ANGIOMYXOMA OF VULVA

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PRESENTATION OF CASE

A 40-year-old female presented with a slow-growing mass on the left labia majora, since three years.

Local examination showed a well-circumscribed pedunculated polypoidal mass measuring 15 cm × 10 cm. The mass was non-tender and firm in consistency.

The inguinal lymph nodes were not enlarged. Her blood investigations and sonography of the abdomen revealed no abnormality. Sonography of the local part showed a large mass with peripheral vascularity, heterogeneous hyperechoic areas, thick echoes and non-vascular areas in the centre. There was a cystic area of size 2.8 × 2.2 near the stalk. Fine needle aspiration cytology smears revealed only blood without cellular elements.

Patient was planned for excision biopsy and all routine investigations were within normal limits [Figure 1].

PATHOLOGICAL DISCUSSION

On histopathology section showed hyperplastic stratified squamous epithelium. The subepithelium is loose and show inflammatory cell comprising of lymphocytes, plasma cells and proliferating capillaries.

There is also presence of dilated and congested thick walled blood vessel. The tumour was composed of spindle and stellate-shaped cells in a myxoid matrix. These cells had eosinophilic cytoplasm and lacked significant nuclear pleomorphism and mitosis. Also seen were variable-sized thin-walled capillaries and thick-walled vascular channels. Some of these vessels showed perivascular hyalinisation of their vascular walls. This was suggestive of AA.

Investigations

All routine investigations were within normal limits and serology was also negative.

Treatment

The main treatment for angiomyxoma is surgery to remove the whole tumour. Depending on where the tumour is, it may be difficult to completely remove it. The medical literature suggests that about 3 out of 10 (30%) aggressive angiomyxoma tumours will come back (recur). Sometimes they can keep coming back. Coping with this can be difficult.

Another treatment you may have is hormone therapy. Some angiomyxomas are hormone sensitive or hormone dependent. This means that they use hormones to grow. Hormone therapies can slow down or stop the growth of a tumour by either-

- Stopping hormones being made, or
- Preventing hormones from making cancer cells grow and divide.
- The hormone treatments that you may have include goserelin and tamoxifen before surgery if you have a large tumour, to make surgery easier or after surgery to reduce the risk of the tumour coming back.

The length of time you need to take is uncertain. This is a relatively new treatment and some women who have stopped taking it found the tumour grew again.

Because angiomyxoma is so rare, there is less information than for more common conditions. Most of what has been written about angiomyxomas in medical journals describes individual cases. This makes it difficult for us to generalise here, because so few people have this condition.

DISCUSSION OF MANAGEMENT

Angiomyxomas are classified either as superficial (also called as cutaneous myxoma). Superficial angiomyxomas usually present in middle-aged adults as a single nodule or a polypoidal lesion in the head and neck region that may be clinically confused with skin tag or neurofibroma. The stroma is made up of mostly oedema with little myxoid material. It occurs almost exclusively in the pelvic and perineal regions of women of reproductive age, but is occasionally reported in men (male-to-female ratio of 1: 6).^[1] The term "aggressive" denotes its propensity for local aggression and recurrences after excision. Usually, this tumour is non-metastasising, but there are reports of multiple metastases in women treated initially by excision and who later succumbed to metastatic disease.^[2,3] About one-fourth of these tumours are pedunculated. There is no complete consensus regarding the tumour pathogenesis. This hormonally responsive tumour is believed to arise from specialised mesenchymal cells of the pelvic-perineal region or from the multipotent perivascular progenitor cells, which often display variable myofibroblastic and fibroblastic features.^[4] This hypothesis is supported by the fact that the tumour cells express desmin and in some cases a smooth muscle acting along with desmin. Recent cytogenetic and molecular studies have identified a variety of genetic alterations involving the chromosome 12 in the region 12q13-15. A gene in this region called high-mobility group protein isoform I-C (HMGIC), which encodes proteins involved in the transcriptional regulation appears to have a role in the pathogenesis of this tumour. Detection of inappropriate HMGI-C expression using the immunoperoxidase technique with anti-HMGI-C antibody

