




Radio-pathological and Clinical Correlation of Aggressive Angiomyxoma: Experience of a Tertiary Care Cancer Hospital

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Abstract

Background Aggressive angiomyxoma (AAM) is a rare mesenchymal tumor predominantly affecting the pelvis and perineum of females. Due to its variable clinical presentation, it is often misinterpreted and treated as other pathologies like Bartholin gland cyst, vulvar abscess, and hernia. The tumor is locally infiltrative, making its complete surgical removal difficult.

Aim The aim of the study is to highlight and illustrate the imaging and pathological features of AAM.

Materials and Methods After obtaining clearance from the Institutional Ethics Committee, a retrospective study was conducted on patients with histopathological diagnosis of AAM, presenting to the dedicated sarcoma clinic between 2016 and 2019. The demographic, clinical, radiological, pathological, and treatment details were obtained from the institute's database. The magnetic resonance imaging (MRI) and computed tomography images were reviewed to assess different imaging parameters. Follow-up imaging was assessed for residual or recurrent mass in the pelvis or perineum and distant metastasis.

Results A total of four patients were included with an age range of 28 to 50 years. The baseline MRI of these four patients shows pelvic mass extending into the perineum, infiltrating along the surrounding fascial planes with the characteristic laminated appearance on T2-weighted image and gradual intense enhancement following contrast administration. All patients had residual disease post-surgery and were put on hormonal therapy.

Conclusion AAMs are locally aggressive, rarely metastasizing mesenchymal tumor that has a specific predilection for the perineum and pelvis of females. MRI features like laminated or striated appearance, post-contrast enhancement, and finger-like infiltrating projections should raise the suspicion of the diagnosis on baseline imaging.

Keywords

- ▶ aggressive angiomyxoma
- ▶ laminated appearance
- ▶ sarcoma

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Introduction

The term aggressive angiomyxoma (AAM) was coined by Steeper and Rosai in 1983¹ while describing this rare mesenchymal neoplasm which is locally aggressive and rarely metastasizes. AAM is a rare soft-tissue tumor that is thought to arise from mesenchyme²; however, the exact cell of origin is not clear and hence, it has been classified under the category of “tumor of uncertain differentiation” under the 2013 Edition of WHO Classification of Bone and Soft-tissue Tumors.³ Only limited literature is available in terms of anecdotal case reports and case series due to the rarity of the tumor.^{4,5} Predominantly affecting females and presenting with nonspecific complaints involving deeper tissues of the pelvis and perineum have been documented.^{5,6}

Due to its rarity and variable clinical presentation, it is often misinterpreted as other common pathologies like Bartholin gland cyst, vulvar abscess, hernia, and lipoma which often leads to inadvertent and incomplete surgical excision.⁷ As the name suggests, the tumor is aggressive indicating its infiltrative nature; AAM tends to flow along the fascial and surrounding fat planes, invading or displacing the tissues, viscera, and vessels which render their complete excision difficult and leads to residual and recurrent disease. The incomplete surgery can be attributed to the lack of prior suspicion, absence of preoperative diagnosis, and imaging depicting the extent of the disease. Recurrence rates range from 25 to 47% with approximately 85% recurring within 5 years of surgery.⁴

For timely diagnosis, it is important to have a high index of suspicion for AAM as the rate of misdiagnosis at initial presentation has been reported up to 82%.^{4,8} Multiple imaging modalities have been used to evaluate AAM including computed tomography (CT), magnetic resonance imaging (MRI), and ultrasonography. It has variable and nonspecific imaging appearance; however, a “laminated” or “swirling” pattern has been described on the T2-weighted sequence of MRI which is considered to be pathognomonic for AAM.⁶ Thus, MRI is considered as the imaging modality of choice by virtue of its better soft tissue resolution which enables it to provide information about the extent of the mass.

Histopathological examination shows a tumor with low to moderate cellularity with abundant myxoid stroma and numerous thick- and thin-walled vascular channels traversing the stroma. The tumor cells are spindle- and stellate-shaped with ill-defined cytoplasm intermingled with collagen fibers. Mitotic activity is usually not seen. The tumor usually invades adjacent soft tissues. Another common finding could be the presence of scattered cells with myxoid features usually zoned around entrapped nerves and vessels. Upon immunohistochemistry, there is usual immunoreactivity for estrogen receptor and progesterone receptor. Focal immunoreactivity for muscle markers can also be seen.⁹ The recent molecular analyses have identified translocation in the 12q13–15 region involving the *HMGGA2* gene.¹⁰

In this study, we aim to discuss the imaging features of histopathologically proven AAM in four patients who presented to our tertiary care cancer hospital.

