EJGO European Journal of Gynaecological Oncology

Aggressive angiomyxoma of the vulva: report of three cases and literature review

Weiguo Song, Zhen Shen, Ying Zhou, Dabao Wu

Department of Obstetrics and Gynecology, Anhui Provincial Hospital, Anhui Medical University, Hefei (China)

Summary

Aggressive angiomyxoma (AAM) is a rare mesenchymal tumour primarily located in the pelvis and perineal regions of women in the reproductive age. Due to the similar presentation to fibroma, lipoma, major vestibular gland cysts, and femoral hernia, the preoperative diagnosis is difficult. The standard treatment for AAM is total excision and strict follow-up. In this paper, the authors report three cases of AAM admitted to the present hospital and review the relative literatures.

Key words: Aggressive angiomyxoma (AAM); Perineal mass; Recurrences.

Introduction

The diagnosis of aggressive angiomyxoma (AAM) is very difficult due to the aspecific presentation. Three cases of AAM are reported and their management is discussed.

Case Report

The first case was a 43-year-old female patient with a vulvar mass admitted to the present hospital on December 3, 2014.Gynecological examination showed a palpable, non-tender, solid mass located on the left side of labia majora. Ultrasound examination showed a well-circumscribed soft tissue mass with heterogeneous internal echoes. The vulva lump resection was performed after the endotracheal general anesthesia on December 9, 2014. Intraoperatively a lobulated cystic mass on the left vulva was found and its boundary was unclear and its size was approximately 3×2×2 cm and 5×4×3 cm, respectively. Postoperative histopathological examination of the specimen showed aggressive angiomyxoma (two spindle-shaped nodules, 4.5×2.5×2 cm, 6×3×2.5 cm). Immunohistochemically, the cells showed positive staining with desmin, SMA, h-Caldesmon, estrogen and progesterone receptors, and CD34. S100 was negative in the tumor cells. Postoperative diagnosis was aggressive vascular mucinous cystadenoma of the left vulva. The patient has not recurred since the last follow-up.

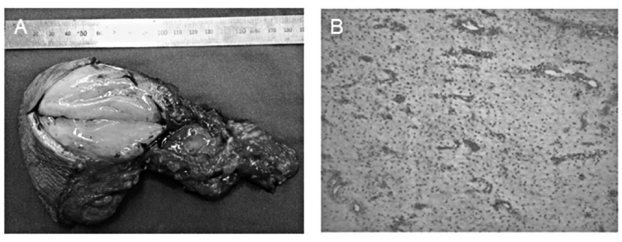

Figure 1. — A) Dissection of the AAM of case 2. B) Pathologic image of AAM of case 2.

The second case was a 26-year-old female patient with a previous history of vulva tumor resection operation and admitted to the present hospital on April 10, 2016. In her medical history, the patient had a complaint of a large painless swelling on the vulva and underwent a vulvar lump resection five years ago. Postoperative histopathological examination showed aggressive angiomyxoma of the vulva. Gynecological examination revealed a mass on the outside of the labia majora and the size was measured about 8×6×5 cm. On April 15, 2016, the patient underwent transperineal surgery with local excision of the mass. Intraoperatively on the right vulva, there was an obvious outward mass, soft, rubbery, with a size of 8×6 cm, and the boundary was unclear. After dissection, the section appeared translucent and gelatinous (Figure 1A). Postoperative histopathological examination revealed that the tumor was composed of spindled and stellate-shaped cells with ill-defined cytoplasm intermingled with collagen fibers and thin vessels in a myxoid background (Figure 1B). On immunohistochemical examination, the cells showed positive staining with desmin, vimentin, SMA, h-Caldesmon, estrogen and progesterone receptors, and CD34. S100 was negative in the tumor cells. Postoperative diagnosis was aggressive angiomyxoma of right vulva. The patient has not recurred since the last follow-up.

