Online Case Report

A solitary encapsulated pelvic aggressive angiomyxoma

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Aggressive angiomyxoma (AAM) was first reported in 1983 as a distinct, slow growing, benign but locally infiltrative, soft tissue tumour. It usually arises in the pelvic and perineal organs, mostly in women. A 47-year-old woman was found to have a large encapsulated retroperitoneal aggressive angiomyxoma. The mass was completely excised via abdomino-perineal approach, and no recurrence noted on MRI at 19 months' follow-up. The encapsulation of this tumour together with other reported rare presentations, suggest an isolated mesenchymal cell origin. A review of the literature is provided.

Key words: Aggressive angiomyxoma – Pelvic neoplasm – Retroperitoneal – Retroperitoneal aggressive angiomyxoma

Aggressive angiomyxoma (AAM) is a rare entity of benign, slowly growing but locally infiltrative soft tissue myxoid neoplasm, thought to be of mesenchymal origin.¹⁻⁸ It affects women 7 times as often as men.^{9,10} It was also reported in children as young as 11 years old of both sexes.¹¹ It has a predilection to occur in the pelvic and perineal organs.¹⁻⁴

AAM was first reported in 1983 by Steeper and Rosai.² We have discovered 180 reported cases, mostly in women; 70 cases were anatomically pelvic and/or retroperitoneal. Four cases only have been reported from the UK.^{6,7} We have found reports of involvement of every pelvic organ, including the urinary bladder¹² and uterus.¹³

The true origin of this tumour is unknown, but few articles have raised the possibility of an isolated mesenchymal cell origin.

In this article, we report an unusual occurrence of an encapsulated pelvic retroperitoneal aggressive angiomyxoma (AAM), as well as a review of the relevant literature.¹⁻¹⁹

Case report

A 47-year-old woman presented with a 3-month history of a vague, poorly defined, painless swelling over the right ischial tuberosity. On examination, the swelling was ill defined, non-tender, and difficult to palpate. The initial differentials were of lipoma or pelvic floor hernia.

MRI scans showed an approximately 17 cm x 10 cm large abnormal soft tissue mass within the pelvis. The mass was iso-intense on T1-images, but became very hyperintense on T2-images, with much whorled solid tissue within (Figs 1 and 2). There was no fluid or fat within it. The mass extended from the pubic symphysis posteriorly to the rectum along the right two-thirds of the pelvis and extended as far as S1 level superiorly, retroperitoneally and inferiorly as far as the level of the anal sphincter. The MRI appearances were suggestive of an unusual mesenchymal tumour. CT scan, abdominopelvic and transvaginal ultrasound and intravenous urography (IVU) confirmed the well-circumscribed appearance of the mass and the marked medial deviation of the right ureter and bladder.

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Figure 1 Sagittal T2-weighted image, slightly to the right of the midline, showing the A–P extent of the tumour.

No abnormal circulation was seen on digital subtraction angiography (DSA) of the abdominal aorta.

At surgery, with a combined abdomino-perineal approach, an extraperitoneal pelvic tumour that extended around the major viscera was resected in its entirety. The tumour was encapsulated, attached to the right side of the bladder, urethra and vagina, highly vascular and measured 16 cm x 14 cm x 3 cm.

Cut section revealed a gelatinous soft and greyish coloured tumour with a fine capsule and no lobulation, necrosis or cystic degeneration.

Microscopic examination showed a relatively hypocellular tumour with myxoid stroma containing numerous vessels, some with thick walls. The cells showed regular nuclei and stellate fine cytoplasm. The features were of an AAM. No mitosis was noted but the tumour reached the margins in places (**Fig. 3**).

Immunohistochemical studies showed the cells to be positive for vimentine and focally positive for desmin and smooth muscle actin. There was strong nuclear positivity for oestrogen and progesterone.

The patient made an uneventful recovery, and went home 9 days later. At 19 months' follow-up, there was no evidence of recurrence either clinically or on MRI.