Materials and Methods

This is a retrospective study that was conducted after obtaining the Institutional Ethics Committee approval; hence, individual patient consent was not obtained separately. Patients presenting to our dedicated sarcoma clinic from 2016 to 2019 with histopathological evidence of AAM were included in the study. The demographic, clinical, radiological, pathological, and treatment details were obtained from the case records available in the institute's database.

Imaging analysis: Baseline imaging was reviewed by our radiologist (E.D.) and the features were recorded on CT and MRI. The CT was studied for the size, extent, and margins of the mass, infiltration of the surrounding structures, vessels, density as compared with muscles, and enhancement pattern. Similarly, MRI was evaluated in terms of signal intensity as compared with muscles on T1 and T2-weighted sequences, enhancement pattern on contrast-enhanced sequences, diffusion-weighted images (DWIs, if available) in addition to the site, size, and extent of the mass. The presence of cystic changes and calcification were also looked for. Follow-up imaging was assessed for residual or recurrent mass in the pelvis or perineum and distant metastasis.

Histopathology: Based on the morphology and immunohistochemistry findings, the diagnosis of AAM was offered. In patients who underwent biopsy/surgery or treatment outside before presenting in our institute; the blocks were reviewed by a dedicated sarcoma pathologist (B.A.) and the features were recorded.

Treatment response: Details about surgery or medical treatment were obtained and the response was assessed on follow-up imaging as per mRECIST 1.1 criteria.

Follow up: Till the last visit in the dedicated sarcoma clinic, i.e., April 2020 was considered.

Results

We had four patients with proven AAMS during the study period of 3 years with a female to male ratio of 3:1 and age ranging from 28 to 50 years. The females presented with complaints of swelling and feeling of a lump in perineum (vagina, labia majora, or perianal location) and the male patient had scrotal swelling. The duration of complaints ranges from 2 to 8 years. The mean age of the patients at presentation was 40.2 years with a mean size of mass 10.7 cm and a range of 7.7 to 13.6 cm. The clinical and demographic details are highlighted in ►Table 1.

Imaging: Baseline MRI was available for all patients with T1 axial, T2 axial, and sagittal images, and postcontrast T1 fat-sat sequences in all. On T1-weighted images, the mass shows a hypointense signal intensity and on T2-weighted images, each patient had a mass with the characteristic “laminated” or “swirled” appearance (►Fig. 1) The mass shows gradual and progressive enhancement on postcontrast sequences with no cystic or calcific changes within. All patients showed laminated appearance on T2-weighted sequence, gradual progressive enhancement after contrast administration, and infiltrative pattern of growth with

Table 1 Clinical and demographic details of the four patients

S. no	Age/ Gender	Clinical presentation	Duration of symptoms (years)	Treatment received	Residual/recurrent disease	Status at last follow-up
1.	44/F	Perianal mass	6	Surgery f/b GnRH analogue	Residual mass stable on medical treatment	On second-line hormonal therapy (PR)
2.	28/F	Vaginal mass	3	Surgery f/b GnRH analogue	Residual stable mass	Mets to lung and axilla—put on hormonal therapy (PR)
3.	50/M	Scrotal swelling	2	Surgery f/b GnRH analogue	Stable residual mass	On hormonal therapy (SD)
4.	39/F	Mass along labia majora	8	Surgery f/b GnRH analogue	Residual stable mass	On hormonal therapy (PR)

Abbreviations: f/b, followed by; PR, partial response, PD, progressive disease; SD, stable disease as per mRECIST 1.1.