The third case was a 28-year-old female patient that presented with a history of an indolent mass on her right vulva of about three years. The mass size was approximately 3×2 cm in size and she did not care about it. After that, she found that the mass had grown gradually and then underwent a vulvar lump resection at local hospital. Postoperative histopathology report suggested that the vulva's fat and fibrous tissues included tumor-like hyperplasia with cystic lesions. After the operation, the mass size decreased, but did not disappear completely. The patient felt the that vulvar lump increased significantly and was admitted to the present hospital for surgical treatment on June 18, 2016. Gynecological examination showed a lump outside of the right vulva, in the shape of an egg. Ultrasound examination displayed that the mass was well-circumscribed soft tissue with uneven internal echoes. The mass measured approximately 3.2×1×2.3 cm. MRI sequences demonstrated a perineal mass located on the right paravulvar region and ischioanal fossa with intermediate signal on T1WI (Fig-

Revised manuscript accepted for publication May 9, 2017

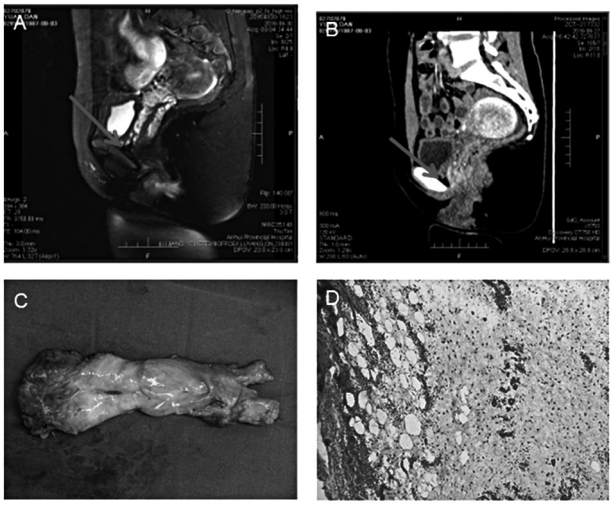

Figure 2. — A) MRI of the AAM of case 3. B) CT of the AAM of case 3. C) Gross image of the AAM of case 3. D) Immunohistochemical staining of the tumor of case 3 (1:10).

ure 2A). On T2WI, the lesion showed a high signal with hypointense strands of fibrovascular tissue mimicking a swirled appearance. Contrast-enhanced abdominal tomography (CT) showed that the right perineal area was a soft tissue density shadow (Figure 2B), the border was less clear, and the largest cross-sectional size was approximately 5.0×3.0 cm. The laparoscopic exploration to rule out the possibility of inguinal hernia and transperineal lump resection was performed on July 13, and a right vulva tumor surface incision of about 6 cm was made and the tumor was completely removed. Histopathological results showed that the right vulva had an aggressive angiomyxoma (11× 6×2 cm) (Figure 2C). Microscopic examination revealed that the tumor cells presented as a short fusiform shape, sarciniform or irregular sparse arrangement, interstitially containing myxoid matrix (Figure 2D). The tumor boundary was unclear and was inserted into the adipose tissue. Immunohistochemical markers demonstrated that the tumor cells showed positive staining with desmin, SMA, and estrogen and progesterone receptors. S100 and CD34 were negative in the tumor cells. Postoperative diagnosis was aggressive angiomyxoma of the vulva on the right side. The patient was not recurred follow-up till now.

Discussion

AAM is an uncommon mesenchymal tumor which is mainly derived from the pelvic and perineal regions including vulva, vagina, bladder, and rectum [1]. It is difficult to define the exact incidence of AAM among the other intra-abdominal mesenchymal tumors due to the reason of its rarity [2, 3]. AAM is considered as an aggressive tumor due to the neoplastic property of the blood vessels [4] and its high inclination of local infiltration [5] and local relapse [6]. The diagnosis of AAM is very difficult because it is often asymptomatic until the tumor reaches a larger size [7]. AAM normally manifests as an indolent swelling [8] located around the genitofemoral region [9]. AAM is often misdiagnosed as a vulvar abscess, Bartholin's gland cyst, vaginal prolapse, or groin/femoral hernia that may lead to pointless surgical interventions [10]. Immunohistochemical staining [11] of AAM discloses high positivity for desmin, vimentin, estrogen and progesterone receptors; however it generally reveals negativity for S-100 protein [12, 13]. USG shows a hypoechoic cystic mass and usually remains insufficient for diagnosis [14]. CT shows a well-defined, hypoattenuated enhanced mass with a swirling appearance which is detected in about 83% of the patients [15,16]. MRI [17] is more useful than the other imaging technologies [18]. On T1-weighted MRI, the tumor shows isosignal compared to the muscles, whereas on T2 high signal intensity is detected. Furthermore, because the size of the tumor is often underestimated by clinical examination, these imaging technologies also help us in deciding the surgical strategy [19].

