CASE REPORT

Perineal aggressive angiomyxoma in a menopausal woman

Corentin Deckers,¹ Livia Dumitru,² Fabrice Corbisier²

¹Université Catholique de Louvain, Brussels, Belgium
²Service de Chirurgie Générale et Digestive, Grand Hôpital de Charleroi, Charleroi, Hainaut, Belgium

Correspondence to
 Corentin Deckers,
 corentindeckers@gmail.com

Accepted 1 November 2017

SUMMARY

Aggressive angiomyxoma (AA) is an uncommon mesenchymal tumour that is principally located in the soft tissues of the pelvis and perineum of young women. The primary features of this benign tumour are a local invasion, a high local recurrence rate and non-specific local clinical signs. We describe the case of a 58-year-old woman, initially treated for a Bartholin's cyst. Histological examination indicated the presence of an AA. The MRI showed a 7 cm soft tissue mass extending from the lateral wall of the vagina, into the left buttock and down into the subcutaneous tissue. We performed a radical excision with wide resection, which is considered the standard gold treatment.

BACKGROUND

To date, fewer than 300 cases of angiomyxoma have been reported in the literature.¹

Given the rarity of aggressive angiomyxoma (AA), we are presenting this case to alert medical practitioners (anatomopathologists, gynaecologists and general surgeons) to the clinical presentation, imaging characteristics, differential diagnosis and treatment of this tumour.

CASE PRESENTATION

We describe the case of a 58-year-old woman who had been menopausal for five years and was being treated with hormonal therapy. Her previous medical history included two natural deliveries. There was no patient history of venereal disease and no family history of gynaecological malignancies.

Over a 2-year period, the patient developed a buttock mass that was symptomatic when she was in a sitting position. A tumefaction in the deep tissue of her left buttock was noted on palpation that had a soft consistency and normal subcutaneous and cutaneous tissue. During this time, the patient underwent surgical excision of a Bartholin's cyst by a gynaecologist. The anatomopathological examination revealed an AA.

We performed an extensive resection with clear margins, which is considered the standard gold treatment,^{2,3} without any complication (figure 1).

The first and second follow-up by MRI performed at 3 and 8 months, respectively, revealed a cicatricial reorganisation of the site with no sign of any tumour recurrence. A CT scan of the thorax was also completely normal at 8 months.

INVESTIGATIONS

The radiological examination consisted of MRI that showed an isointense signal related to muscle on T1-weighted images and a hyperintense signal on T2-weighted images. A swirled appearance is commonly seen in most cases and was also present in our patient (figure 2).

The MRI performed at 8 months did not show any sign of recurrence (figure 3).

The immunochemical staining of our tissue sample was positive for oestrogen and progesterone receptors, desmin and vimentin.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of AA must include several other conditions, such as a Bartholin's cyst, groin or femoral hernia, vulvar abscess and other tumours, such as superficial angiomyxoma, angiofibroblastoma, myxoid lipomatous tumours and myxoid leiomyoma.

Angiofibroblastomas are usually located in the superficial vascular soft tissue in the vulva. They are well circumscribed and are usually smaller than 5 cm, in contrast to the larger size and infiltrating nature of angiomyxomas. The tumour cells appear rounded or more epithelioid than those in angiomyxomas and are often aggregated around blood vessels.⁴

Superficial angiomyxomas are often located at extragenital sites, such as the head and neck, although they can also involve the vulvovaginal region. On gross examination, angiomyxomas have a distinct lobular or multinodular architecture. Histologically, they are characterised by myxoid features but lack large-calibre vessels such as those seen in AA.⁴

Another entity that mimics the appearance of AA is the benign or malignant lipomatous tumour. The most important lesions are myxolipoma and myxoid liposarcoma, which have compositions similar to that of mature adipose tissue with a myxoid stroma that contains stellate or spindle cells, similar to AA. The characteristic finding in myxoid liposarcoma is the abundant adipose tissue containing well-organised lipoblasts and arborising capillary networks, in contrast to the disorganised thin-walled and thick-walled blood vessels characteristic of AA.

