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CASE REPORT

Laparoscopic Approach for Recurrent Huge Vulval Mass

Kavitha Yogini Duraisamy[1] · Devi Balasubramaniam[1] · Malathi Ezhilmani[1] · Palanivelu Chinnusamy[2]

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Introduction

Aggressive angiomyxoma is a benign mesenchymal tumor with very low potential to metastasize. The tumor was termed as aggressive due to its slow growth with extensive local invasion [1]. Most commonly it is diagnosed only on histopathology. Etiology is unknown and usually affects vulva, perineal region, buttocks of women in reproductive age. It is a rare and recurrent tumor with less than 250 cases reported in the literature [2]. Considering its nature of high local recurrence and extensive invasion, proper diagnosis, management and long-term follow-up are very essential. No treatment modality has proven to be of any benefit for complete cure for this condition. However, total excision of the mass should be opted to prevent the chance of recurrence. Despite complete surgical removal, recurrence rate of aggressive angiomyxoma is reported as high as 72%.

Dr. Kavitha Yogini Duraisamy MS., Fellow in Endogynecology, FMAS, is Head of Department in the Department of Endogynecology of Gem Hospital and Research Centre, Coimbatore, Tamil Nadu, India; Dr. Devi Balasubramaniam MS., Fellow in Endogynecology, FMAS, is Consultant in the Department of Endogynecology of Gem Hospital and Research Centre, Coimbatore, Tamil Nadu, India; Dr. Malathi Ezhilmani, MS., is Fellow in the Department of Endogynecology of Gem Hospital and Research Centre, Coimbatore, Tamil Nadu, India; Dr. Palanivelu Chinnusamy, MS, MCh, DNB, FACS, FRCS(Hon) Ed., is Director in Gem Hospital and Research Centre, Coimbatore, Tamil Nadu, India.

✉ Kavitha Yogini Duraisamy
yoginianandvij@rediffmail.com

[1] Department of Endogynecology, GEM Hospital and Research Centre, 45A, Pankaja Mills Road, Ramanathapuram, Coimbatore, Tamil Nadu 641045, India

[2] Gem Hospital and Research Centre, Coimbatore, Tamil Nadu, India

Case Report

We present a case of a 28-year-old unmarried girl with a large, fleshy, pedunculated painless mass on the left labia majora with difficulty in walking. She underwent local surgical excision four times in the past with history of recurrence within 3–6 months following every surgery. Last surgery was done 3 years back and swelling appeared 3 months following surgery and gradually progressed to present size. The previous histopathology reports were not available. There was a huge soft non-tender swelling in left labia majora around 18 × 10 cm involving labia minora, fourchette distorting the vulval introitus and urethra. It was ill-defined having variegated appearance with smooth surface. We decided to proceed with MRI in view of recurrent soft tissue mass. MRI suggested dumbbell-shaped mass in the left vulva with pelvic extension with thick vascular pedicle. Hemangioma was ruled out, and hence, we did not do MRI angiogram/venogram. FNAC done showed hemorrhagic material. In view of benign nature of huge mass, recurrence following multiple local excision and MRI showing pelvic extension, combined laparoscopic and vulval excision was planned for the patient to achieve complete removal of the mass. Patient was followed for 12 months and had no recurrence (Fig. 1).

Surgical Technique

Laparoscopic findings: uterus normal size, tubes and ovaries were normal. With the help of the vaginal assistant, vulval mass was pushed inside. A diffuse vague mass was seen lateral to left uterosacral ligament. As MRI suggested a large vascular pedicle supplying the mass-unilateral (left) temporary occlusion of anterior division of internal iliac artery ligation was done before proceeding with excision. Peritoneum was incised lateral to left ureter and anterior division of internal iliac artery was ligated with No 1-0 PDS. Incision was extended down along left lateral pelvic wall and posterior leaf of broad ligament up to left uterosacral ligaments. Left para-rectal space was developed by dissecting between left internal iliac artery