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may potentially be a useful marker for microscopic residual disease.^[5]

Clinically, it may be misdiagnosed as Bartholin cyst, lipoma, labial cyst, Gartner duct cyst, levator hernia or sarcoma. Fibroepithelial stromal polyp, superficial angiomyxoma, angiomyoibroblastoma, cellular angiofibroma and smooth muscle tumours also need to be considered in the differential diagnoses of a polypoidal mass in the perineum. It is an infiltrative tumour, whereas angiomyoibroblastoma is well circumscribed (this characteristic can also be identified on Magnetic Resonance Imaging [MRI]). Also, it has thick-walled vessels, which are less numerous than the thin-walled vessels in angiomyoibroblastoma. On Computed Tomography (CT) scan, these tumours have a well-defined margin with attenuation less than that of the muscle. On MRI, these tumours show high signal intensity on T2-weighted images. The attenuation on CT and high signal intensity on MRI are likely to be related to the loose myxoid matrix and high water content of angiomyxoma.^[6] Recurrences may occur from months to several years after excision (2 months to 15 years).^[7] Despite the name is not that aggressive with only a 30% chance of recurrence, which is eminently treatable by excision with a 1 cm margin. Most of the patients have only one recurrence. Radiation therapy and chemotherapy are considered less-suitable options due to low mitotic activity. Hormonal manipulation with tamoxifen, raloxifene and gonadotropin-releasing hormone analogues has been shown to reduce the tumour size and may help to make complete excision feasible in large tumours and in the treatment of recurrence.^[8] Angiographic embolisation may also help in subsequent resection by shrinking the tumour as well as making it easier to identify it from surrounding normal tissues.^[9] As late recurrences are known, all patients need to be counselled about the need for long-term followup.

Magnetic resonance imaging is the preferred method for detecting recurrences.

REFERENCES

- [1] Han-Geurts IJ, van Geel AN, van Doorn L, et al. Aggressive angiomyxoma: multimodality treatments can avoid mutilating surgery. *Eur J Surg Oncol* 2006;32(10):1217–21.
- [2] Siassi RM, Papadopoulos T, Matzel KE. Metastasizing aggressive angiomyxoma. *N Engl J Med* 1999;341(23):1772.
- [3] Blandamura S, Cruz J, Vergara FL, et al. Aggressive angiomyxoma: a second case of metastasis with patient's death. *Hum Pathol* 2003;34(10):1072–4.
- [4] Alameda F, Munne A, Baro T, et al. Vulvar angiomyxoma, aggressive angiomyxoma and angiomyoibroblastoma: an immunohistochemical and ultrastructural study. *Ultrastruct Pathol* 2006;30(3):193–205.
- [5] Nucci MR, Fletcher CD. Vulvovaginal soft tissue tumors: update and review. *Histopathology* 2000;36(2):97–108.
- [6] Outwater EK, Marchetto BE, Wagner BJ, et al. Aggressive angiomyxoma: findings on CT and MR imaging. *AJR Am J Roentgenol* 1999;172(2):435–8.
- [7] Behranwala KA, Thomas JM. Aggressive angiomyxoma: a distinct clinical entity. *Eur J Surg Oncol* 2003;29(7):559–63.
- [8] McCluggage WG, Jamieson T, Dobbs SP, et al. Aggressive angiomyxoma of the vulva: dramatic response to gonadotropin-releasing hormone agonist therapy. *Gynecol Oncol* 2006;100(3):623–5.
- [9] Magtibay PM, Salmon Z, Keeney GL, et al. Aggressive angiomyxoma of the female pelvis and perineum: a case series. *Int J Gynecol Cancer* 2006;16(1):396–401.