Case Report

Giant Fibroepithelial Stromal Polyp of the Vulva: Diffusion-Weighted and Conventional Magnetic Resonance Imaging Features and Pathologic Correlation

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A B S T R A C T

Background: Fibroepithelial stromal polyp (FESP) is a rare benign mass, usually presenting at the vagina. Herein we report, to our knowledge, the first case of contrast-enhanced magnetic resonance imaging (MRI) with diffusion-weighted images of a giant vulvar FESP, and compare the MRI features with the histopathologic results.
Case: A 14-year-old girl presented with a huge mass as large as 20 cm that originated from the labium majora. Preoperative MRI showed a polypoid mass consisting of a central stalk and surrounding stroma. Different signal intensities on MRI were correlated with various histopathologic features. The mass was cured by complete excision without remnant lesion.
Summary and Conclusion: Contrast-enhanced MRI with diffusion-weighted images can help us differentiate FESP from other vulvovaginal stromal tumors with a complete evaluation of the external and internal structures and the depth of invasion.
Key Words: Vulvar diseases, Fibroepithelial stromal polyp, Adolescent, Magnetic resonance imaging, Diffusion magnetic resonance imaging

Introduction

Fibroepithelial stromal polyp (FESP), also known as mesodermal stromal polyp, cellular pseudosarcomatous fibroepithelial polyp, or pseudosarcoma botryoides, is a rare benign polypoid subepithelial mesenchymal proliferation that occurs at the vagina, vulva, or cervix.[1] The clinical features are unusual, often overlapping with other genital stromal tumors or malignant neoplasms, and the various histologic appearances and the rarity of this disease also make its diagnosis challenging.[2–4] Despite diagnostic difficulties, the radiologic features of FESP are rarely published, and there have only been 2 case reports, which used ultrasonography (US), non-contrast-enhanced (CE) computed tomography (CT) findings, and magnetic resonance (MR) images of FESP.[5,6] Herein we report on a FESP as large as 20 cm in a 14-year-old girl and introduce CE MR imaging (MRI) with diffusion-weighted images for the first time in such a case. We also correlate the imaging features with the histopathologic findings and discuss the role of MRI in differentiating FESP from other vulvovaginal stromal tumors.

Case

A 14-year-old girl presented with a huge, exophytic, polypoid mass arising from her right labium majora. It had first been noticed 2 years earlier as a small nodule; however, it had grown rapidly during the past several months. Although the mass was asymptomatic, sudden mild pain and discomfort had developed 3 days earlier. Physical examination revealed a nontender, soft, polypoid mass attached to the right labium majora with a stalk. The surface of the mass showed numerous shallow skin folds without ulcer, bleeding, or discharge. She was nulliparous and took no hormonal drugs. There was no relevant family history. For further evaluation, she underwent CE-MR examination (Skyra 3T; Siemens).

The MR image showed a 20-cm, pedunculated, polypoid mass arising from the right vulva. It was composed of a stalk attached to the vulva and a stroma that occupied most of the mass (Fig. 1). Specifically, the central stalk consisted of 3 areas of different signal intensities: (1) hyperintense area on T1-weighted images with a signal drop on fat saturation technique (Fig. 2); (2) abundant tortuous vessels spreading out from the attachment site; and (3) multiple, small locules that were hyperintense on T2-weighted images, hypointense on T1-weighted images, and not enhanced after contrast injection. The stroma of the polypoid mass consisted of 2 areas of different signal intensities: (1) hypointense area on T2-weighted images, homogeneously enhanced; and (2) hyperintense area on T2-weighted images, less enhanced (Fig. 3). Diffusion-weighted images showed no foci of diffusion restriction. The subcutaneous fat at the attachment site was intact without invasion by the mass. Pelvic organs such as the uterus, ovaries, and urinary bladder were normal. There were no enlarged lymph nodes in the pelvis. Because these MRI findings suggested no foci of malignancy in the mass, no invasion of adjacent

