Case Report

A case of male perineal aggressive angiomyxoma with expressions of female hormone receptors

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Introduction: Aggressive angiomyxoma is a rare mesenchymal tumor in females of reproductive age that occurs in the pelvis and perineal zone with a high risk of local infiltration and recurrence. Male aggressive angiomyxoma in perineal zone is very rare.
Case presentation: A 63-year old male presented to our hospital with chief complaint of perineal mass. He was diagnosed with perineal lipoma by needle biopsy 3 years before. Computed tomography revealed a 16 cm perineal tumor without lymph node or distal metastasis. As the tumor had gradually increased, we performed tumor resection. The histological diagnosis of tumor using immunohistochemistry was aggressive angiomyxoma with complete resection. Moreover, expressions of both estrogen and progesterone receptors were recognized. No recurrence was seen 9 months after surgery.
Conclusion: Male aggressive angiomyxoma is a very rare tumor, which has expressions of female hormone receptors. Hormonal therapy might be effective for perioperative therapy or recurrence.

Key words: aggressive angiomyxoma, female hormone receptor, perineal tumor.

Keynote message

Male aggressive angiomyxoma is a rare mesenchymal tumor. Female hormonal therapy based on expression of ER and PgR might be effective for preoperative therapy or in case of recurrence.

Introduction

Aggressive angiomyxoma (AAM) is a rare mesenchymal tumor that is usually found in the pelvis and perineum of reproductive female patients. Male AAM is very rare and preoperative diagnosis is difficult. Herein, we report a case of a male AAM in perineal zone that has expression of female hormone receptors.

Case presentation

A 63-year old male presented at our institution with a chief complaint of perineal mass. Physical examination revealed a soft painless, smooth surface perineal mass. Laboratory examination and tumor markers were unremarkable. Three years before, computed tomography (CT) revealed a 7 cm low density mass in the left perineal area (Fig. 1a). At this point, the pathological diagnosis by needle biopsy was lipoma. This tumor grew to 16 cm without enhancement in the perineal area after 3 years (Fig. 1b). No enlarged lymph node and distal metastasis could be observed. Magnetic resonance imaging (MRI) showed a large perineal mass with isointense signal on T1-weighted images and hyperintense signal on T2-weighted images (Fig. 1c 1d). Fluorodeoxyglucose-positron emission tomography (FDG-PET)-CT scan showed a perineal mass with mild accumulation. As the tumor had increased markedly, we performed a resection of it by transperineal approach. The incision of the operation was on the left side of the scrotum, and the length was 10 cm. The tumor was soft and well-