Discussion

The rarity of this condition makes the pre-operative diagnosis fairly difficult.^{1–20} Its radiological features have been described, being a displacing mass rather than infiltrating, with high signal intensity on T2-weighted images,^{1,14,15} but these features can be attributed to the

Figure 2 Coronal T2-weighted image with fat suppression showing a hyperintense mass inferior to a fibroid uterus and the urinary bladder compressed to the left side.

Figure 3 The histological view of the aggressive angiomyxoma (H&E stain).

myxoid component of the tumour, shown by many other soft tissue tumours of mesenchymal origin.

On gross examination, it was different in being well circumscribed non-lobulated and encapsulated,^{1–20} but the gelatinous appearance, greyish discoloration on cross section and the large size of the tumour are common features of AAMs. These findings are attributed to the myxoid stroma that is rich in collagen fibres.^{1–4} The diagnosis is based on histopathological and immunohistochemical studies. In this case, the finger-like projections into surrounding tissues was not seen,^{1–8} but the low cellularity in the myxoid stroma with numerous vessels of varying wall thickness along with the spindle-shaped and stellate-shaped cells with regular nuclei and absence of mitotic figures, added to the Immunohistochemical features, are all typical for AAM.^{1–8}

The documented studies for immunohistochemical data in two series showed nearly 97% positive response to desmin, 94% for smooth muscle actin, 93 and 90% for oestrogen and progesterone receptors, respectively, but 0% for S100 protein.¹

Given its propensity for local recurrence, it is very important to differentiate this tumour from other benign myxoid neoplasms, *e.g.* angiomyofibroblastoma, myxomas of intramuscular and juxta-articular types, myxoid leiomyoma, and pelvic fibromatosis. Equally important is to differentiate it from the low-grade myxoid sarcoma, *i.e.* myxoid liposarcoma, and low-grade myxoid malignant fibrous histiocytoma (*e.g.* myxofibrosarcoma).¹⁻⁴

This type of tumour may have a mesenchymal origin, but few papers reflect on its aetiology. The cytologic and architectural similarities of this tumour with the normal stroma of the vulvar, perineal and pelvic region add to the debate on the line of differentiation.¹⁻³

Clonal cytogenic aberrations have been reported in some aggressive angiomyxomas,¹ including loss of an X chromosome¹⁶ and a translocation involving 12q14-15.¹⁷ The latter has been frequently seen in a variety of other mesenchymal tumours and it seemed to be the most frequent chromosomal abnormality associated with irritation of human mesenchymal neoplasms.¹⁷

Using immunohistochemistry or ultrastructural analysis, it was proposed that the neoplastic cells of AAM exhibit fibroblastic and myofibroblastic features and appear to be hormonally influenced.^{1,18} The possibility that a progenitor cell has the capacity for smooth muscle differentiation has also raised.^{1,3,4} Other tumours with similar features (*i.e.* angiomyofibroblastoma) further suggest the above possibility as these two lesions seem related and possibly derived from a primitive mesenchymal cell normally found in the female genital tract that has the capability of myofibroblastic differentiation.^{1,19}

The unusual encapsulation of the well-circumscribed tumour in this case is also in favour of an isolated origin. In addition, sporadic reports of recurrences in sites different than the originally excised mass, and the reports of unusual locations or presentations of this tumour, *e.g.* multifocal AAM,²⁰ the bladder,¹² and the uterine cavity¹³ (*e.g.* polypoidal mass in the last two sites, occasionally), *etc.*, are all in favour of an isolated primitive mesenchymal cell origin.

The wide range of patients' age, and the occurrence of AAM in men, limit the possibility of hormonal dependency.^{4,18}

Complete excision is the treatment of choice.¹⁻²⁰ We believe that 1-2 yearly intervals of follow-up with alternate MRI and sonography is sufficient in view of the slow growing and non-metastasising nature of this tumour.

Conclusions

- Routine MRI or sonographic pelvic examination is recommended for any labial or ischial mass, to avoid inadequate preparation for unusual findings.
- Complete excision of the tumour is the treatment of choice.
- Follow-up with alternate MRI and sonographic examination at 1-2 yearly intervals should suffice.

- The well-circumscribed and encapsulated nature in our case, along with the occasional abnormal presentations of this tumour, favour the possibility of isolated primitive mesenchymal cell origin.

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