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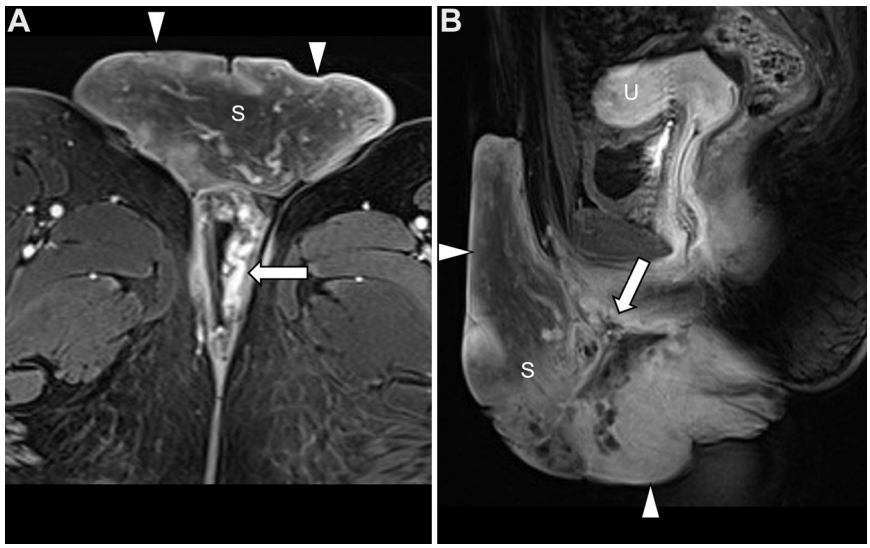

Fig. 1. (A) Axial and (B) sagittal contrast-enhanced T1-weighted images (gradient-echo, fat saturation, TR/TE 3/1) showing the central fibrovascular core (arrows), polypoid stromal proliferation (S), and covering surface epithelium (arrowhead). Uterus (U) and cervix are normal. TR, repetition time; TE, echo time.

subcutaneous fat, and no pelvic lymphadenopathy, the surgeon decided to remove the mass by simple excision.

During the procedure, the patient was placed in the lithotomy position, and surgeons exposed the stalk, approximately 8 cm in size, at the right labia majora and surgically excised it. The excised mass was measured at 20 × 12.5 × 3.5 cm, weighed 509 g, and the cut surface was whitish in appearance (Fig. 4). On histopathologic examination, the mass was found to be composed of sparsely distributed stellate stromal cells that showed various cellularities. The different T2 signal intensities of the stroma were correlated with these various cellularities on histopathologic analysis; areas with scanty cellularity showed T2 hyperintensity with scarce enhancement on MRI. In contrast, areas composed of more stromal cells showed T2 hypointensity with homogeneous enhancement (Fig. 3). Adipose cells

