



Case Report

Aggressive angiomyxoma of vulva in 28-years old patient: A case report of second recurrence

Mehrangiz zamani^a, Mina Mollabashi^b, Narges Mehrabi^c, Shima Alizadeh^{d,*}

^a Department of Obstetrics & Gynecology, Hamadan University of Medical Science, Hamadan, Iran

^b Department of Radiology, Hamadan University of Medical Science, Hamadan, Iran

^c Hamadan University of Medical Sciences, Hamadan, Iran

^d Gynecologist; Board Certified at Tehran University of Medical Science, Tehran, Iran

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ABSTRACT

Introduction: Aggressive angiomyxoma is an uncommon mesenchymal tumor in women who are in reproductive age, that occurring in the pelvis and perineal zone with a high risk of local infiltration and recurrence.

Case presentation: We describe a case of a 28-year-old woman with a huge recurrent vulvar aggressive angiomyxoma.

Clinical Discussion: Our patients underwent surgery and tumor resection for two times but had relapsed every 2 years through 5 years and finally she underwent total vulvectomy. The patient received Decapeptide for 3 months to prevent tumor recurrence after surgery and to date, there has been no evidence of local recurrence.

Conclusion: Aggressive angiomyxoma is a rare tumor with high recurrence rate. The best treatment is surgical resection by experienced Gyn-oncologist in teamwork and tertiary Hospital.

1. Introduction

Aggressive angiomyxoma (AA) is an uncommon myxoid mesenchymal tumor, involves the vulvovaginal, perineal and pelvic zone especially in middle-aged women [1]. This tumor has a high trend to local recurrence but a low trend to metastasize [2]. The tumor cells have desmin, smooth muscle actin, vimentin, and estrogen and progesterone receptors [3]. It is expected to grow during pregnancy and sensitive to hormonal manipulation. For AA of the vulva, which is a rare presentation of AA, radical surgery with wide margins is the first line of treatment although treatment with gonadotropin releasing hormone agonists is an adjunct treatment [3,4].

2. Case presentation

A 28-year-old Iranian virgin girl presented with a gradually increasing huge pedunculated mass arising vulva and pubis, first on 2011. The family history was negative. The patient justified for being a Case report and she signed a written consent. At the first time the tumor was resected by general surgeon (2011), and the pathology revealed aggressive angiomyxoma. She didn't have any follow-up. The tumor had

relapsed after 2 years and she went under surgery again by Gynecologist. medical therapy (Decapeptide) was used for patient to prevent recurrence (2013). The tumor has relapsed again after 2 years, on May 2015 we visited the patient in oncology ward of Fatemeh hospital. Local examination showed a well-circumscribed pedunculated polypoidal mass measuring about 20 × 15 × 10cm. The mass was painless, soft and spongy in inspissations with no detectable inguinal lymph nodes (Fig. 1).& (Fig. 2).

The pelvic MRI was performed to assess any invasive lesion in anal or bladder or any intra pelvic area. MRI revealed a huge pedunculated solid tumor in supra and pre pubic which involved labium majora and minora. There were some invasive lesions in anal canal with circumferential view in posterior of anal. She underwent excision of the tumor by the Gyn-Oncologist at tertiary referral hospital. Plan of surgery was total vulvectomy, the whole tumoral tissue removed totally up to endopelvic fascia (Fig. 3). Anal canal and its margin were checked carefully by colorectal surgeon at the time of surgery but no evidence of tumoral tissue was detected. Due to the fact that the patient had two surgeries and the resulting changes occurred in soft tissue after operation, a signal change occurs that can remain for a long time, even a few years, and is false positively similar to tumor invasion in the MRI. We

* Corresponding author.

E-mail addresses: m.zamani@umsha.ac.ir (M. zamani), Vah.mina@yahoo.com (M. Mollabashi), narges.mehrabi95@gmail.com (N. Mehrabi), Alizadeh.Shima@gmail.com (S. Alizadeh).

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Fig. 1. The appearance of the tumor before surgery; lateral view.