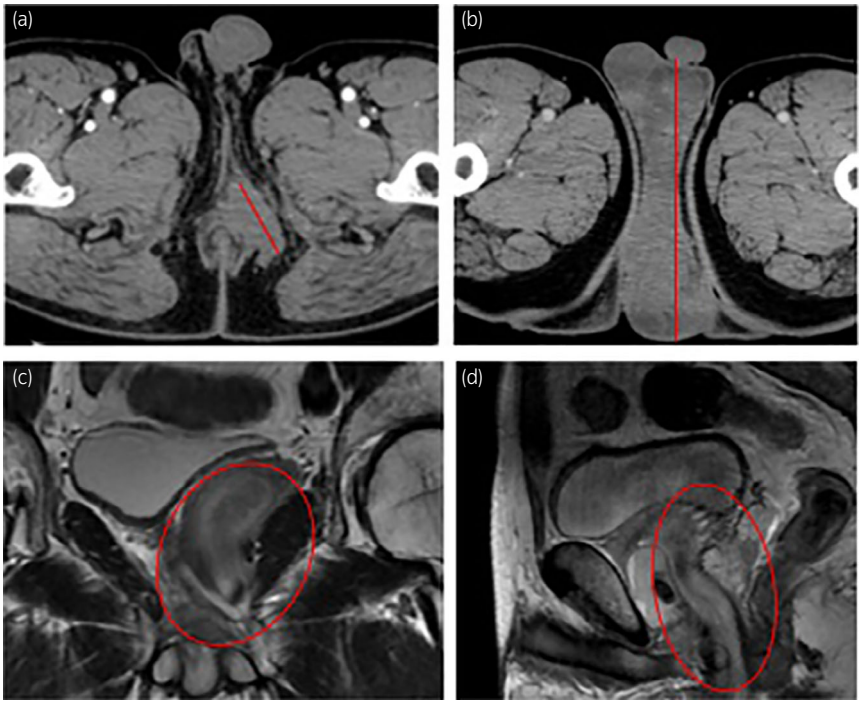

Fig. 1 (a) Enhanced computed tomography (CT) showed a 7 cm mass lesion in perineal zone 3 years before. The red bars indicate the length of the mass. (b) Enhanced CT shows a 16 cm mass lesion without enhancement. (c) Sagittal and (d) Coronal MRI showed a high intensity lesion with swirled appearance on the T2-weighted images. The red circle indicates the "swirling and layering pattern".

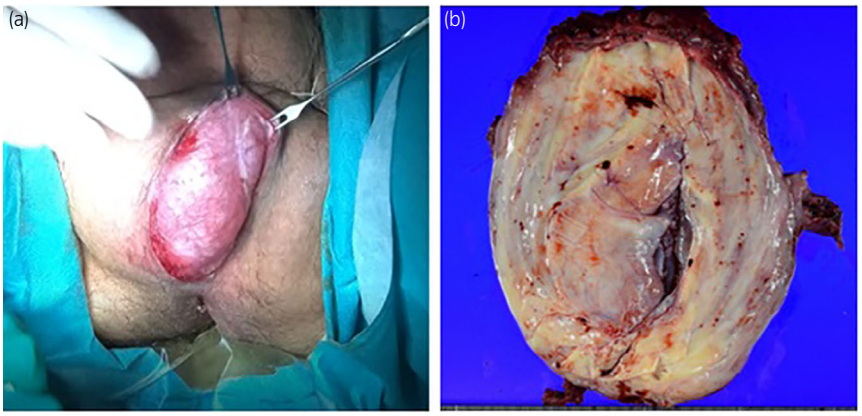

Fig. 2 (a) Intraoperative findings. The tumor was soft and well-circumscribed with capsule formation. (b) Macroscopic figure of the tumor. The cut surface was yellowish-white, and there was included a little mucus component.

circumscribed, having a capsule formation (Fig. 2a). The cut surface was yellowish-white, and there was a little mucus component (Fig. 2b). Microscopic examination showed fibrotic stroma and vascular growth of unequal size throughout the tumor. This area showed spindle-shaped cells and prominent thickening of vascular smooth muscle cells (Fig. 3a,b). The surgical margin was negative. Immunohistochemical studies a showed positive staining of tumor cells for desmin, α-SMA, and CD34, and a negative one for S100 (Fig. 3c–f). These features led to the diagnosis of aggressive angiomyxoma. Currently, 9 months after surgery, the patient is in remission without sign of recurrence.

Discussion

AAM is a rare entity most frequently affecting reproductive age women (male/female: 1/7). Steeper and Rosay have first described it in 1983[1] and only about 350 cases have been reported thus far. This tumor most commonly originates from the vulvar region, pelvis and perineum in females and from the scrotum and inguinal region in males, respectively. In most cases, angiomyxoma is an asymptomatic gradually enlarging mass, its main symptoms being swelling without pain, abdominal distention, and hematuria or vaginal hemorrhage in women.[2] In our case, the only symptom was the perineal mass. There were 57 cases in male AAM described in the literature as far as we investigate. The median age of these patients is 55 years, ranging from 13 to 82 years. AAM is derived from scrotum (38%); inguinal region (16%); perineal region (12%); and other organs such as testicle, ureter, and prostate. Yoshitomo et al. have reported that 13% of AAM in males have originated from the perineal region.[3] Preoperative diagnosis of AAM is difficult; however, it has a characteristic imaging finding that T2-weighted MRI shows well-circumscribed T2 high-signal tumor with central low T2 signal intensity and is called "swirling and layering pattern."[4,5] This might assist in preoperative diagnosis and it was also observed, retrospectively, in our case. Some cases

