CASE REPORT

An unusual vaginal lump

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SUMMARY

A 46-year-old woman presented with a 2-week history of a painless golf ball sized lump in her vagina. Despite transvaginal ultrasound scan, biopsy and MRI, it was only after surgery had been performed that it became clear what the nature of the mass actually was.

Immunohistochemistry revealed the lump to be an aggressive angiomyxoma. Follow-up MRI showed residual (or recurrence of) angiomyxoma which was successfully treated with monthly injections of *Prostap* (leuporelin acetate), a gonadotropin-releasing hormone agonist. Further MRI showed complete resolution of the lesion.

for these specific characteristics, as the correct imaging modality needs to be selected, and the radiologist needs to be aware and informed about which particular characteristics to look out for. Similarly, a specialised laboratory may need to be accessed to determine the specific immunohistochemical features of the lesion.

Delays in these investigations and their interpretation can potentially lead to mismanagement or delay of these aggressive lesions, so it is worth being aware of this type of mass and how to identify it.

This case report presents a typical case and describes the main characteristics to look for on imaging and histology.

BACKGROUND

The term angiomyxoma describes an extremely rare benign tumour of unknown aetiology suspected to be myofibroblastic in origin.¹ Two types are recognised; 'superficial' and 'aggressive'. This report describes a case of aggressive angiomyxoma, a locally invasive neoplasm found predominantly in the pelvis and perineum of women of reproductive age, with women ~10 times more likely to be affected than men.²

Surgical management by wide local excision is the most effective treatment, although the rate of local recurrence is reported as being between 30 and 70%. The discovery that some of these tumours are hormone sensitive has led to the use of drugs such as gonadotropin-releasing hormone (GnRH) agonists and Selective Oestrogen Receptor Modulators (SERMs) to arrest growth or prevent recurrence.^{3 4}

Histological features of aggressive angiomyxoma include spindle and stellate cells in a loose hypocellular connective tissue matrix. There is typically low mitotic activity, and vessels within the lesion often display a range of thicknesses.⁵ In addition to sensitivity to progesterone and oestrogen, immunohistochemistry is often positive for desmin, smooth muscle actin, muscle-specific actin vimentin and negative for S100.^{1 6}

MRI is the gold standard investigation for these rare lesions which have characteristic appearance on the different modalities which can be an invaluable (and non-invasive) way to diagnose, assess and follow-up such lesions.^{7 8}

Although aggressive angiomyxoma is generally accepted to be a rare condition, (only a few hundred recorded cases)³ we are aware of three such cases in Wales in the past few years. These can be identified either by their characteristics on MRI or via immunohistochemistry.

However, unless clinicians are aware of the possibility of aggressive angiomyxoma they will not look

CASE PRESENTATION

A 46-year-old woman presented to her general practitioner with a 2-week history of a painless lump in her vagina which she assumed to be a prolapse. There were no associated urinary symptoms or dyspareunia, and no history of intermenstrual or post-coital bleeding. The patient had delivered three healthy children vaginally without complication and had been attending regularly for cervical smear tests with negative results. Her menstrual cycle was regular; 5 days of bleeding every 28–30 days with no associated menorrhagia or dysmenorrhoea. The patient was otherwise well with no medical problems and denied any weight loss or reduction in appetite.

On examination a large golf ball sized swelling was evident in the left lateral vaginal wall and visible at the vaginal opening. The mass was fixed to underlying structures, was soft but not reducible and there was no uterine descent. It was initially thought to be a cystic lesion but did not yield fluid on aspiration.

INVESTIGATIONS

The nature of the mass was not immediately apparent. MRI was undertaken to explore the nature and extent of the lesion, and seemed to demonstrate a fluid-filled structure in the left vaginal wall. The structure extended inferiorly to the perineum and superiorly to the upper third of the vaginal wall, with a maximal diameter of 8.8 cm sagittally and 6 cm axially. The origin of the mass appeared to be mucosal, and there was no evidence of connection to bowel, and no solid component or suspicious features seen.

MRI pelvis prior to surgery showed the lesion in the left lateral vaginal wall extending into the perineum through puborectalis and levator ani muscles (figures 1 and 2).

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Figure 1 T2 sagittal preoperative.

Figure 2 T2 coronal preoperative.

Transvaginal ultrasound scan was also carried out, which confirmed that it was in fact solid and demonstrated increased vascularity (figure 3).

Biopsies were sent for histopathological analysis, which reported squamous epithelium covered mucosa expanded by a myxoid lesion comprising prominent dilated and congested blood vessels in oedematous stroma with occasional scattered fibroblasts. No evidence of cellular atypia, stromal overgrowth or cellularity was seen and the impression was of a benign fibroepithelial polyp.