The up-to-date treatment of AAM is complete surgical excision with tumor-free margins [20, 21]. However, there is still a debate about the treatment because of high recurrence rate in spite of wide surgical excision scope [22]. The recurrence rate is reported with a wide range, from 33% to 83% [23]. Recurrence occurs mostly within the first three years. All adjuvant treatment approaches remain debatable. Chemotherapy exerts no beneficial results for adjuvant therapy because of low mitotic activity of the tumor [24]. Embolization of the tumor has been reported as an alternative approach. However, it remains insufficient due to the extensive vascular network of the tumor. The main localization of AAM, which is limited to reproductive organ region, and the positive estrogen and progesterone receptors' status of the tumor suggest that AAM may be a hormone-responsive tumor [25]. Several beneficial results with tamoxifen or GnRH agonist [26] have been reported [27]. Although the majority of the authors have reported no advantage in using radiotherapy, it can be a good alternative treatment in patients who are resistant to antihormonal therapy, those with recurrence, or in those with a tumor excision that could generate a high death rate [28]. In conclusion, AAM of the vulva is a rare benign neoplasm with local invasive nature and has a good prognosis despite its risk to recur. Regular follow-up should be recommended after surgical treatment.

Acknowledgements

This study was supported by the National Natural Science Foundation of China (grant nos. 81272881 and 81001168).

References

[1] H.C. Lin, C.C. Liu, W.Y. Kang, H.L. Ke, C.C. Li, W.J. Wu, C.H. Huang: "Huge aggressive angiomyxoma: a case report and literature review". *Kaohsiung J. Med. Sci.*, 2006, *22*, 301.

[2] Das B.P., Baruah D., Medhi K.B., Talukder B.: "An aggressive angiomyxoma of vulva - A rare entity - A case report". *J. Midlife Health*, 2016, *7*, 140.
[3] Kura M.M., Jindal S.R., Khemani U.N.: "Aggressive angiomyxoma of the vulva: An uncommon entity". *Indian Dermatol. Online J.*, 2012, *3*, 128.
[4] Cheah P.L., Looi L.M., Sivanesaratnam V.: "Aggressive angiomyxoma of the vulva with an unusual vascular finding". *Pathology*, 1993, *25*, 250.
[5] Narayama C., Ikeda M., Yasaka M., Sagara Y.S., Kan-no Y., Hayashi I., *et al.*: "Aggressive Angiomyxoma of the Vulva with No Recurrence on a 5-year Follow up: A Case Report". *Tokai J. Exp. Clin. Med.*, 2016, *41*, 42-45.
[6] Sengupta S.K., Saha S.P., Roy H., Sarkar A.N.: "Recurrent aggressive angiomyxoma of the vulva - a rare presentation". *J. Clin. Diagn. Res.*, 2014, *8*, OD01.
[7] Gungor T., Kaleli A., Kuzey G.M.: "Aggressive angiomyxoma of the vulva and vagina. A common problem: misdiagnosis". *Eur. J. Obstet. Gynecol. Reprod. Biol.*, 2004, *112*, 114.
[8] Mandal S., Roy S., Khurana N.: "Aggressive angiomyxoma of the vulva presenting as a pedunculated swelling". *Indian J. Pathol. Microbiol.*, 2008, *51*, 259.
[9] Varras M., Akrivis C., Lekkou P., Kitsiou E., Demou A., Antoniou N.: "Aggressive angiomyxoma of the vulva: our experience of a rare case with review of the literature". *Eur. J. Gynaecol. Oncol.*, 2006, *27*, 188.
[10] Sozutek A., Irkorucu O., Reyhan E., Yener K., Besen A.A., Erdogan K.E., *et al.*: "A Giant Aggressive Angiomyxoma of the Pelvis Misdiagnosed as Incarcerated Femoral Hernia: A Case Report and Review of the Literature". *Case Rep. Surg.*, 2016, *2016*, 9256749.
[11] Q. Wang, X. Lin, W. Zhang, Y. Gao: "Aggressive angiomyxoma of the vulva: intra-operative pathological diagnosis is useful in deciding the scope of surgery and reducing recurrence". *Acta Chir. Belg.*, 2012, *112*, 79.
[12] H.Q. Wei, J. Liu, J.J. Feng: "Clinicopathological analysis of aggressive angiomyxoma". *Zhonghua Yi Xue Za Zhi*, 2012, *92*, 1553.
[13] Amezcua C.A., Begley S.J., Mata N., Felix J.C., Ballard C.A.: "Aggressive angiomyxoma of the female genital tract: a clinicopathologic and immunohistochemical study of 12 cases". *Int. J. Gynecol. Cancer*, 2005, *15*, 140.
[14] Wolf B., Hom L.C., Handzel R., Einenkel J.: "Ultrasound plays a key role in imaging and management of genital angiomyofibroblastoma: a case report". *J. Med. Case Rep.*, 2015, *9*, 248.
[15] H. Wu, W. Liu, H. Xu, D. Wang, A. Ouyang A.: "Aggressive angiomyxoma of the pelvis: a series of four cases and literature review". *Eur. J. Gynaecol. Oncol.*, 2015, *36*, 610.
[16] Outwater E.K., Marchetto B.E., Wagner B.J., Siegelman E.S.: "Aggressive angiomyxoma: findings on CT and MR imaging". *AJR Am. J. Roentgenol.*, 1999, *172*, 435.
[17] Tariq R., Hasnain S., Siddiqui M.T., Ahmed R.: "Aggressive angiomyxoma: swirled configuration on ultrasound and MR imaging". *J. Pak. Med. Assoc.*, 2014, *64*, 345.
[18] Brunelle S., Chetaille B., Lelong B., Piana G., Sarran A.: "Aggressive angiomyxoma with diffusion-weighted magnetic resonance imaging and dynamic contrast enhancement: a case report and review of the literature". *Case Rep. Oncol.*, 2013, *6*, 373.
[19] X. Li, Z. Ye: "Aggressive angiomyxoma of the pelvis and perineum: a case report and review of the literature". *Abdom. Imaging*, 2011, *36*, 739.
[20] Choi H., C. Park, Y.I. Ji: "Alternative surgical approaches for aggressive angiomyxoma at different sites in the pelvic cavity". *Obstet. Gynecol. Sci.*, 2015, *58*, 525.
[21] Iyibozkurt A.C., Yasa C., Schmieta D.E., Gungor-Ugurlucan F., Topuz S., Berkman S.: "Aggressive angiomyxoma of the pelviperineum: surgical treatment through a perineal incision". *J. Minim. Invasive Gynecol.*, 2011, *18*, 541.
[22] Salman M.C., Kuzey G.M., Dogan N.U., Yuce K.: "Aggressive angiomyxoma of vulva recurring 8 years after initial diagnosis". *Arch. Gynecol. Obstet.*, 2009, *280*, 485.
[23] Kiran, G., Yancar S., Sagar H., Kiran H., Coskun A., Arikan D.C.: "Late recurrence of aggressive angiomyxoma of the vulva". *J. Low. Genit. Tract Dis.*, 2013, *17*, 85.
[24] Wang Z., Liu Y., Yang L., Gu L., He Y., Huang D., Zhu G.: "Maxillary aggressive angiomyxoma showing ineffective to radiotherapy: a rare case report and review of literature". *Int. J. Clin. Exp. Pathol.*, 2015, *8*, 1063.
[25] McCluggage W.G., Patterson A., Maxwell P.: "Aggressive angiomyxoma of pelvic parts exhibits oestrogen and progesterone receptor positivity". *J. Clin. Pathol.*, 2000, *53*, 603.
[26] Lee C.W., Yoon J.H., Park D.C., Lee S.J.: "Aggressive angiomyxoma of the vulva treated by using a gonadotropin-releasing hormone agonist: a case report". *Eur. J. Gynaecol. Oncol.*, 2011, *32*, 686.
[27] McCluggage W.G., Jamieson T., Dobbs S.P., Grey A.: "Aggressive angiomyxoma of the vulva: Dramatic response to gonadotropin-releasing hormone agonist therapy". *Gynecol. Oncol.*, 2006, *100*, 623.
[28] Ribaldone R., Piantanida P., Surico D., Boldorini R., Colombo N.,Surico N.: " Aggressive angiomyxoma of the vulva". *Gynecol. Oncol.*, 2004, *95*, 724.

Corresponding Author:
YING ZHOU, M.D.
Department of Obstetrics and Gynecology
Anhui Provincial Hospital, Anhui Medical University
#17 Lujiang Road
Hefei City, Anhui Province (China)
e-mail: caddie1234@gmail.com