

Huge Pedunculated Angiomyofibrosarcoma Arising from The Vulva: A Case Report with MR Findings and Review of the Literature

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1. Abstract

Angiomyofibrosarcoma (AMF) is a benign mesenchymal tumor of the genital tract, which is more common in young and middle-aged women. It was first reported by Fletcher in 1992[1]. It mostly occurs in vulva, but there are also a few cases in vagina, cervix, fallopian tube, broad ligament, scrotum, urethra and so on. Here, the case of a 32-year-old female patient, who presented with huge pedunculated AMF arising from vulva, is reported.

2. Introduction

Although vulva appears to be the most frequent sites of origin for the AMF. But AMF usually appears as a small subcutaneous nodule, and the formation of a large pedunculated mass is rare. We present a case of AMF that arose from the vulva, with an emphasis of MR imaging features of the disease.

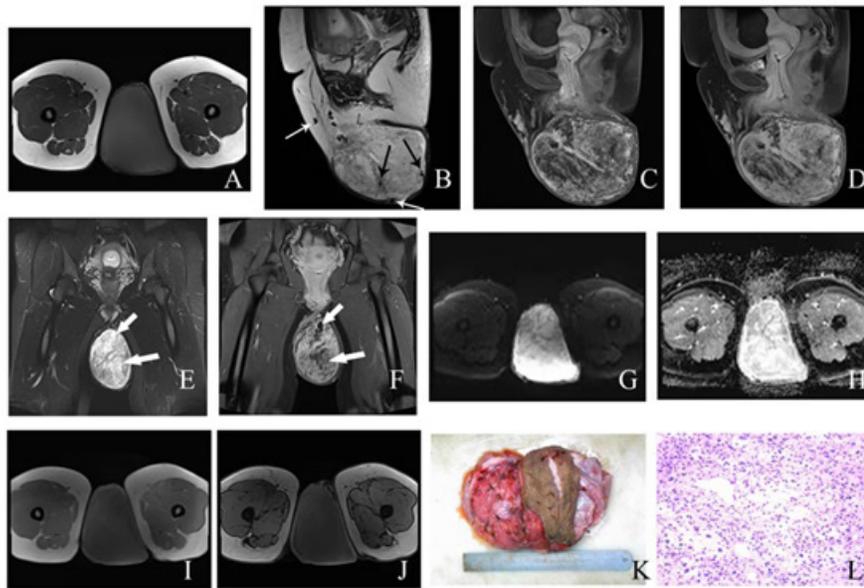
3. Case Presentation

A 31-year-old woman presented with a vulva mass that was detected three years ago. It was clinically diagnosed as vestibular gland cyst at the local hospital. Later, the tumor gradually increased, and the patient began to feel pain, which could be relieved in one minute. The tumor formed a large pedunculated mass, measuring 16 cm in maximal dimension, which hung down from the left labium majus.

The mass was demonstrated as a well-defined, oval shaped mass in the left vulva that measured 15.6×12.6×12.2cm. It showed a homogeneously low intensity on a T1-weighted image (Figure A) and heterogeneously high intensity on a T2-weighted image (Figure B). Some flow void of the vessels were seen in tumor body and in the subcutaneous tissue of the vulva. Diffusion Weighted Imaging (DWI) (Figure G) and Apparent Diffusion Coefficients(ADC) (Figure H) both show high signal. The opposed-phase image (Figure J) did not decrease significantly compared with the in-phase image (Figure I). In contrast enhanced dynamic MR images (Figure C D), they demonstrated a large, well-circumscribed subcutaneous tumor showing a fast and persistent uneven enhancement.

At surgery, a tumor of about 20×10×8cm in size was found in the left labia major, and three large blood vessels with a diameter of about 1cm were found on the surface. The skin and fascia on the surface of the tumor were cut in a fusiform shape about 5cm near the base of the tumor, and the tumor was completely removed from the base.

With immune stains, the tumor cells were positive for vimentin, desmin, CD31 and CD34, the Ki-67 proliferation index was less than 5%. The tumor was negative for S-100, CK, MDM2, SMA, and melanoma. These distinctive pathological and immunohistochemistry features conformed the diagnosis of AMF.



On T1-weighted image (Figure A), the signal intensity of the mass was similar to that of skeletal muscle. The T2-weighted sagittal image (Figure B) showed a huge mass in the left vulva. Some flow void of the vessels was seen in tumor body (black arrow) and in the subcutaneous tissue of the vulva (white arrow) on T2-weighted image. Sagittal T2-fat-saturated MRI images with contrast enhancement (Figure C, D) demonstrated showing an intense and delayed uneven enhancement. Coronal fat-suppressed T2-weighted MR image (Figure E) showed high signal intensity (arrow) on it and the enhancement on fat-suppressed T2-weighted image (Figure F) displayed hypointensity (arrow). DWI (Figure G) and ADC (Figure H) both showed high signal. The opposed-phase image (Figure J) did not decrease significantly compared with the in-phase image (Figure I). The gross specimen (Figure K) is grayish white and grayish red with smooth surface and capsule. There is also a piece of skin and subcutaneous tissue on the lateral side of the left labia major. A photomicrograph of the tumor tissue (Figure L) showed spindle shaped myofibroblasts and abundant thin-walled vessels.

4. Discussion

AMF is characterized by superficial soft tissue mass, usually painless and clear margin, which is often misdiagnosed as vestibular gland cyst [2], lipoma or invasive angiomyxoma. Luis et al [3] reported some examples of AMF contain mature adipocytes as an integral component of the tumor, mature adipocytes accounted for about 50% in 1 case, and even more than 90% in the other 2 cases. Similar to Sims's preoperative diagnosis [4], our case was also misdiagnosed as invasive angiomyxoma. On T2 weighted images, vortex like high signal and low signal fibrovascular tissue were seen in invasive angiomyxoma, with obvious delay and layered enhancement on enhanced scan [5]. Ill-defined margins with surrounding tissues infiltration was often found in Aggressive angiomyxoma.

Lim et al [6] described MR findings of a case of AMF originating from the posterior space of bladder. On T1-weighted images, the signal intensity of the mass was similar to that of skeletal muscle. After injection, the mass showed strong and homogeneous enhancement on T1-weighted images. On contrast enhanced dynamic MR images, the mass showed fast and persistent enhancement. They thought this marked delayed contrast enhancement may be due to abundant fibrous tissue in the tumor. In our case, a strip of low signal was seen in the mass at T2-weighted images, and enhanced on a contrast-enhanced MR images. This phenomenon can prove the existence of fibrous components in the mass. Shintaku et

al [7] and Ito-Miyazaki et al [8] described a case of AMF from the right labium majus respectively. They both showed homogeneously low intensity on T1-weighted images and heterogeneously high intensity on T2-weighted images. The lesions were irregularly enhanced by the administration of a contrast medium. Geng J et al [9] supported that the high signal intensity seen on T2-weighted images and the enhancement on T1-weighted images after administration of gadolinium reflect the high myxomatous content of the tumor and the vascularity of the tumor respectively when correlated with the pathological specimen.

5. Conclusion

MRI is a just a clinical tool and the MR features for AMF are non-specific. However, a well-defined and slowly growing mass in the reproductive system of young and middle-aged women presented with long T1 and long/short T2 signals and obvious uneven continuous enhancement, with many blood vessels in and around the tumor, angiomyofibroblastoma should be considered.

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