DIFFERENTIAL DIAGNOSIS

The most likely differentials for the lump were sarcoma and lymphoma, which have serious consequences for the patient, but require different modes of treatment. Histopathology reported squamous epithelium covered mucosa expanded by a myxoid lesion comprising prominent dilated and congested blood vessels in oedematous stroma with occasional scattered fibroblasts. No evidence of cellular atypia, stromal overgrowth or cellularity was seen and the impression was of a benign fibroepithelial polyp.

Figure 3 Transvaginal ultrasound Doppler scan of the lesion showing a solid structure with increased vascularity.

TREATMENT

Surgical excision of the structure was performed. Removal of the vaginal mass was performed in the lithotomy position, under general anaesthetic. Bivalve speculum and bimanual vaginal examinations were performed to confirm preoperative findings. The patient was catheterised to facilitate the surgery and prevent trauma to the urethra.

The mass was identified in the left lateral wall. The mass had increased in size by this time and was now protruding beyond the introitus by 4 cm. To develop the tissue planes, normal saline was infiltrated around the mass and a circumferential incision was made at the base of the mass. The mass was separated from the vaginal wall by blunt enucleation and dissection, until the base and pedicle of the mass was reached. The structure was multilayered and did not contain fluid. The base of the pedicle was deep in the ischio-rectal fossa, extending through puborectalis and levator ani muscles. The pedicle was clamped and cut and the mass was removed. After haemostasis was achieved, the vaginal wall was closed and a vaginal pack and urinary catheter were inserted. These were left in situ for 24 hours. The mass was sent for further histopathological investigation and later immunohistochemistry.

OUTCOME AND FOLLOW-UP

The surgical specimen was sent for further histological analysis, the results of which changed the differential diagnosis. The initial histology report of the surgical sample described a soft tissue neoplasm comprising spindle and stellate cells in an oedematous and collagenous matrix with both thin and thick-walled blood vessels, a dense lymphocytic infiltrate surrounded many blood vessels. Some collagen condensation was seen around a few blood vessels. Occasional multinucleate giant cells were also noted. No evidence of mitotic activity was seen and no atypical cells found (figure 4). The histologist reported that the excision margins of the specimen were clear.

This information narrowed the list of differentials to aggressive angiomyxoma, angiofibroma and cellular angiofibroma.

Immunohistochemistry of the sample was positive for vimentin, oestrogen receptors (ER), progesterone receptors (PR), desmin and focally with smooth muscle actin, which supported a diagnosis of aggressive angiomyxoma (figure 5).

MRI performed 3 months after the operation showed a small lesion, suggestive either of residual disease or local recurrence (figures 6 and 7).

The lesion is best seen on 3 plans T2-weighted sequences with similar signal characteristics as the original lesion.

The regional multidisciplinary team discussion concluded that despite the initial report of a clear excision margin, the lesion

Figure 4 Spindle and stellate cells in an oedematous and collagenous matrix (H&E, ×200).

Figure 5 Immunohistochemical staining (desmin, ×100).

was more likely to be residual disease than recurrence of the lesion, and a posterior exenteration would be necessary to achieve a clear excision margin. Considering the hormonal status of the lesion and the fact that there are few reports of GnRH analogue treatment showing complete response and regression of recurrent aggressive angiomyxoma tumours,⁹ it was decided to put the patient on a monthly injection of 3.75 mg of Prostag for 6 months and perform follow-up MRI after 3 months. The potential side effect of menopausal symptoms was explained to the patient.

After 3 months follow-up MRI showed no sign of residual disease. The patient needs no further treatment at present but will be followed up long term in case of future recurrence.

Figure 8 shows complete resolution of the lesion after treatment with Prostag.

DISCUSSION

Similar published cases:

Aggressive angiomyxoma was first named in an American case series of nine in 1983. The authors coined the term aggressive angiomyxoma:

...to emphasize the neoplastic nature of the blood vessels and its locally infiltrative and recurrent nature

The cases were treated surgically, and after long-term follow-up four patients experienced recurrences and none developed distant metastases.¹⁰

Figure 6 T2 axial postoperative.

Figure 7 T2 sagittal postoperative.

Figure 8 T2 axial posthormone.

Another series of nine was reviewed in 1985 which focused on the pathological characteristics of aggressive angiomyxomas, including their reactivity to actin. Two of the patients were men, all of the six cases which were followed up experienced recurrence, and again there were no cases of metastases.¹¹

An example of a case managed medically is that of a 34-year-old woman who twice experienced recurrence after surgical excision and was successfully treated with 6 months of a GnRH agonist.⁴

The radiological features of aggressive angiomyxoma were explored in a 2003 paper which recommended the use of MRI if such a neoplasm was suspected.⁷

There are two documented cases of metastasising aggressive angiomyxoma, both were fatal cases in women involving the lungs.¹²

Aggressive angiomyxoma in men was explored in an American paper in 2006 which cited 39 cases in men; finding that male lesions can also be sensitive to oestrogen and progesterone.¹³