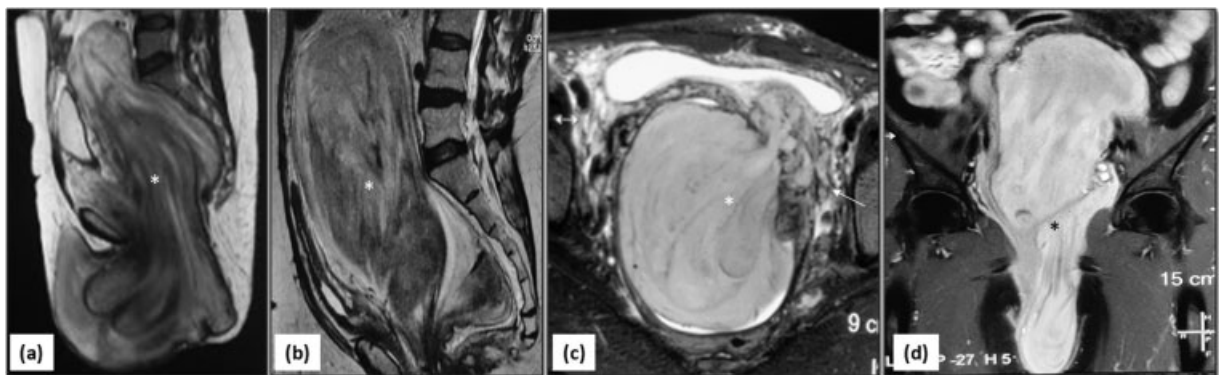


Fig. 1 Laminated appearance on MRI in the four patients of aggressive angiomyxoma: (a, b) T2-weighted sagittal image in patients 1 and 3, respectively, shows the striated layered appearance (*asterisk*) as alternating high and low signal bands. (c) The axial T2 weighted image of patient 2 also shows the laminated appearance (*asterisk*) in the mass with finger-like projections along its left lateral border (*arrows*). (d) Significant enhancement is seen in the postcontrast coronal image of patient 4 with the laminated morphological feature within the mass (*asterisk*).

finger-like projections along the periphery. DWI images were available for one patient where the mass showed diffusion restriction and a mean ADC value of $1.6 \times 10^{-3} \text{ mm}^2/\text{s}$ (**Fig. 2**) **Table 2** describes imaging features on MRI in detail.

Follow-up CTs of the chest and abdomen are available for one patient (patient 2) which showed a residual mass in pelvis demonstrating progressive heterogeneous enhancement on single venous phase contrast-enhanced CT. This patient had nodules in the lungs and enlarged necrotic axillary lymph nodes suspicious for metastasis. The primary residual mass showed infiltration of the urinary bladder and finger-like projections along its margins.

Pathology (**Fig. 3**): Histological examination in all the tumors shows a low to moderate cellularity with abundant myxoid stroma. The tumor cells were spindle- and stellate-shaped with ill-defined cytoplasm. The cells exhibit mild nuclear pleomorphism with round hyperchromatic nuclei with few cells showing small nuclei. Mitotic activity was infrequent. The margins were infiltrative with the encasement of normal nerves and vessels at places. Immunohistochemistry of the estrogen and progesterone receptors was

diffusely positive in all the cases. Focal immunoreactivity for smooth muscle actin and desmin was noted in cells showing myxoid differentiation of light microscopy. CD34 was focal in one of the cases. However, S100 was negative. One of the patients showed moderate cellularity with the cells exhibiting oval to spindle morphology and mild nuclear pleomorphism. However, there was no mitotic activity and no significant atypia. The remaining cases were mildly cellular with traversing blood vessels and abundant myxoid stroma with monomorphic spindle to stellate-shaped cells.

Treatment and follow-up: Patients underwent incomplete surgery followed by adjuvant hormonal therapy (GnRH analogues). Only one patient progressed on first-line hormonal therapy and later responded to second-line hormonal therapy; the remaining two patients showed partial response and one patient had stable disease on first-line hormonal therapy only (**Fig. 4**). One patient developed lesions in the lungs and axilla (within 6 months of diagnosis of AAM) which were evaluated for metastases. The axillary lymph node biopsy did not yield any viable tissue and the patient refused for a repeat biopsy; hence, she was put on Goserelin (GnRH analogue). Since the axillary lymph nodes

