

Aggressive Angiomyxoma of Vulva: Wide local excision case report

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Abstract

Aims and Background: Aggressive angiomyxoma is rare tumor of pelvic and perineal organs, occurring usually in women of reproductive age, and carrying a high tendency to local infiltration and relapse. It is non-malignant but can be locally aggressive and frequently recurs. Misdiagnosis is frequent. Wide excision is the preferred method of treatment but the challenge is to perform the degree of extirpation necessary to prevent recurrence. Most literature about Aggressive angiomyxoma consists of isolated case reports.

Presentation of Case: We reported a case of 35-year-old Indian woman complaining of a slow and progressive growth of a left vulvar labia majora pedunculated tumor with long a stalk was detected second time after first resection 2yrs back, measuring of 15.0x8.0 cm. Wide local surgical resection of the tumor were performed. Histopathology diagnosed a large aggressive angiomyxoma with uninvolved resection margins. Patient remains without recurrence at 8.0-months follow up.

Discussion and Conclusion: Non Aggressive angiomyxomais rare but should be excluded in any large vulval mass. We expect that awareness accompanied with wide free safe margin excision has important role to prevent the recurrence of tumor.

Keywords: Aggressive angiomyxoma, vulva.

1. Introduction

Aggressive angiomyxoma (AA) is a rare, locally aggressive myxoidmesenchymal neoplasm arising in the pelvis and perineal regions. The name angiomyxoma was chosen because of the similarity to myxoma and the notable vascular component. The term Aggressive angiomyxoma was not coined until 1983 but similar tumors were described as early as in the 1860s. AA was first described in 1983 by Steeper and Rosai.[1] They presented AA as a vulvar polyp clinically and diagnosed it histologically. It usually occurs in women, especially middle-aged; 95% of total cases are found in females.[2] However, it has later been reported in males. In men, the tumour involves analogous sites including the scrotum and inguinal area and usually appears at an older age.[3] It's suspected there might be a relation with hormonal status that might explain a female to male ratio of slightly more than 6:1. Have been reported about 150 cases in the world medical literature.[4] This benign tumor usually presents in middle aged adults as single nodule or polypoid lesion that may be clinically confused with skin tag or neurofibroma.[5]

2. Case report

35-year-old Indian woman para four, presented with polypoidal, slow-growing painless mass for two year

duration, arising from the left labia majora which was causing her difficulty in walking and restricting her free movement. There were no associated symptoms like pain and no urinary or bowel complaints. Past and menstrual history was unremarkable. Patient was admitted in August 2016, and clinical examination showed a large skin covered non-tender, pedunculated polypoidal mass involving the middle part of left labia majora. The soft doughy tumor mass attached arising from vulva of size 15.0x8.0 cm (Figure 1 and 2).



Figure 1: The soft doughy tumor mass attached arising from vulva



Figure 2: The soft doughy tumor mass attached arising from vulva

Speculum examination for vagina and cervix were also normal. The inguinal lymph nodes were not enlarged. An ultrasound revealed a subcutaneous hypo-echoic cyst in the left labium major. The uterus, adnexa, and other pelvic organs were normal. CT scans showed a mass of soft tissue originating from the left side of the vulva, without infiltration. Laboratory tests for tumour markers were negative.

The patient underwent wide local excision of the tumor remnants and partial vulvectomy surgery. The pedicle was ligated at the base, excised and labial reconstruction was done. (Figure 3 and 4)

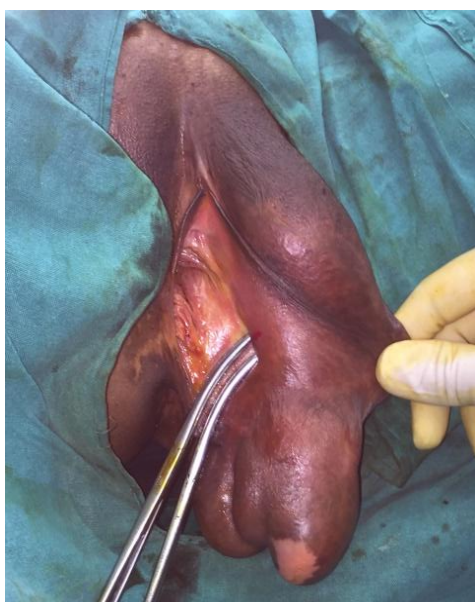


Figure 3: The pedicle was ligated at the base, excised and labial reconstruction was done.



Figure 4: The pedicle was ligated at the base, excised and labial reconstruction was done

On slicing, cut section was homogenous, white fleshy, soft to feel. No areas of haemorrhage or necrosis were identified.

Microscopically, the tumor was composed of spindle and stellate-shaped cells embedded in a loose myxoid matrix. These cells showed low to moderate cellularity and had eosinophilic cytoplasm with no significant nuclear pleomorphism and mitosis. Variable-sized thin-walled capillaries and thick-walled vascular channels were haphazardly arranged in the stroma. Some of these vessels showed perivascular hyalinization in the vascular walls. (Figure 5)