Myxoid leiomyoma is also a pelvic tumour that can grow very large and, like AA, is often mistaken for a Bartholin's cyst. Important histological features can differentiate these two entities, namely

CrossMark

To cite: Deckers C, Dumitru L, Corbisier F. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-222622

Figure 1 (A) Intraoperative image highlighting the extension of the tumour into the pelvis. (B) The resected specimen appears as a well-circumscribed and well-encapsulated, fleshy, gelatinous mass with finger-like projections into neighbouring tissues. (C) Once this tumour was removed, its marked tendency to displace rather than invade perineal structures could be seen.

a more focal myxoid change and the more classic smooth muscle differentiation. Additionally, myxoid leiomyoma contains more abundant eosinophilic cytoplasm than AA.⁴

TREATMENT

In all cases described in the literature, surgery is the standard of care in the treatment of AA,^{5,6} which is why we chose to perform a wide resection with clear margins (figure 1).

Even though achieving negative resection margins can be difficult in some cases, and complex and complete resection of the deep-seated tumours may cause fertility problems, extensive surgery remains the gold standard.⁶

According to a review of 111 cases by Chan *et al*⁶ there was no significant difference in disease-free maintenance between groups of patients with positive and negative resection margins or incomplete removal of the tumour. There was also no significant morbidity.⁶

Figure 2 (A) The preoperative sagittal MRI shows a 7×4×2 cm soft tissue mass extending from the lateral vaginal wall, into the left buttock and down into the subcutaneous tissue (sagittal T2-weighted MRI). (B) The tumour grew around the structures of the pelvic floor and had a well-defined margin. There is a hypointense signal on this T1-weighted image. (C) There is a typical 'swirled aspect' or layered internal structure of the angiomyxoma on this axial T2-weighted coronal image. (D) Coronal MR (T1-weighted image with gadolinium injection) shows close contact between the aggressive angiomyxoma and the lateral vaginal wall.

Figure 3 MRI follow-up performed at 8 months showed no sign of recurrence and a cicatricial reorganisation of the pelvic-perineal region. (A) Sagittal MRI (T2-weighted MR image). (B) Axial T1-weighted image. (C) T2-weighted coronal image. (D) Coronal MR T1-weighted image with gadolinium injection.

However, given that this tumour may potentially metastasise, we recommend achieving a clear margin resection if technically possible.

Although chemotherapy and radiotherapy are two other methods of management, they have no well-defined roles in the treatment of AA.⁷

The embolisation of the tumour vessels, together with radiation therapy in the preoperative period, may decrease the risk of recurrence.^{8,9}

Another adjuvant treatment method (described by Fine *et al*¹⁰) involves hormonal therapy, which is used mainly because the tumour occurs in premenopausal females and has oestrogen and progesterone receptors.¹¹

Only two case reports described the complete radiographic resolution of the tumour after treatment with gonadotropin-releasing hormone agonists.¹⁰

We did not start any adjuvant therapy in our patient.

OUTCOME AND FOLLOW-UP

On gross examination, it was a well-encapsulated tumour measuring 9×2.8×2.5 cm with a rubbery consistency and a gelatinous surface (figure 4).

Histological examination (figure 5) showed a sparsely cellular tumour with an eosinophilic stroma that contained haphazardly arranged blood vessels (capillaries and venules). The stroma was myxoid with collagen fibrils. The tumour cells were cytologically bland and had an ovoid or stellate appearance. No mitotic figures and no atypical presentation were described.¹²

Despite the associated morbidity, the prognosis of patients with AA is excellent.¹³

The recurrence rate at 5 years ranges from 25% to 47% in patients treated primarily with surgery. Only two cases of metastases have been described in the literature. The first patient had lung metastases, and the second patient, a 27-year-old woman, died from multiple lung metastases.¹⁴

The MRI (figure 3) and CT scan of the thorax performed at 8 months did not show any sign of relapse. We plan to perform the same follow-up every 6 months.