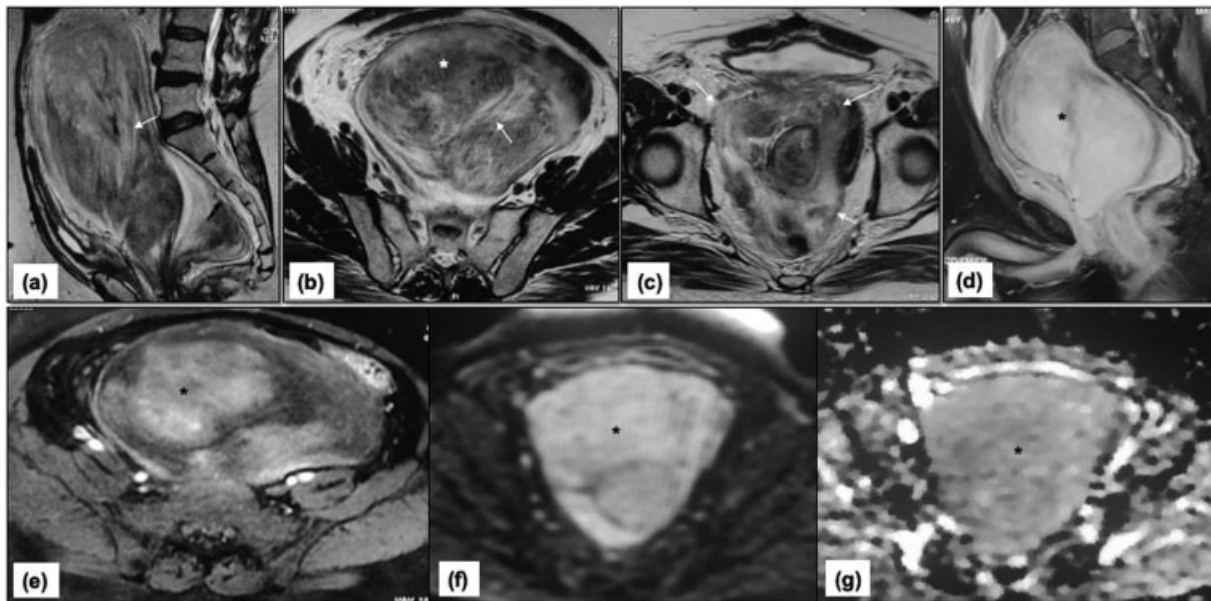


Fig. 2 MRI features of AAM: the pelvic mass shows a predominant hypointense signal on T2-weighted image (a) with an internal laminated or striated appearance (arrows in a and b) seen as alternate high and low signal bands. Its infiltrative nature is seen as finger-like projections along the ill-defined borders extending into the surrounding planes (arrows in c). (d, e) The mass shows gradual intensity enhancement after the administration of contrast (asterisk) and restriction on DWI and ADC images (f, g). AAM, aggressive angiomyxoma; ADC, apparent diffusion coefficient; DWI, diffusion-weighted image.

Table 2 Imaging features of aggressive angiomyxomas on MRI

Patient	Size at baseline	Extent	T1	T2	Enhancement on CEMR	Cystic changes	Calc	Finger-like projections	Laminated appearance
1	45 × 25 cm	Rectovesical location with extension to perineum	Hypo	Hyper	+	–	–	+	+
2	8.6 × 7.2 cm	Retrovesical location with extension to lateral pelvic walls and vagina	Hypo	Hyper	+	–	–	+	+
3	12.8 × 10.4 × 9.6 cm	Scrotal lesion with extension to inguinal canal	Hypo	Hyper	+	–	–	+	+
4	35 × 25 × 10 cm	Rectouterine lesion with presacral extension	Hypo	Hyper	+	–	–	+	+

decreased by 40% and lung lesions were stable, the lesions were considered metastatic, and the response was categorized as a partial response.

All patients are alive till the last clinic visit with a duration of follow-up ranging from 13 to 90 months.

Discussion

Angiomyxoma is usually a benign tumor that displaces neighboring organs rather than invading them. It is more common in women of reproductive age and mostly develops

in the pelvis, vulva, or perineum in the fourth to fifth decade.¹ In men, it may develop in the perineum or inguinoscrotal region.⁵ The incidence in females is significantly higher than in males (female-to-male ratio of 6:1)⁶ which was observed in our study as well (a female to male ratio of 3:1).