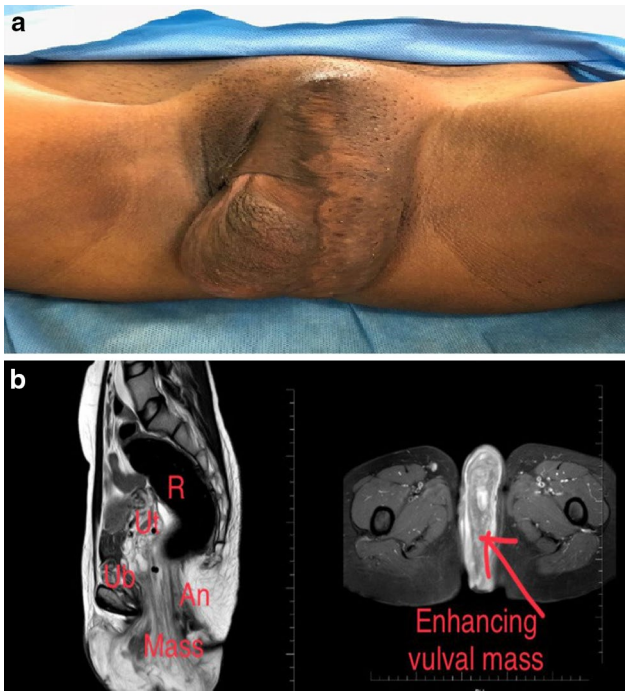

Fig. 1 **a** Preoperative image of a huge left vulval mass. **b** (i) Sagittal T2 MRI image showing mass, Ub—urinary bladder, Ut—uterus, R—rectum, An—anal canal. (ii) Coronal T1 MRI image showing contrast enhancing vulval mass

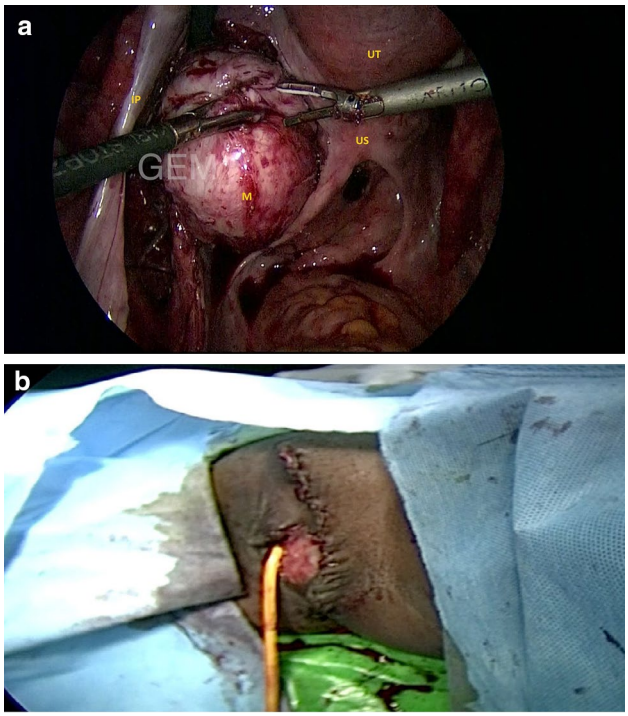

Fig. 2 **a** Intra-operative image of removal of left vulval mass showing M—vulval mass with pelvic extension, US—uterosacral ligaments, UT—uterus, IP—infundibulopelvic ligament. **b** Postoperative image after complete excision

and left ureter. Anteriorly uterovesical fold was opened, and bladder was dissected down. On deep dissection between left ureter and uterosacral ligament, a thin-walled cyst of 6×6 cm was noted which drained mucinous fluid. Cyst wall was peeled from adjacent structures. Ureterolysis was done. Another ill-defined soft tissue mass 8×8 cm was noted posterior to uterine vessels and lateral to uterosacral ligament in left parametrium on the lower part of cervix extending into left para-colpos. With blunt and sharp dissection using harmonic, mass was released from adjacent structures and multiple feeding vessels were coagulated/clipped. Ill-defined mass 12×8 cm extending into para-colpos and left lateral border of vagina and inferior to ureter and uterine vessels extending to floor of para-rectal space into levator ani and into left vulva was excised with vulval skin with the aid of vaginal assistant by pushing the mass inside through the rent in the levator ani. Levator ani was sutured laparoscopically with No 2-0 PDS continuous sutures. Excess vulval skin was excised, and vulval reconstruction was done (Fig. 2).