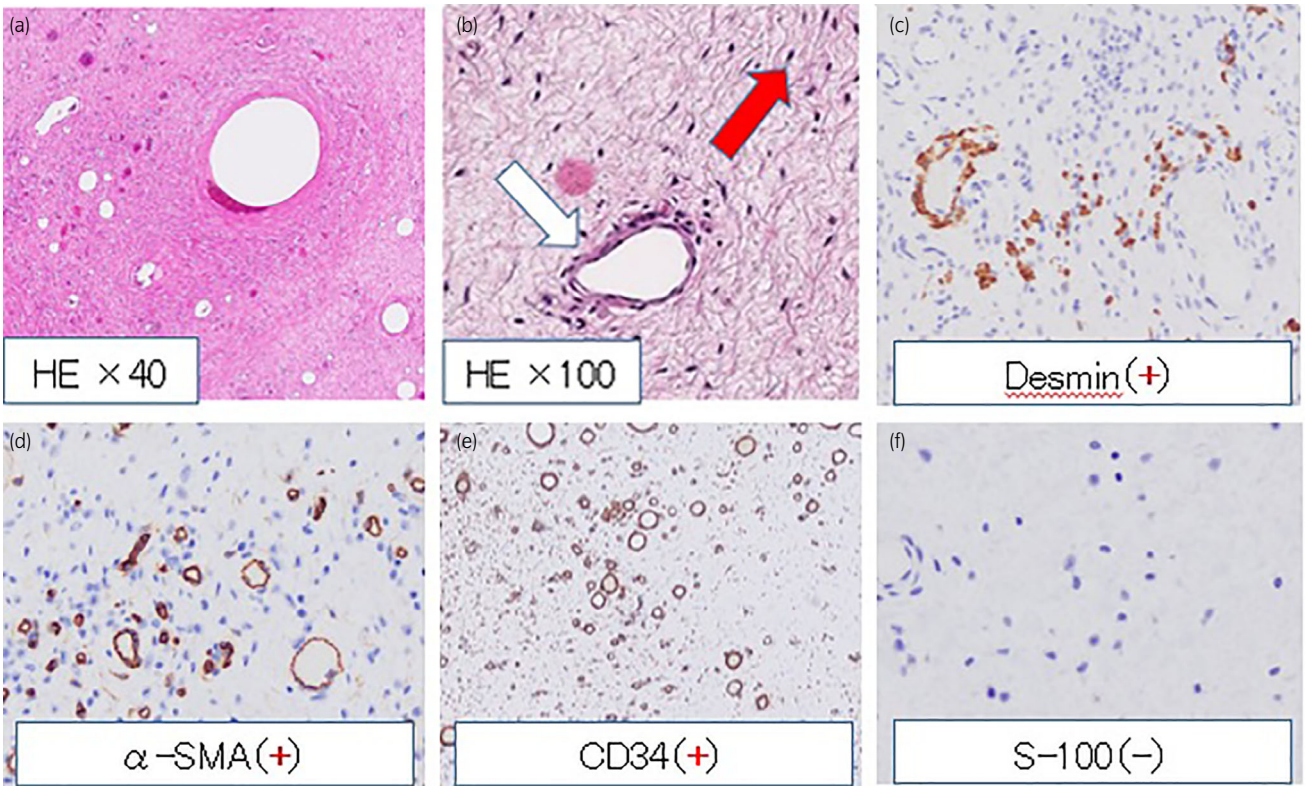

Fig. 3 Histological findings. (a) Hematoxylin–eosin staining showed fibrotic stroma and vascular growth of unequal size throughout the tumor. (b) In strong magnification, the same area showed spindle-shaped cells (red arrow) and prominent thickening of vascular smooth muscle cells (white arrow). Immunohistochemical findings showed that staining of tumor cells for desmin (c), α-SMA (d) and CD34 (e) is positive, while for S100 (f) is negative.

