Aggressive Angiomyxoma of Vulva in Pregnancy: A Case Report

Rinchen Zangmo¹ · Sunesh Kumar¹ · Neeta Singh¹ · Jyoti Meena¹

Received: 8 January 2016 / Accepted: 19 March 2016 / Published online: 31 March 2016
© Federation of Obstetric & Gynecological Societies of India 2016

About the Author

Dr. Rinchen Zangmo has done her MBBS from Government Medical College Srinagar. She obtained the degree of M.D. Obstetrics and Gynecology from All India Institute of Medical Sciences, New Delhi, in June 2013. She is also a Diplomate of National Board (DNB) in Obstetrics and Gynecology. Presently, she is doing senior residency in the Department of Obstetrics and Gynecology at All India Institute of Medical Sciences.

Introduction

Aggressive angiomyxoma is a rare soft-tissue tumor occurring on the genital, perineal and pelvic organs in women of childbearing age. Steeper and Rosai first described it in 1983 [1]. This condition can, however, occur in perimenopausal women, men and children. It is a slow-growing tumor, but it

is locally invasive and 30–40 % cases present with relapse after treatment. It is important to diagnose this condition correctly because the tumor is locally infiltrative and requires wide excision and follow-up [1].

Case Report

A 21-year-old female presented to our outpatient department at 8 months of pregnancy with a vulval mass. She was primigravida at 32 weeks of pregnancy with a painless vulval mass. She noticed the mass at 5 months of pregnancy when it was the size of an almond. The mass increased to the present size in 3 months duration, but without any pain or discomfort. She gave a history of mass in the same location twice in the past. First was 18 months back, which was excised in a local hospital, and it reappeared after 8 months of excision for which repeat excision of the mass was done. The mass was around 5 cm in size on both the occasions according to the patient. No

Dr. Rinchen Zangmo is a senior resident in Department of Obstetrics and Gynecology at All India Institute of Medical Sciences; Dr. Sunesh Kumar is a Professor in Department of Obstetrics and Gynecology at All India Institute of Medical Sciences; Dr. Neeta Singh is a Professor in Department of Obstetrics and Gynecology at All India Institute of Medical Sciences; Dr. Jyoti Meena is an Assistant professor in Department of Obstetrics and Gynecology at All India Institute of Medical Sciences.

✉ Rinchen Zangmo
rinchen.zn@gmail.com

¹ Department of Obstetrics and Gynecology, All India Institute of Medical Sciences, New Delhi, India

histopathological records of the previous excision procedures were available.

On examination, uterus was 32 weeks size with a single live fetus in cephalic presentation. Local examination of the perineum revealed a vulval mass arising from the upper right labia minora measuring around 7×8 cm. The mass was soft in consistency with bosselated surface and was non-tender. There was no ulceration or bleeding on the surface and no vaginal or rectal involvement. The patient was asked to follow-up after 2 weeks or earlier if needed. She did not turn up till 37 weeks of pregnancy when she started having pain in the mass. On examination, the mass had increased in size measuring 15×15 cm this time (Fig. 1). There was no tenderness and no signs of secondary infection in the mass. Vaginal examination for pelvic assessment was attempted, but it was not possible as the mass was obstructing the introitus almost completely. Patient was admitted, and an elective cesarean section was done at 38 weeks on May 12, 2014 under antibiotic cover. Postoperative period was uneventful, and the patient was discharged in stable condition on day 4 of surgery. She was asked to follow-up after 6 weeks for excision of the mass. A wide local excision of the mass was done on July 1, 2014 under spinal anesthesia. Its size was 18×15 cm with no gross infiltration of surrounding areas. The mass was sent for histopathological examination. Patient was discharged on the third day of surgery with an advice to follow-up after 2 weeks with the histopathology report. The report confirmed angiomyxoma of vulva (Fig. 2) with 2.5 cm of tumor-free margin. The patient was then asked to follow-up after 6 months. There was no new growth at 6 months (Fig. 3). She is on 6-monthly follow-up since then. Her last visit was on December 17, 2015, and she was doing fine with no recurrence.

