Recurrent transperineal aggressive angiomyxoma: a case report

Xiaoli Li
Zhejiang University School of Medicine Second Affiliated Hospital
Chunxia Zhou
Zhejiang University School of Medicine Second Affiliated Hospital
Jun Lin (linjun@zju.edu.cn)
Women's Hospital, School of Medicine, Zhejiang University

Case report

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Recurrent transperineal aggressive angiomyxoma: a case report

Xiaoli Li[1], Chunxia Zhou[1], Jun Lin[2]

1 Department of Gynecology, The Second Affiliated Hospital of Zhejiang University School of Medicine, Hangzhou, Zhejiang, People's Republic of China.

2 Women's Hospital, School of Medicine, Zhejiang University, Hangzhou, Zhejiang, People's Republic of China. Electronic address: linjun@zju.edu.cn.

Abstract

Background: aggressive angiomyxoma is a rare disease that may cause misdiagnosed in the clinical work and the characteristic of this disease is low potential malignancy. This case shows a recurrent case of aggressive angiomyxoma 7 years after the surgery which was mistakenly diagnosed at that time.

Case presentation: the patient presented a large mass in the right labium majus without any pain. This mass was recurrent 7 years after she received a surgery about the mass occurred at the same place which was diagnosed as vulvar angiomyofibroblastoma. She took the ultrasound scan and MRI scan and underwent the surgery again. This mass was excision completely. At this time, pathologist checked the HE stained slides and immunohistochemistry staining slides, then come the the conclusion that this mass was aggressive angiomyxoma. We rechecked the pathological slides 7 years ago and found out it was misdiagnosed at that time.

Conclusions: Aggressive angiomyxoma is a rare tumor. This case presents a rare disease that may be misdiagnosed as other benign vulvar disease even after the surgery which get pathologic evidence. So we need to know more about this disease.

Background

Aggressive angiomyxoma is a rare stromal disease that most frequently occurs in the premenopausal women, especially between second and forth decade, which is usually arise from mesenchymal tissue and presents low potential malignancy. This tumor is often misdiagnosed as Bartholin’ s cyst, angiomyofibroblastoma or hernia because of its non-specific clinical and radiological aspects[1]. In this case, we’ ll describe a case about a female patient who was mistakenly diagnosed as angiomyofibroblastoma after initial surgery and recurrent 7 years later with the correct diagnosis at the same time.

Case presentation

A 38-year-old female came to the outpatient department of our hospital, presented with a progressive enlargement asymptomatic mass in the right labium majus with a duration of 1 year growth [Figure 1]. The patient complained that the nontender mass was recurrent, with a history of mass excision she underwent at the same site as the primary lesion 7 years ago. At that time, pathological diagnosis of the mass was vulvar angiomyofibroblastoma. The patient also had a medical history about her right breast cancer which received 4 times neoadjuvant EC chemotherapy regimens and 4 times docetaxel combined with Herceptin chemotherapy regimens. After that, she underwent right breast-conserving surgery on 2015 and continued 11 times of Herceptin target therapy every 3 weeks for the next half a year. Because of the breast cancer, she took tamoxifen pills till now.

At first, the patient took a superficial ultrasound scan about her mass. The result of ultrasound scan suggested a solid mass inside the soft tissue measuring 7.7*2.4cm. The mass was well-circumscribed with septation inside and the lower edge was attached to the anal canal. Colored Doppler showed there were rich flow signals on the circumference and inside the lesion. Then MRI scan was performed to evaluate the mass. On Magnetic Resonance Imaging, a 44*58*83mm irregular shape lesion was demonstrated on the right side of vulva, anterior to the right of the anal canal with a well-defined margin (Figure.1). T1-weighted imaging showed hypointense while T2-weighted images showed high signal intensity compared to muscle at the same time. There was mild enhancement after gadolinium injection. No obvious enlarged lymph node was observed in pelvic. The MRI diagnose was right perineum mass, intended to consider as benign tumor like angiomyolipoma or

hemangioma。

On palpation, the physical examination showed a mass on the right labia majora which was about 8*4cm in size, with obvious pigmentation, soft texture and no tenderness (Figure.2).