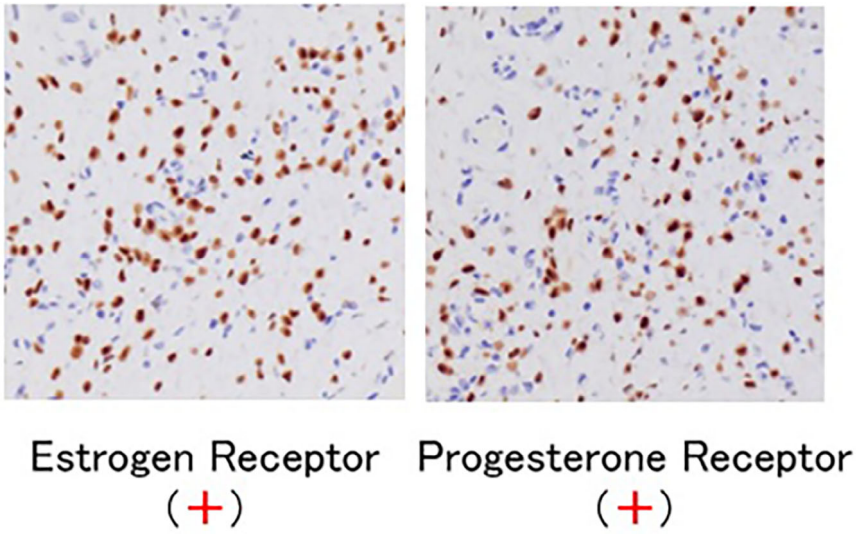

Fig. 4 The expressions of hormone receptor for both estrogen and progesterone were positive.

are diagnosed with AAM by needle biopsy before surgery. The possibility of the correct diagnosis will be increasing by performing biopsy for a size larger than 10 cm of AAM.[2,6] In our case, the needle biopsy performed 3 years before could not deliver the AAM diagnosis probably because the suspect mass had a size of less than 10 cm, and the sampling tissue was of considerably a small amount to confirm diagnosis.

Perineal tumors include epithelial, mesenchymal, and secondary tumors. Mesenchymal tumors also include sarcoma and benign mesenchymal tumors. The benign mesenchymal tumor has mainly three differential diagnoses as superficial angiomyxoma, angiomyofibroblastoma, and aggressive angiomyxoma.[7] Differential diagnosis of these three tumor types is performed histologically by hematoxylin–eosin staining and immunostaining. The typical findings of AAM

include stellate to spindle-shaped tumor cells with fibromyxoid stroma and hyalinized thin-to-thick wall vessels, and no necrosis or cystic changes. Moreover, AAM has as a specific characteristic immunoreactivity for desmin and αSMA, and CD34 is positive and S-100 is negative.[8] Our case was consistent with these pathological characteristics including immunohistochemistry.

There are several treatments for AAM such as surgery, hormonal therapy, radiation, and angiographic embolization, but surgical excision is the only curative treatment.[8] Even if complete surgery is performed, local relapse occurs in 50% of the cases.[9] It was suggested that AAM is associated with female hormones because the expressions of estrogen receptor (ER) and progesterone receptor (PgR) were observed in female cases.[10] Therefore, the hormonal therapy has been administered among perioperative therapy in some cases.[11] Bensalah et al reported that female patients with AAM of the pelvis received preoperative neoadjuvant hormone therapy using GnRHa. After decreasing the diameter of the tumor, they underwent surgery with complete resection.[12] In cases where the tumor had progressed from the perineum to the pelvis and complete resection or organ preservation was difficult, preoperative hormone therapy with GnRHa was considered a suitable treatment for tumor shrinkage. Therefore, the expression of ER and PgR in the resected tumor was examined and it was found that both of these hormone receptors were positive (Fig. 4). These results suggested that preoperative hormone therapy for tumor shrinkage might have been effective in this case. Moreover, hormonal therapy was considered effective when recurrence was subsequently observed.

Conclusion

We report on a case of male perineal aggressive angiomyxoma. Hormonal therapy based on the expression of ER and PgR might be effective for perioperative therapy or recurrence.

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Author contributions

Mizuki Hisano: Conceptualization; data curation; investigation; methodology; project administration; writing – original draft. Tomohiko Matsuura: Project administration. Renpei Kato: Project administration. Shigekatsu Maekawa: Project administration. Yoichiro Kato: Project administration. Mitsugu Kanehira: Project administration. Masamichi Suzuki: Investigation; project administration. Manabu Nakayama: Investigation; project administration. Ryo Takata: Supervision; validation. Wataru Obara: Supervision; writing – review and editing.

Conflict of interest

The authors declare no conflict of interest.

Approval of the research protocol by an institutional reviewer board

Not applicable.

Informed consent

Not applicable.

Registry and the registration no. of the study/trial

Not applicable.

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