Fig. 2. The appearance of the tumor before surgery; Anterior view.

received one ovoid shaped mass measuring $12 \times 12 \times 10$ cm. External surface is predominantly covered by gray and wrinkled skin except for an area 7cm in diameter which is rough and adipose tissue attached to it. Cut surface are tan-yellow, solid and glistening which was sent to pathology ward, Microscopy showed a polypoid lesion covered by skin and composed of a hypo cellular stroma without atypia or obvious mitotic activity and variably sized vessels with dilated Lumina compatible with AA and free of tumor margins. **She visited annually in the office. Fortunately, there is no evidence of recurrence. She has her menstrual cycles.** Decapeptide began and till now (6years) the patient is free of AA.

3. Discussion

AA is usually found in the genital, perineal, and pelvic area [5]. Angiomyxomas usually grows slowly and a high capacity for local infiltration and local recurrence. Our patient experienced surgery three times due to recurrent recurrence. Although angiomyxomas are locally aggressive, far metastasis (to the lung) has been reported in only two cases [6,7]. Although the nature of tumor is non-metastasizing, but there are some reports of multiple metastases in women treated first by total excision and later affected by metastatic disease [6,7]. Fortunately in this Case despite locally recurrence, we didn't detect any metastasis. According to investigations, one-fourth of these tumors are pedunculated [2]. As the feature of our patient, the common presentation of AA is a large pedunculated, lobulated and painless bank or an ill-defined swelling of the vulva or pelvic region [8]. The origin of tumor is mesenchymal cells or multipotent perivascular progenitor cells of the pelvic-perineal region, with myofibroblastic and fibroblastic features [9]. From the Microscopic point of view, the tumor consists of stellate and spindle-shaped neoplastic cells set in a loosely myxoid stroma with



Fig. 3. The tumor side after total resection.

scattered vessels of varying caliber. These tumors cells have Estrogen Receptor (ER), progesterone receptors (PR), vimentin, and desmin [6, 10]. The specimen in our case was examined by expert pathologist, and also evaluated for ER and PR receptors.

Lipoma, Gartner duct cyst, Bartholin cyst, myxolipoma, myxomas,

myxoid leiomyoma, myxofibrosarcoma, angiomyofibroblastoma, and myxoid liposarcoma are in the list of AA differential diagnosis [11–13]. The obvious vascular component in AA, like what is in the myxoid liposarcomas, helps in ruling out most of these neoplasms. The absence of lipoblasts in AA rule out liposarcoma [13]. Comparing angiomyofibroblastoma, AA has less vessels with thicker walls [2,3].

Primary treatment with tamoxifen, raloxifene or gonadotropin-releasing hormone analogues is effective. In addition, preoperative prescription of GnRH agonists leads to tumor shrinkage which might increase the chance of complete excision [14].

Because of the infiltrative nature of the tumor and absence of well-defined capsule, it is difficult to excise the tumor thoroughly, while the surgical excision is the choice of treatment [15,16]apsule. This Case managed by general surgeon at the first time and we don't know about the margin of the tumor. She didn't receive any hormonal therapy. She had no surveillance until the recurrence. Since the rarity of vulvar tumors, we recommend management in team work under the GYN-Oncologist leadership. Nearly all authors believes complete resection of tumor with tumor free margin reduces recurrence rate (16) but according to further investigations, 50 %–70 % relapse are reported after surgical resection [17]. Recurrences may occur from months to several years after excision (2 months–15 years) [12].

In our Case, finally the tumor excised completely up to endopelvic facias with tumor free margin and postoperative period is still uneventful. **We confronted with a kind of recurrent tumor and anatomical disruption. Due to past perineal surgeries and the extensive recurrence we had no choice but to have total vulvec-tomy for achieving free margin.**

Furthermore, we suggested radiotherapy to the patient before surgery in the hope that she underwent more limited resection. She refused radiotherapy because of adverse effects of radiation in young women.

The patient received Decapeptide for 3 months to prevent tumor recurrence after surgery and to date, there has been no evidence of local recurrence. As late recurrences are known, all patients need to be counseled about the need for long-term follow-up. This Case report has been reported in line with the improved SCARE checklist. The SCARE Guidelines were published in 2020 [18].

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent was obtained from the patient for publication of this Case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Availability of data and materials

All data generated or analysed during this study are included in this published article [and its supplementary information files].

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

Alizadeh SH; Writing - Review & Editing; critical revision; Mollabashi Mina: data collection, Investigation, Zamani Mehrangiz: performed the surgery. Investigation; Narges Mehrabi: Writing - Original Draft

All authors have read and approved the manuscript.

Registration of research studies

Not Applicable

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