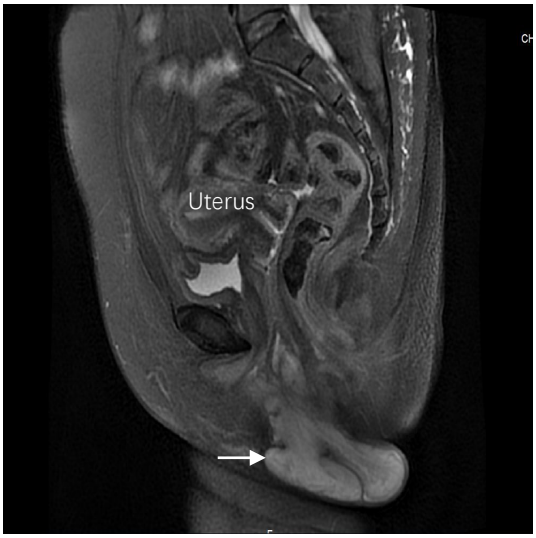


Figure.1: sagittal section of MRI showing the solid mass on the right side of vulva

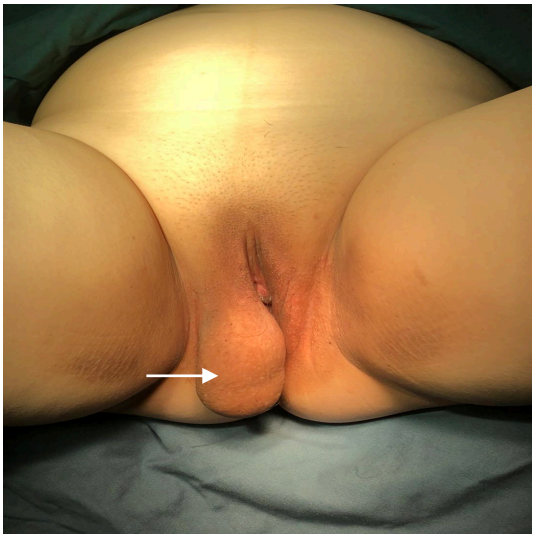
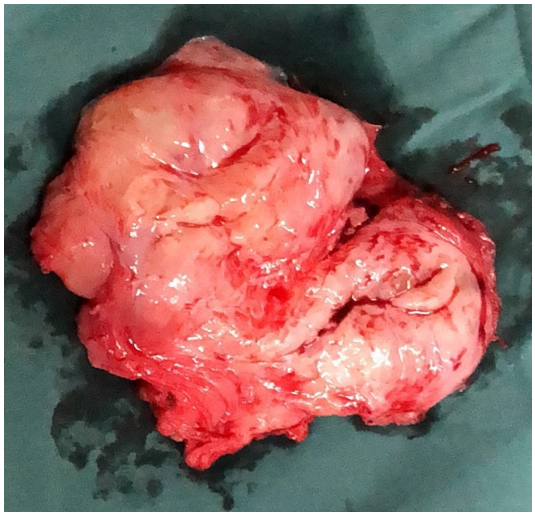


Figure.2: the arrow showing the mass on the right labia majora

Hence, a surgical operation was performed, which in the proceed, we could find the upper edge of the mass is the pubic symphysis, the right edge is the pubic arch, the left edge is the right vaginal wall, partly protrudes toward the vagina, and the lower edge is the level of the sacral ligament. The mass suffered complete excision and on macroscopic view, the sectioned surfaces were soft accompanied by edema and mucoid degeneration change.

The gross appearance of the tumor was large, which was about 8 cm in maximum diameter. Microscopic observation revealed that the tumor volume was large, unclear boarder and growed deeper and infiltrated into surrounding fat tissue. There was even and consistent mucus inside the tumor with low cell density and mild morphology. The cells were little atypical and no mitosis phase. A lot of vessels could be found in the background of mucous which the vessel walls varies in diameter and thick-walled vessels and hyaline degenerated vessels could be seen. The immunohistochemistry staining results of the mass were positive for smooth muscle actin, desmin, vimentin, CD31 and CD34, meanwhile negative for S-100.



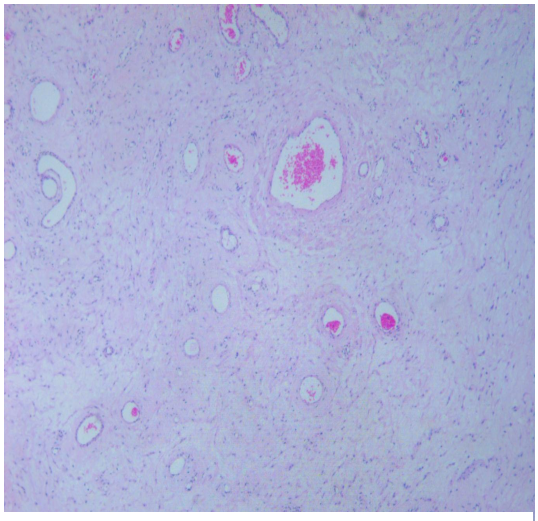


Figure.3: Cut surface of myxoid mass showed edema and mucinous degeneration

Figure.4: Vascular with uneven wall thickness proliferation can be seen in the mucoid stroma , and the vascular lumens are irregular.(H&E X400).