Most of the AAM patients present with a slow-growing mass in the pelvic and perianal regions that is either asymptomatic or associated with regional pain, dyspareunia, or a pressure-like sensation. Clinical diagnosis before surgery is difficult since the differential diagnosis on clinical

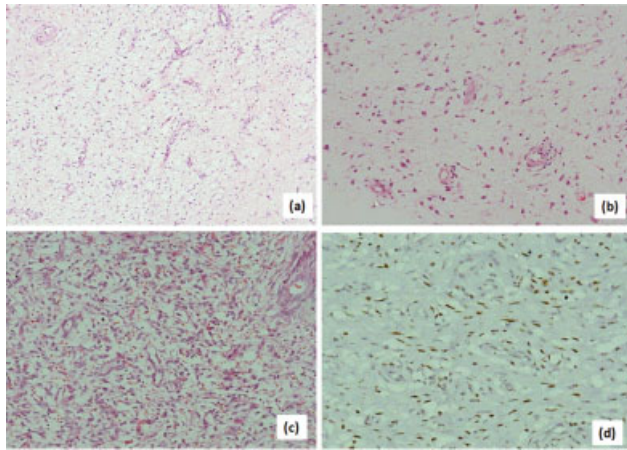


Fig. 3 Histopathological findings in aggressive angiomyxoma: (a) Low power picture from a case of aggressive angiomyxoma showing relatively low cellularity with abundant myxoid matrix and many traversing thin-walled blood vessels. (H&E 100x). (b) On high power, the paucicellular tumor shows monomorphic spindle-to stellate-shaped cells embedded in myxoid stroma. The nuclei are small with scanty cytoplasm and indistinct cytoplasmic boundaries (H&E 200x). (c) Another case of aggressive angiomyxoma showing moderate cellularity with cells exhibiting mild nuclear pleomorphism and oval-to spindle-shaped nuclei and scanty to moderate amount of cytoplasm (H&E 200x) and (d) Immunostain for estrogen receptor showing nuclear immunoreactivity in spindle cells.

examination includes lesions like vulval abscess, Bartholin gland cyst, cyst of mesonephric duct, vaginal hernia, or levatorani muscle hernia. This often leads to misdiagnosis and incomplete surgical excision. The female patients in our study presented with initial symptoms of nontender mass in perianal, vaginal, and labia majora and the male patient had swelling in the inguinoscrotal region. All these patients underwent excision of the mass before presenting to our institute with residual/recurrent mass. It emphasizes the role of preoperative imaging in determining the exact size and extent of the tumor.

A review of the radiology literature revealed limited information on the imaging features of AAMs. In a series of 10 patients, Outwater et al¹¹ found the typical laminated tumor morphology on T2-weighted imaging and contrast-enhanced MRI. In other case reports, the typical MRI findings of laminated T2 signal intensity characteristics have been described.^{12,13} Our study elaborates on previously published literature by describing the CT and MRI characteristics of these tumors and specifically assessing T2-weighted imaging and contrast-enhanced CT in the detection of internal laminated appearance. The ultrasonography (US) and CT features of AAM have been described.¹⁴ Upon US, the mass may appear hypo-echoic or cystic. The CT features are variable and include hypoattenuating or isoattenuating mass involving the pelvis and perineum with enhancement in the contrast-enhanced scan, which may show the classical swirled appearance. In the series of six cases reported by Outwater et al,¹¹ only 50% of the cases showed the classical swirling architecture on CT.

The imaging differential diagnosis includes myxoma, infiltrating angioliipoma, and infiltrating liposarcoma. Myxoma

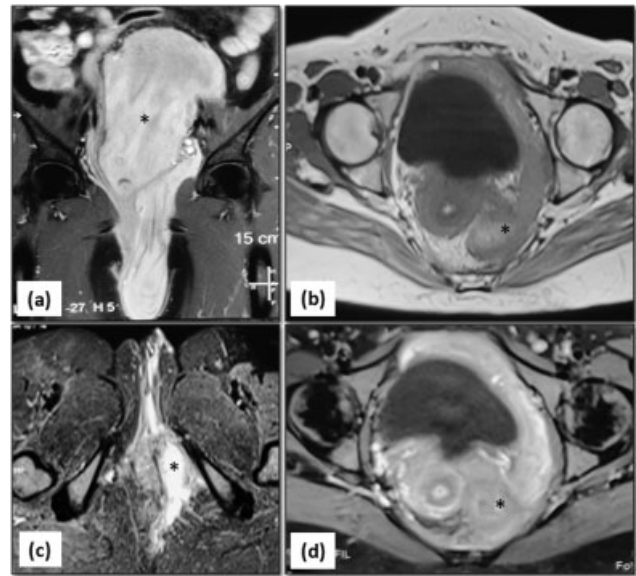


Fig. 4 Follow-up and response assessment: (a) Baseline MRI shows large enhancing mass in the pelvis extending to the perineum into left labia majora (asterisk). (b, c) Postoperative MRI shows residual mass (asterisk) along the left side of the pelvis extending along the labia majora (c) which remains stable on hormonal therapy as seen on MRI after 6 months of initiating the medical treatment (d).

