May 16th, 11:30 AM - 1:30 PM

Case of an adolescent girl with familial vulvar leiomyoma

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Case of an Adolescent Girl with Familial Vulvar Leiomyoma

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BACKGROUND

Uterine leiomyomas affect approximately 50% of premenopausal adult women[1] and can arise anywhere smooth muscle cells are located in the body.

Vulvar leiomyomas are a rare diagnosis comprising 0.07% of vulvar tumors, thought to arise from smooth muscle within the round ligament, erectile tissue, and dartos muscle.[2]

CASE DESCRIPTION

A 16yo cis-female noticed a painless vulvar mass that had increased in size over 7 months. Her mother reported having a history of recurrent vulvar leiomyoma requiring multiple surgical excisions.

Single digit vaginal exam revealed a solid well-circumscribed vulvar mass that involved the right labia majora and perineum, extending caudally to the level of ischial spine.

Pelvic MRI completed, with findings consistent with large vulvar mass measuring over 9cm (Figure 1).

A multidisciplinary surgical procedure with pediatric gynecology and colorectal surgery was performed. A paramedian skin incision was made along the right labia majora, extending inferiorly the perineum (Figure 3). The mass was excised in its entirety from within the pararectal space, and dead space closed in layers with interrupted sutures followed by skin closure. Patient admitted overnight with vaginal packing, which was removed after 24 hours and the patient discharged to home on post-op day one.

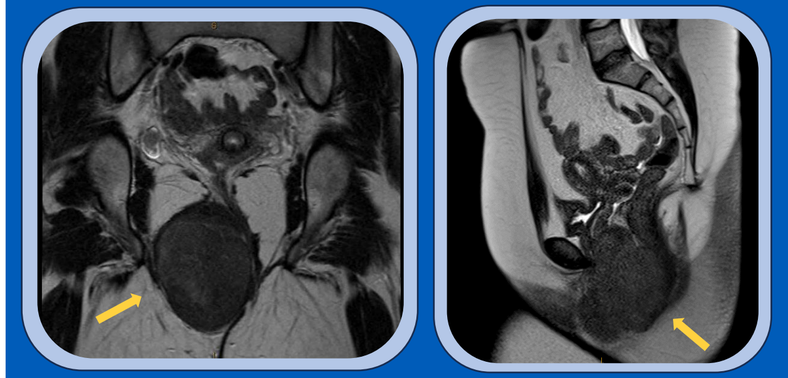

Fig 1. Pelvic MRI revealed lobular right-sided vulvar mass measuring 9.6 x 9.1 x 7.2cm, displacing the rectum to the left without evidence of tumor invasion. MRI features most suggestive of leiomyoma.

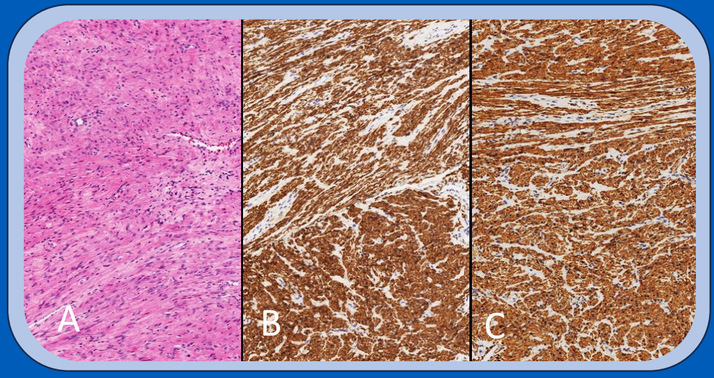

Fig 2. A. Spindle cells arranged in intersecting fascicles, with indistinct borders, eosinophilic fibrillary cytoplasm, and cigar-shaped nuclei **B.** Positive Desmin staining, **C.** Positive smooth muscle actin staining

Fig 3. Initial paramedian incision made on the labia majora. Mass was excised using blunt dissection and bipolar cautery device. The specimen was 12.2cm in diameter, weighing 319g

COMMENTS

There have been over 100 cases of vulvar leiomyoma reported in the literature.[3] ***Only 3 cases have been previously reported in adolescents.***[4-6]

No cases of familial vulvar leiomyoma have been described, although there are known syndromes of hereditary cutaneous leiomyomas, uterine fibroids, and renal cancer.[7]

Preoperative MRI was useful for delineation of anatomic location of the mass, surgical planning, and confirmation of clinical suspicion of leiomyoma given smooth muscle consistency on T2 weighted images.[2]

Prior reports of surgical management of vulvar leiomyoma describe performing excision at the mucocutaneous junction of the labia.[4] In our case, surgical excision was performed via a ***paramedian skin incision*** to preserve the integrity of the vagina given extensive involvement of mass within the pararectal space.

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