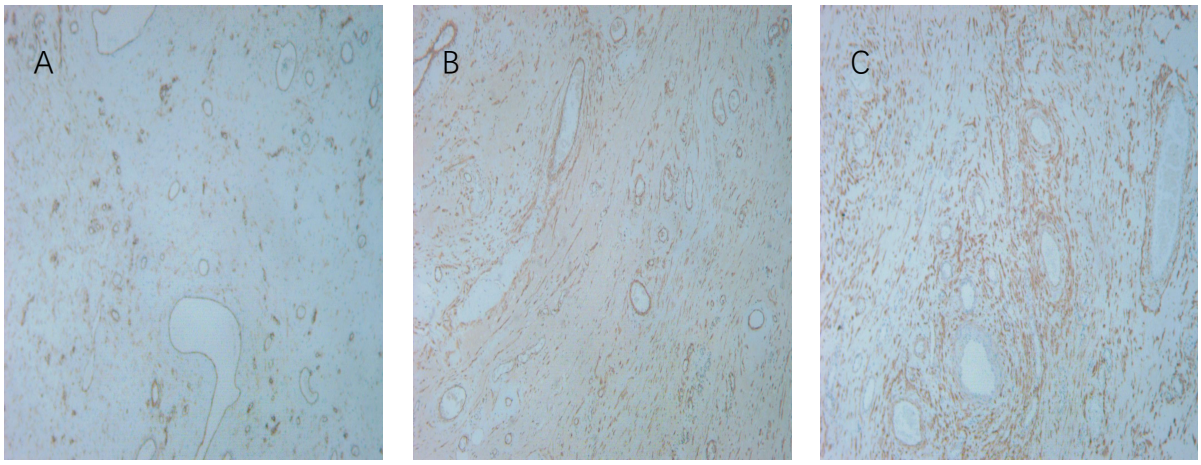


Figure.5: immunohistochemical staining for tumor : A, positive for CD34, stain highlights blood vessels, B, positive for desmin, C, cytoplasm positive for SMA, B and C displayed the component of smooth muscle in the neoplasm

Discussion and conclusions

Aggressive angiomoyxma is uncommon benign mesenchymal tumor with a predilection for perineum, which characteristics are a high risk of relapse after resection[2] and usually have a large size. It was first reported in 1983 by Steeper and Rosai[3]. It looks morphologically

similar as angiomyofibroblastoma which both grow at a very slow rate that hence patients will be asymptomatic for a long time before they are conscious of the mass[4].

Usually, histopathologic examination of aggressive angiomyxoma revealed numerous thick-walled blood vessels in a hypocellular myxoid stroma, the irregular boundary and abundant mucous matrix with hyaline change in the vessels' wall. Immunohistochemically, the stromal cells showed expression of desmin, smooth muscle actin, vimentin and CD34 in which atypical vessels cound be found in. In pathology it needs to identify with angiomyofibroblastoma which is rare vascular soft tissue neoplasma in vulva. Angiomyofibroblastoma is morphyologically familiar to aggressive angiomyxoma as they share many features such as painless and slow-growing mass. But histopathologically it displays superficial vascular mucinous, clear boundary, more smooth muscle components in stromal matrix and presents more pathological thin-walled blood vessels[5]. It' s usually negative for desmin, smooth muscle actin, vimentin and CD34 as the same time.

Considering of its high local recurrence and characteristic of aggressive, a complete surgical excision is very important[6], which patients will get benefit from it. Tumor-free margin surgical excision is considered as a usual way to get the treatment. But unfortunately, even patients get the complete resection of the mass, it still has the possibility to recurrent[7] and it usually occur at the same site as the primary one[8]. So a long-term follow-up is very necessary.

As to this patient, she was misdiagnosed 7 years ago after her first surgery. The mass she got excision was diagnosed as angiomyofibroblastoma as we reviewed the pathological slides this time in which revealed thick wall vessels and vascular proliferation can be seen in the mucoid stroma, which showed typically aggressive angiomyxoma. But at that time, pathologist was lack of experience and couldn' t tell the difference between the two disease. It was until 7 years after the patient got the correct diagnose.

The patient has been under follow-up for 2 years now, and remains asymptomatic and with no radiological evidence of recurrence.

Aggressive angiomyxoma is a rare tumor and surgical excision is the first option to cure the disease[9]. Pathologic is the gold standard for diagnose. This case presents a rare disease that may be misdiagnosed as other benign vulvar disease even after the surgery which get pathologic evidence. Hence experienced pathology doctors are very important. But we can still get some clues by review medical history and investigate the imaging features.

Ethics approval and consent to participate

The present study was approved the Institutional Review Board of The Second Affiliated Hospital of Zhejiang University School of Medicine.

Consent for publication

Consent was obtained from the patient

Availability of data and material

The datasets supporting the conclusions of this article are included within the article and its Additional files.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

XL wrote the manuscript. CZ prepared all the figures. All authors read and approved the final manuscript.

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