is a benign mesenchymal neoplasm that most frequently occurs in older patients. However, myxomas are mainly intramuscular and show no significant vascularity.¹⁵ Infiltrating angioliipoma is usually found in the thigh region and as the name implies it infiltrates and replaces the involved muscle and subcutaneous tissue.¹⁶ The lack of high fat content is a key feature in differentiating the angiomyxoma from myxoid liposarcoma and infiltrating angioliipoma.¹⁷

Due to the rarity of this disease, treatment recommendations are based on case reports and small series. A wide surgical resection is the treatment of choice, sometimes after preoperative angiographic embolization to decrease the size of the tumor.¹⁸ But, because of its frequently large size and finger-like extensions along the organs (urogenital tract and rectum), wide excision is not always possible and/or may cause significant morbidity in young patients. In this series, upon initial imaging evaluation, the tumors appeared bulky with a mean long-axis dimension of 10.7 cm (range, 7–14 cm) with 50% of cases showing a maximal diameter of more than 10 cm. Furthermore, the prognostic value of extensive surgical resection to obtain clear margins is questioned. A review of over 100 cases by Chan et al¹⁹ showed that patients with clear resection margins have similar chances of remaining disease free (50% at 10 years) compared with those having tumor-involved resection margins (40%). Later publications confirmed these findings.^{5,18} These observations explain that incomplete resection is acceptable nowadays if followed by long-term follow-up and careful monitoring for timely identification and prompt resection of relapse.

Treatment involving chemotherapy and radiotherapy has been disappointing, probably due to the low mitotic activity of the tumors.^{18–20} AA is likely hormone dependent as

suggested by the reproductive age of the patients, the few cases reported during pregnancy, and the frequent expression of hormone receptors.⁵ Thus, gonadotropin-releasing hormone agonists have been administered to AA patients in the postoperative and in the preoperative setting with dramatic responses, facilitating surgery, or avoiding mutilating surgery.^{21–23}

After treatment, long-term follow-up is required as late recurrences have been described up to 14 years following treatment¹⁹ and around 71% of recurrences occur in the first 3 years regardless of the initial tumor size and age of the patient.¹⁹ In our series, all the patients had residual disease on follow-up. Distant metastasis from AAM is extremely rare, and currently, there are three cases reported around the world.^{24–26} The first case is a 63-year-old woman with lung and lymph node metastasis. The second case is a 27-year-old woman with multiple postoperative recurrences, who was diagnosed with lung metastasis after a few years and died from her disease. The third case is a recurrent AAM; the tumors invaded into the inferior vena cava and right atrium and also into both the lungs. These three cases had lung metastases.²⁶ In our experience, we had one patient who had suspicious metastases to lung and axillary lymph nodes. The patient had a partial response to hormonal therapy till the last follow-up. Because of this unpredictable biological behavior of AAM, some groups recommended classifying AAM as moderate malignant tumors.

Periodic clinical examination may be insufficient and imaging studies, such as MRI, are fundamental.² They may detect recurrence early, when the tumor is smaller in size, making surgery easier to perform or allowing the use of less morbid percutaneous ablative therapeutic strategies such as radiofrequency ablation or cryotherapy.²

Our study is limited by its retrospective study design and the small number of patients from a single cancer center, owing to the rarity of the neoplasm.

Conclusion

AAMs are locally aggressive, mesenchymal tumors that have specific predilections for the perineum and pelvis of females in the fourth decade of life. The angiomyxomas typically show local recurrence and are extremely rare to metastasize. The MR appearances of AAMs are characteristic, with the “laminated” appearance on the T2 weighted sequence, being diagnostic for pelvic AAMs. While surgery remains the mainstay of treatment, adjuvant treatment in the form of hormonal/chemotherapy is routinely prescribed due to the higher chances of local recurrences and long-term follow-up is also required.

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Conflict of Interest
None declared.

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