Figure 4 Macroscopic appearance.

Figure 5 Histopathological examination showing a loose myxoid stroma, widely scattered bland spindle cells with a stellate, ovoid appearance and variable-sized vascular channels. ((A) H&E $\times 5$. (B) H&E $\times 40$.)

DISCUSSION

Described for the first time by Steeper and Rosai in 1983, AA is a rare vulvovaginal mesenchymal benign neoplasm that occurs primarily in young women.² To date, fewer than 300 cases have been described in the literature. The diagnosis of AA is challenging because the tumour is usually asymptomatic.^{2,3} Histological examination and MRI are the keys to diagnosing aggressive angiomyxoma. AA has an excellent prognosis despite a high rate of recurrence and few cases of metastasis. Many therapies are described, but surgical excision with a long follow-up remains the gold standard treatment of these aggressive tumours.

Learning points

- Aggressive angiomyxoma is an uncommon mesenchymal tumour of the pelvic–perineal region in young women that is associated with a high rate of local recurrence and is often clinically mistaken for more common entities.
- Early recognition requires a high clinical suspicion because the tumour is rare, and the diagnosis is usually made histologically and with MRI.
- Surgery with clear margins is the gold standard treatment of aggressive angiomyxoma.

Contributors CD collected the data about the angiomyxoma, wrote our first draft in French, organised the figure, wrote the cover letter and submitted the case to BMJ. He also made the corrections for resubmission. LD translated our French draft in English, described the histological pattern, the differential diagnosis, and asked for the consent of the patient. FC revised and approved the final version of this paper.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2017. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- Husso A, Savola J, Gunn J. Successful surgical treatment of an aggressive pelvic angiomyxoma with intracardiac extension. *Ann Thorac Surg* 2017;104:e23–e25.
- 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.
- Fetsch JF, Stenman G. Deep aggressive angiomyxoma. In: Fletcher CDM, Unni K, Feds M, *Tumors of soft tissue and bone*. Lyon: IARC press, 2002.
- Sutton BJ, Laudadio J. Aggressive angiomyxoma. *Arch Pathol Lab Med* 2012;136:217–21.
- Fletcher CD, Tsang WY, Fisher C, *et al*. Angiomyofibroblastoma of the vulva. A benign neoplasm distinct from aggressive angiomyxoma. *Am J Surg Pathol* 1992;16:373–82.
- Halder K, Martinek IE, Kehoe S. Aggressive angiomyxoma: a case series and literature review. *Eur J Surg Oncol* 2010;36:335–9.
- Chan YM, Hon E, Ngai SW, *et al*. Aggressive angiomyxoma in females: is radical resection the only option? *Acta Obstet Gynecol Scand* 2000;79:216–20.
- Nyam DC, Pemberton JH. Large aggressive angiomyxoma of the perineum and pelvis: an alternative approach. Report of a case. *Dis Colon Rectum* 1998;41:514–6.
- Rhomberg W, Jasarevic Z, Alton R, *et al*. Aggressive angiomyxoma: irradiation for recurrent disease. *Strahlenther Onkol* 2000;176:324–6.
- Suleiman M, Duc C, Ritz S, *et al*. Pelvic excision of large aggressive angiomyxoma in a woman: irradiation for recurrent disease. *Int J Gynecol Cancer* 2006;16(Suppl 1):356–60.
- 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;81:120–2.
- Htwe M, Deppisch LM, Saint-Julien JS, *et al*. Hormone-dependent, aggressive angiomyxoma of the vulva. *Obstet Gynecol* 1995;86:697–9.
- McCluggage WG. A review and update of morphologically bland vulvovaginal mesenchymal lesions. *Int J Gynecol Pathol* 2005;24:26–38.
- Fetsch JF, Laskin WB, Lefkowitz M, *et al*. Aggressive angiomyxoma: a clinicopathologic study of 29 female patients. *Cancer* 1996;78:79–90.

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow