Moreover, hormonal therapy may also help to achieve complete excision in large tumors and can be used to treat recurrences [13]. Subsequent tumor resection followed by angiographic embolization may also be helpful because this procedure shrinks the tumor and makes it easier to identify the tumor from surrounding normal tissues [14]. However radiation therapy and chemotherapy are considered less-suitable options due to the low mitotic activity of this tumor.

Recurrences are known to happen late, so all patients must be counseled about the need for long-term follow-up. Unfortunately, there are few reports about the long-term follow-up outcome despite its significance. Our patient has been followed up annually for 7 years without local recurrence.

## Conflict of interest

No potential conflict of interest relevant to this article was reported.

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