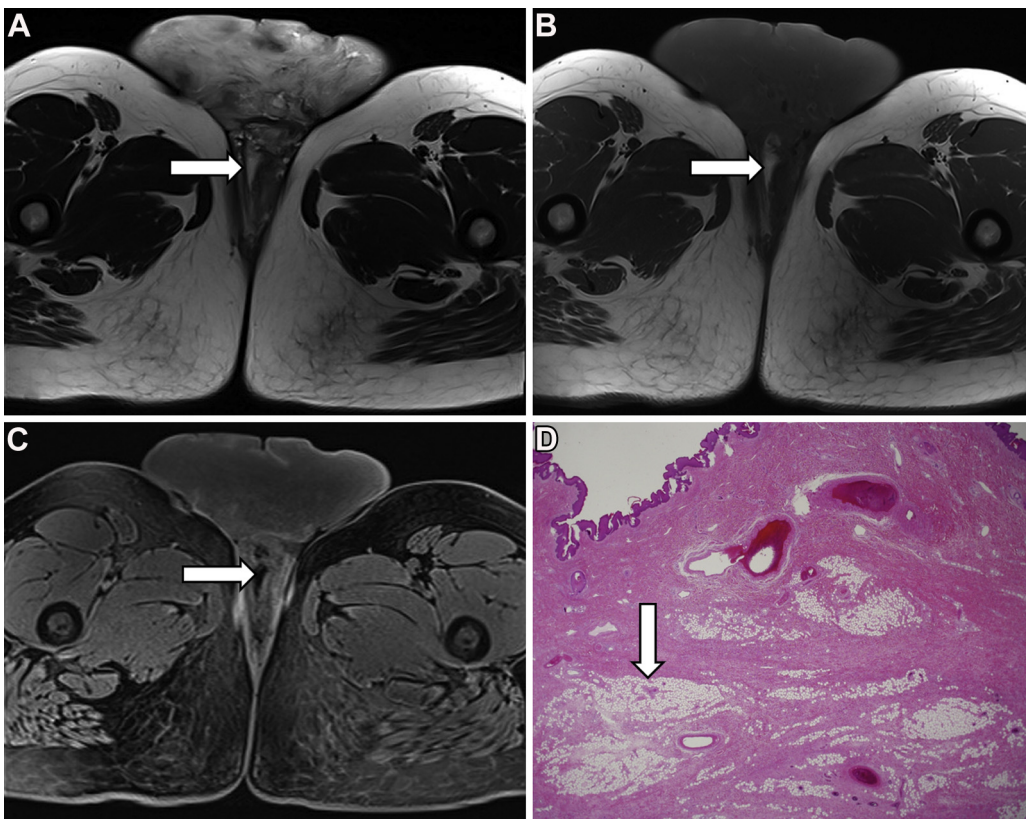

Fig. 2. Fat-containing tissue on axial (A) T2-weighted (fast spin-echo, TR/TE 5140/84), (B) T1-weighted (fast spin-echo, TR/TE 500/11), and (C) T1-weighted images with fat saturation (gradient-echo, fat-saturation, TR/TE 3/1) confirmed as adipose cells on (D) histopathologic specimen (hematoxylin and eosin ×40). TR, repetition time; TE, echo time.

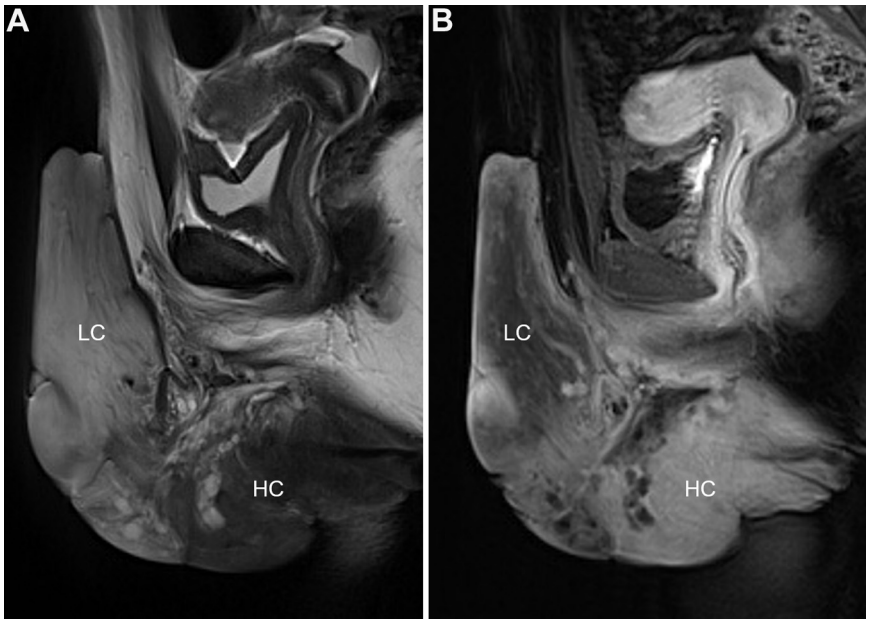

Fig. 3. Different signal intensities of stroma on sagittal (A) T2-weighted (fast spin-echo, TR/TE 5140/84) and (B) contrast-enhanced T1-weighted images (gradient-echo, fat-saturation, TR/TE 3/1) were correlated with a low cellular area (LC) and a relatively high cellular area (HC) on histopathologic examination, respectively.

aggregated close to the stalk were correlated with T1 hyperintensity on the MR image that was suppressed by the fat saturation technique. The results of immunohistochemistry staining were positive for estrogen receptor and desmin, but negative for CD34, smooth muscle actin, and myogenic differentiation 1(Rhabdomyosarcoma marker). Ki-67 showed low proliferative activity. There was no area of myxomatous stroma, densely aggregated spindle cells, or cellular atypia.

After excision, the patient has been stable without any complication during 1 year of follow-up.