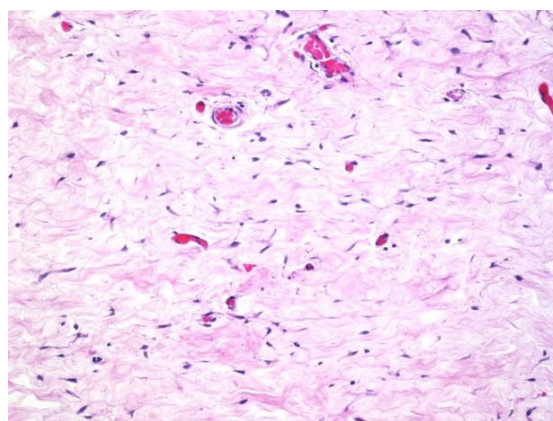


Figure 5: vessels showed perivascular hyalinization in the vascular walls

3. Discussion

At least 100 cases of aggressive angiomyxoma involving the female pelvis or perineum, or both, have been described. At least 14 of these primarily involved the vulva. It is an unusual tumor derived from fibroblasts or myofibroblasts with nuclei that have no atypical features or

mitotic activity. There have been no reported deaths attributed to this tumor. All of the reported cases of AAM of the vulva have presented as painless, slow-growing, polypoid, cyst-like masses in females between the ages of 15 and 77 years. The peak incidence of AAM is at 31-35 years of age.[6]

Our case is in this age range. Most AAM of the vulva are >10 cm in their maximal dimension on presentation.[7] While the AAM in our case is considerably larger than this, it is by no means the largest. Chen *et al* reported an AAM, arising from the right labium majus and extending into the retroperitoneum, which weighed 19.8 kg.[8]

AA is a hormonally responsive tumor; it is positive for ER and/ or PR and is believed to arise from specialized mesenchymal cells of the pelvic or perineal region.

Clinically, the differential diagnosis includes vulval abscess, Bartholin abscess, Gartner's duct cyst, vaginal cyst, vaginal mass or polyp, vaginal prolapse, pelvic floor hernia, obturator and levator hernia, vulval lipoma, pedunculated vulval leiomyoma, and vulvar hypertrophy with lymphedema.

Clinical diagnosis is usually difficult but with clinical information, histologic diagnosis of AAM can be made without the need for further studies.

As these tumors have a strong propensity for local recurrence, wide excision without causing morbidity is recommended. Recurrences generally occur in the first 5 years after primary surgery, and about 70% occur in the first 3 years, but late recurrences up to 14 years have been reported.[9] Preoperative angiographic embolization, preoperative external beam irradiation, and intraoperative electron beam radiotherapy are useful to decrease the chances of local recurrences.[10]

Various methods have been used to treat recurrences, and many options have been successful. Hormonal treatment with tamoxifen, raloxifene or gonadotropin-releasing hormone analogues has been shown to reduce tumor size. Moreover, hormonal therapy may also help to achieve complete excision in large tumors and can be used to treat recurrences.[11] Hormonal treatment with a gonadotropin-releasing hormone agonist (GnRh-a) can be applied for small primary AAM, in addition to adjuvant therapy for residual tumors.[12]

4. Conclusion

Although a rare diagnosis, AAM can present with unusual features. Detailed radiological examination is helpful in detecting the problem, but histology is the gold standard for diagnosis. Wide excision is curative and prognosis of such tumors is good. Long-term follow-up is necessary and MRI is the preferred method for detecting recurrences.

References

- [1] Steeper TA, Rosai J. Aggressive angiomyxoma of the female pelvis and perineum. Report of nine cases of a distinctive type of gynecologic soft-tissue neoplasm. *Am J Surg Pathol* 1983; 7:463-75.
- [2] Han-Geurts IJ, van Geel AN, van Doorn L, M den Bakker, Eggermont AM, Verhoef C. Aggressive angiomyxoma: multimodality treatments can avoid mutilating surgery. *Eur J Surg Oncol* 2006; 32:1217-21.
- [3] Behranwala K. A. and Thomas J. M., "Aggressive' Angiomyxoma: A Distinct Clinical Entity," *European Journal of Surgical Oncology*, 2003; 29,(7): 559- 563.
- [4] Mathieson A., Chandrakanth S., Yousef G. *et al.*, "Aggressive Angiomyxoma of the Pelvis: A Case Report," *Canadian Journal of Surgery*, 2007; 50 (2): 228-229.
- [5] Allen PW, Dymock RB, Mac Cormac LB. Superficial angiomyxomas with or without epithelial components. Report of 30 tumors in 28 patients. *Am J Surg Pathol*, 1988; 12(7): 519-530.
- [6] Chan YM, Hon E, Ngai SW, Ng TY, Wong LC. Aggressive angiomyxoma in females: is radical resection the only option? *Acta Obstet Gynecol Scand* 2000; 79: 216-20.
- [7] Behranwala KA, Thomas JM. 'Aggressive' angiomyxoma: a distinct clinical entity. *Eur J Surg Oncol* 2003; 29: 559-63.
- [8] Chen L, Schink JC, Panares BN, Barbuto D, Lagasse LD. Resection of a giant aggressive angiomyxoma in the Philippines. *Gynecol Oncol* 1998; 70: 435-9.
- [9] Haldar K, Martinek IE, Kehoe S. Aggressive angiomyxoma: a case series and literature review. *Eur J Surg Oncol* 2010; 36:335-9.
- [10] Nyam D, Pemberton JH: Large aggressive angiomyxoma of the perineum and pelvis: An alternative approach. *Diseases of the Colon & Rectum* 1998; 41: 514-516.
- [11] McCluggage WG, Jamieson T, Dobbs SP, Grey A. Aggressive angiomyxoma of the vulva: dramatic response to gonadotropin-releasing hormone agonist therapy. *Gynecol Oncol* 2006; 100:623-5.
- [12] Shinohara N, Nonomura K, Ishikawa S, Seki H, Koyanagi T: Medical management of recurrent aggressive angiomyxoma with gonadotropin-releasing hormone agonist. *International Journal of Urology* 2004; 11: 432-435.