A 2010 literature review from a British Hospital stated that fewer than 250 cases had been reported since 1983 worldwide including their case series of 7 women.³

Features of aggressive angiomyxoma on MRI: MRI is the best imaging modality to show tissue characteristics and local extension of the lesion.⁷

Typical MRI features of aggressive angiomyxoma are swirled strands aligned with the craniocaudal axis,⁸ best seen on T2-weighted images where the overall lesion is hyperintense relative to adjacent pelvic muscle and the swirled strands are slightly hypointense. On T1-weighted images the lesion has a homogeneous isosignal to muscles, and the swirled strands enhance strongly on T1-weighted PC. The lesion shows hyper-signals on both diffusion-weighted imaging and apparent diffusion coefficient indicating T2 shine-through effect similar to some other vascular lesions like haemangiomas.

As seen earlier, ultrasound scan confirms the solid and vascular nature of the lesion, when its true nature is confused to be a cyst or polyp on MRI, as in this case.

On CT scan the lesion is usually hypodense to muscles.

Conclusion: Aggressive angiomyxoma is a rare but treatable condition. Diagnosis can be made on the basis of immunohistochemical characteristics or the MRI findings. MRI is also valuable tool in its preoperative planning and postoperative

Learning points

- ▶ Aggressive angiomyxoma occurs in the reproductive system of women of reproductive age.
- ▶ Although it is benign, it is locally invasive.
- ▶ Aggressive angiomyxoma can be identified preoperatively on MRI or immunohistochemistry by distinctive characteristics.
- ▶ Treatment can be surgical or hormonal depending on the extent of the invasion, the needs and wishes of the patient, and the sensitivity of the individual lesion.

assessment. A multidisciplinary approach may be necessary using the expertise of gynaecologists, histopathologists, urologists, radiologists and colorectal surgeons. Treatment may be conservative, medical and/or surgical depending on the site and size of the lesion and the individual needs of the patient.

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Contributors SCS assisted with surgery, interviewed the patient, gained consent for the report, and wrote the case report except the radiological and surgical details. HS contributed the sections on the radiological features of the case. IA identified the case, performed the surgery, identified other cases in the locality, found original papers relevant to the literature review, wrote the section on the surgical details, edited the final paper and is guarantor.

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REFERENCES

- 1 Amezcua CA, Begley SJ, Mata N, *et al.* Aggressive angiomyxoma of the female genital tract: a clinicopathologic and immunohistochemical study of 12 cases. *Int J Gynaecol Cancer* 2005;15:140–5.
- 2 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.
- 3 Haldar K, Martinek IE, Kehoe S. Aggressive angiomyxoma: a case series and literature review. *Eur J Surg Oncol* 2010;36:335–9.
- 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;81:120–2.
- 5 McGluggage WG. A review and update of morphologically bland vulvovaginal mesenchymal lesions. *Int J Gynaecol Pathol* 2005;24:26–38.
- 6 Fetsch JF, Laskin WB, Lefkowitz M, *et al.* Aggressive angiomyxoma: a clinicopathologic study of 29 female patients. *Cancer* 1996;78:79–90.
- 7 Jayadevan NN, Sohaib SA, Thomas JM, *et al.* Imaging features of aggressive angiomyxoma. *Clin Radiol* 2003;58:157–62.
- 8 Karwacki GM, Stöckli M, Kettelhack C, *et al.* Radiographic diagnosis and differentiation of an aggressive angiomyxoma in a male patient. *J Radiol Case Rep* 2013;7:1–6.
- 9 Bai HM, Yang JX, Huang HF, *et al.* Individualized managing strategies of aggressive angiomyxoma of female genital tract and pelvis. *Eur J Surg Oncol* 2013;39:1101–8.
- 10 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;7:463–76.
- 11 Bégin LR, Clement PB, Kirk ME, *et al.* Aggressive angiomyxoma of pelvic soft parts: a clinicopathologic study of nine cases. *Hum Pathol* 1985;16:621–8.
- 12 Blandamura S, Cruz J, Vergara L, *et al.* Aggressive angiomyxoma: a second case of metastasis with patient's death. *Hum Pathol* 2003;34:1072–4.
- 13 Idrees MT, Hoch BL, Wang BY, *et al.* Aggressive angiomyxoma of male genital region. Report of 4 cases with immunohistochemical evaluation including hormone receptor status. *Ann Diagn Pathol* 2006;10:197–204.

Patient's perspective

Until a biopsy of my vaginal growth was suggested, I had not been particularly concerned about its nature. Luckily, the biopsy was performed within one week; but six weeks of waiting for biopsy results was excruciatingly difficult. I feel extremely fortunate and relieved that the tumour was benign. I do have concerns that the tumour may recur now that the anti-hormone injections have stopped. My consultant is discussing options with other medical professionals who have experience with angiomyxoma cases. I have been very impressed, over the past two years, with the care and dedication of healthcare professionals involved with my case.

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