Fig. 1 Vulval mass at 37 weeks pregnancy (approx. 15×15 cm in size)

Fig. 2 Histopathology slide with features of angiomyxoma

Fig. 3 Follow-up after 6 months; no new growth

Discussion

Steeper and Rosai first described aggressive angiomyxoma in 1983. The pathogenesis of AAM is not clear. Studies have demonstrated a translocation at the level of chromosome 12 with a consequent aberrant expression of the high-mobility group protein isoform IC involved in DNA transcription. Almost exclusively, it involves women of child-bearing age, although very rare cases have been diagnosed in perimenopausal women, men and children. On gross examination, these tumors are characteristically soft, bulky masses with a smooth external surface, measuring from <5 cm to >10 cm [1]. A mixture of spindle or stellate cells in a loosely myxoid stroma characterizes the microscopic appearance of these neoplasms [1]. Several studies have demonstrated estrogen receptor (ER) and/or progesterone receptor (PR) positivity within aggressive angiomyxoma [2]. This explains the rapid increase in size of the tumor during the last 2 months of pregnancy in our case.

Although aggressive angiomyxoma is a locally invasive tumor, it may rarely metastasize to distant organs as in frank malignancies. Junzu et al. reported a case of aggressive angiomyxoma encasing the ureters and colon with invasion of the inferior vena cava and associated pulmonary metastasis. Thus, aggressive angiomyxomas can be regarded as tumors of intermediate malignancy having an unpredictable and even sometimes unfavorable outcome, in a small percentage of patients [3].

Multiple treatment modalities have been described for the treatment of angiomyxoma. Complete surgical excision should be the treatment of choice whenever possible. Recurrence may, however, occur even with negative surgical margins. Thus, multimodal therapies may be required to treat recurrent angiomyxoma. Adjunct hormonal treatment with gonadotropin-releasing hormone analogs (GnRHa) has been described. The response with GnRHa has been attributed to the fact that many of these tumors express estrogen and progesterone receptors and are therefore sensitive to hormonal therapy. Second recurrence of a vulvar angiomyxoma following two prior surgical excisions in a 34-year-old female was strongly positive estrogen and progesterone receptors. The patient was treated with 3 months of GnRHa which led to complete resolution of the tumor [4]. Radiotherapy and chemotherapy are unlikely to be useful because of the low mitotic activity of the tumor.

To conclude aggressive angiomyxoma is an uncommon mesenchymal myxoid tumor. The rate of growth is increased in pregnancy owing to its hormone receptor

positivity. It is characterized by frequent local recurrence and is regarded as a non-metastasizing tumor. However, certain case reports have stated that this tumor can metastasize. Thus, periodic follow-up of the patient after treatment is required.

Compliance with Ethical Standards

Conflict of interest Authors Rinchen Zangmo, Sunesh Kumar, Neeta Singh and Jyoti Meena declare no conflict of interest.

Ethical Approval All procedures performed involving the human participant 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.

References

1. Fetsch JF, Laskin WB, Lefkowitz M, et al. Aggressive angiomyxoma: a clinicopathologic study of 29 female patients. *Cancer*. 1996;78:79–90.
2. McCluggage WG, Patterson A, Maxwell P. Aggressive angiomyxoma of pelvic parts exhibits oestrogen and progesterone receptor positivity. *J Clin Pathol*. 2000;53:603–5.
3. Junzu G, Bofeng C, Liping W. Aggressive angiomyxoma: an unusual presentation. *Korean J Radiol*. 2012;13(1):90–3.
4. Fine BA, Munoz AK, Litz CE, et al. Primary medical management of recurrent aggressive angiomyxoma of the vulva with a gonadotropin-releasing hormone agonist. *Gynecol Oncol*. 2001;81(1):120–2.