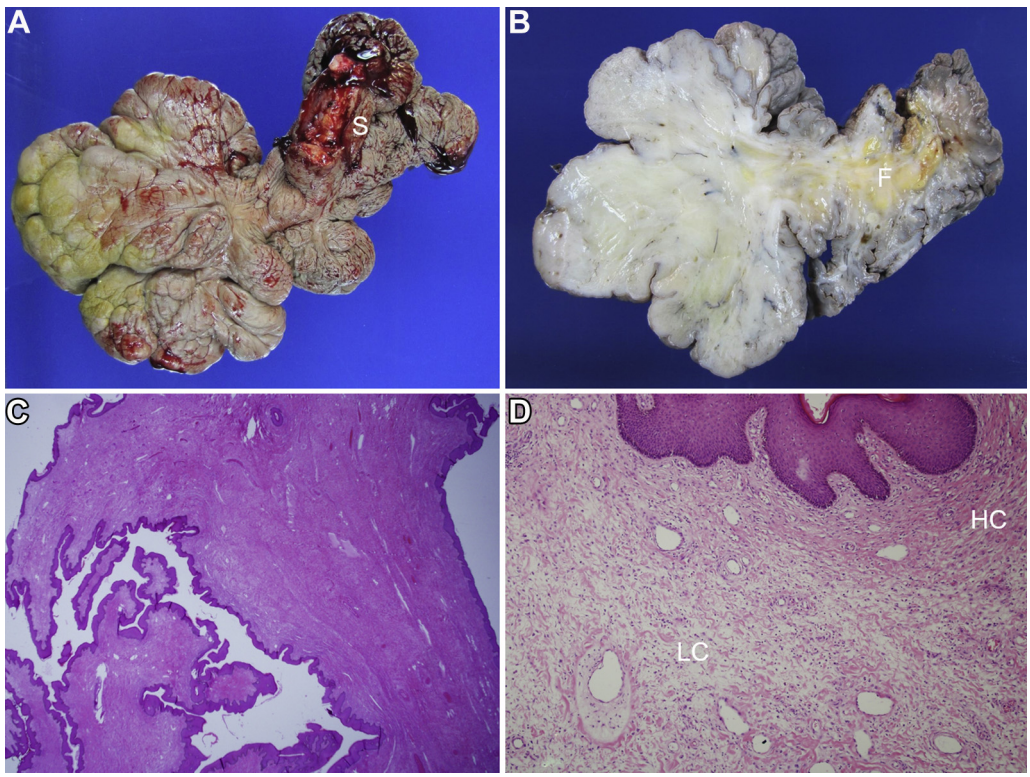

Fig. 4. (A) Gross photograph showing a huge polypoid mass with a central stalk (S). (B) Cut surface photograph showing whitish appearance with yellow fat (F) in the stalk. (C) Histopathologic specimen revealing fibrous stroma with various cellularities and covering surface squamous epithelium (hematoxylin and eosin ×20). (D) Stellate cells formed the stroma with various cellularities (hematoxylin and eosin ×100). Multinucleated cells are typically located under the hyperplastic squamous epithelium. HC, high cellularity; LC, low cellularity.

Summary and Conclusion

FESP is a rare benign soft tissue tumor that occurs in the vulvovaginal region. It likely originates from subepithelial stromal cells or subepithelial mesenchyma of the vulvovaginal area from the endocervix to the vulva. It occurs most frequently in the vagina, followed by the vulva, and the cervix, and rarely occurs in the extragenital area.[1,3] The common manifestation is 1 or more painless polyps. The pathogenesis of FESP is not yet well understood. However, a strong relationship with hormonal stimulation is suggested on the basis of the points of evidence that: (1) it frequently occurs during pregnancy and regresses after delivery; (2) it is associated with hormone replacement therapy or tamoxifen treatment; and (3) the stromal cells of FESP are reactive to estrogen receptor, progesterone receptor, and desmin.[1,4,7,8] The rapid growth seen in our case is thought to result from hormones in adolescents. However, sometimes symptoms such as bleeding, discharge, and discomfort can also be associated with FESP, depending on the size of the mass, which is usually smaller than 5 cm. However, it might be as large as 20 cm and might sometimes undergo torsion and develop stromal edema.[2–4]

Although most genital stromal tumors are small, usually less than 5 cm, several giant masses in the vulvovaginal region in the literature have been found to be FESP or aggressive angiomyxoma.[2,3,5,6] FESP is benign, with no reported destructive local recurrence or metastasis, unless the resection margin is positive.[9] Aggressive angiomyxoma is the only genital stromal tumor with a 30%-40% recurrence rate, although this rate has recently been reduced to less than 10% with the use of an aggressive surgical procedure.[1] Thus, it is important to distinguish aggressive angiomyxoma from FESP and some researchers have reported distinguishing features to help differentiate them.