Discussion

Clinically, it may be misdiagnosed as angiomyxolipoma, hamartoma, hemangioma, Bartholin cyst, lipoma, Gartner cyst, angiomyofibroblastoma and other smooth muscle tumors. Grossly, they are soft, partly circumscribed masses with gelatinous appearance on cut section. Microscopic appearance of these lesions shows numerous thick-walled vessels in a loose collagenous stroma with stellate-shaped neoplastic cells. Exact pathogenesis is not known. MRI is the gold standard for diagnosis [3]. It is superior to a CT when determining the extent of the tumor and its relation to the pelvic floor.

Partial excision can be opted in view of high operative morbidity. It requires multimodal treatment combining surgical and medical management to treat recurrent angiomyxoma. In general, excision of these lesions is challenging as they have the same consistency as that of normal connective tissue and hence have a high propensity for local recurrence (36–72%) [4]. Recurrences usually occur in first 5 years after surgery in which about 70% recurrence in first 3 years and late recurrences up to 14 years have been reported.

Wide surgical excision is the best treatment of choice. To achieve complete excision, we may need to remove adjacent normal fascia and muscles. However, major organs such as rectum or bladder are spared as this highly morbid extensive surgery cannot be justified for such recurrent tumors. If the tumor extends above the pelvic diaphragm, an abdominal-perineal approach is the best treatment option. The high recurrence is commonly due to incomplete excision mainly

because of improper diagnosis. Radiation and chemotherapy are less effective due to its low mitotic activity.

This tumor is common during reproductive age due to hormonal dependency. This is explained by its positive estrogen and progesterone receptors. Hormonal treatments with gonadotropin-releasing hormone analogues and tamoxifen have been tried, but its role is not clearly defined. Another modality of treatment is angiographic embolization of the tumor. This helps by shrinking the tumor size before surgery. However, recurrences after initial embolization are common. As late recurrences are well known, these patients should be counseled about the need for long-term follow-up. Periodic clinical examination may be insufficient to detect recurrence. Imaging studies such as MRI can detect recurrence early, but there are no guidelines about their follow-up. This woman has been advised regular checkups. After follow-up for 1 year, she had no recurrence.

Conclusion

This is a challenging case for a surgeon, due to its wide extension and difficulty in differentiating it from normal adjacent structures. If we can define its extent of invasion and anatomical location, any vulval tumor can be successfully managed by surgical excision. If one can achieve complete excision, the recurrence rate will be low. It is reported that aggressive angiomyxoma needs long-term follow-up. Laparoscopic approach for vulval mass is difficult, but it provides various advantages for the patient. We conclude that laparoscopic approach is also feasible and safe; we can achieve complete removal of the tumor. In our patient, the procedure was successfully performed by laparoscopy.

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Compliance with Ethical Standards

Conflict of interest Dr. Kavitha Yogini Duraisamy, Dr. Devi Balasubramaniam, Dr. Malathi Ezhilmani and Dr. Palanivelu Chinnusamy declare that they have no conflict of interest.

Human Rights Statement There is no violation of human rights.

Informed Consent Informed written consent for publication of this article was obtained from patient and her relative.

Ethical Statement All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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About the Author

Dr. Kavitha Yogini Duraisamy Dr. Kavitha Yogini Duraisamy is an experienced gynec laparoscopic surgeon. She currently heads the Department of Endogynecology at GEM institute. She graduated from Kasturba Medical College, Manipal, and completed postdoctoral fellowship in Endogynecology from GEM institute. She was awarded honorary FMAS by AMASI for her extensive work in Gynec laparoscopy. She is well versed with advanced laparoscopic surgeries including pelvic and para-aortic lymphadenectomy, paravaginal cystocele repair, laparoscopic sacrocolpopexy, frozen pelvis and severe pelvic endometriosis, gynec oncological surgeries. She has organized regular laparoscopic training courses for the past 8 years. She has been a faculty and organizing member in several national and international conferences. She has a keen interest in research, and her special interests are robotic and laparoscopic gynecologic oncology and urogynecology.