Chan et al have mentioned that aggressive angiomyxoma show more infiltrative borders and a deeper location, whereas FESP has a circumscribed margin with a superficial subepithelial location on histopathologic examination.[2] Our results suggest that MRI is useful for determining the depth of invasion. Kato et al have emphasized that the detection of fatty tissue on MRI is a specific finding that suggests FESP.[6] We also observed adipose cells aggregating close to the stalk, suggesting that MRI is helpful for detecting fat, especially when using the fat saturation technique.

It is also important to exclude botryoid embryonal rhabdomyosarcoma when FESP exhibits pseudosarcomatous morphology. The differential point is that botryoid embryonal rhabdomyosarcoma is typically diagnosed in prepubertal girls and is reactive to specific markers of skeletal muscle differentiation such as myogenin and myogenic differentiation 1(Rhabdomyosarcoma marker) staining, whereas those are negative in FESP, as in our case.[1,3] The differential imaging features between these 2 lesions are yet to be studied.

The radiologic features of vulvar FESP have rarely been reported. The review of the literature revealed 2 case reports showing US and non-CE CT and MRI of FESP.[5,6] Our study is the first report, to our knowledge, to present CE-MRI and diffusion-weighted images as well. Bozgeyik et al have postulated that US is a more suitable modality for the initial examination, because it is more convenient to use and less expensive, but shows the blood supply and the extent of the mass.[5] However, we used MRI as the first imaging study with our patient, because the mass was as large as 20 cm upon gross inspection, and we thought MR or CT imaging might better include and evaluate the entire mass. In addition, MRI could also provide information about all pelvic structures, including the uterus, ovaries, and lymph nodes without radiation exposure, an important consideration for children and adolescents. As a result, in our case, MRI proved to be a useful modality for evaluating the mass and the pelvic structures without radiation exposure, to exclude malignancy on the basis of diffusion-weighted imaging and help establish the surgical plans.

Upon histopathologic examination, FESP is typically composed of 3 components: a central fibrovascular core, pedunculated or polypoid stromal proliferation, and a covering surface squamous epithelium.[1,2] FESP can exhibit a wide range of histopathologic morphologies, including features typical of sarcomas, such as hypercellularity, bizarre nuclear features, numerous mitotic figures of more than 10 mitoses per 10 high-power field, and atypical stromal cells.[9] When these features are present, the term cellular pseudosarcomatous fibroepithelial polyp or pseudosarcoma botryoides can be used. In our case, MRI clearly demarcated these 3 histopathologic components and the worrisome histologic features were not detected, although the mass was as large as 20 cm. In addition, different T2 signal intensities and differential enhancements on CE-MRI of the stroma were correlated with different cellularities of the histopathologic features.

That there was no area of diffusion restriction in our patient's diffusion-weighted images might support the results of pathology that showed there were no foci of sarcomatous transformation within this huge mass. However, the feasibility of diffusion-weighted images should be further studied because the diffusion-weighted MRI features of other vulvar masses have not been previously reported.

To this point, the lesion has not recurred during 1 year of follow-up. We believe that the possibility of recurrence is very low, because the mass was completely excised, and the resection margin was negative. However, we intend to pay attention to whether the mass recurs, particularly if the patient becomes pregnant or begins to take tamoxifen.

In conclusion, FESP is a hormone-related subepithelial stromal proliferation in the vulvovaginal region typically occurring in women of reproductive age that can grow to greater than 20 cm. Its differentiation from aggressive angiomyxoma is important and CE-MRI with diffusion-weighted images is helpful for this purpose. FESP can be cured by complete excision. Therefore, the physician's awareness of the differential features of FESP is needed to avoid unnecessarily aggressive management of the disease.

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