

CASE REPORT

DOI: 10.5336/caserep.2021-87801

Aggressive Angiomyxoma of the Pelvis and Perineum: A Case Report with Radiologic-Pathologic Correlation

Gökhan YILMAZ^a, Mehmet Haydar ATALAR^b, Nisa BAŞPINAR^b, Neşe YELDİR^c,
Ali Cihan YILDIRIR^d

^aClinic of Radiology, Başakşehir Çam and Sakura City Hospital, İstanbul, Türkiye

^bDepartment of Radiology, Sivas Cumhuriyet University Faculty of Medicine, Sivas, Türkiye

^cDepartment of Pathology, Sivas Cumhuriyet University Faculty of Medicine, Sivas, Türkiye

^dDepartment of General Surgery, Sivas Cumhuriyet University Faculty of Medicine, Sivas, Türkiye

ABSTRACT Aggressive angiomyxoma is a rare mesenchymal tumor mostly observed in women of childbearing age. Although it is often located in the vulva, it can also be observed in the paravaginal, perirectal areas, perineum and pelvis. It appears as a locally aggressive, large, infiltrative mass. Complete surgical excision is often difficult and has high recurrence rates. Magnetic resonance imaging (MRI) plays an important role in preoperative diagnosis of aggressive angiomyxoma. The typical swirling appearance on T2-weighted and post-contrast T1-weighted MRI series is diagnostic for aggressive angiomyxoma. Here, we present a 51-year-old female patient with pelvic-perineal aggressive angiomyxoma, accompanied by radiological and pathological findings.

Keywords: Magnetic resonance imaging; myxoma; pathology; pelvis; perineum

Aggressive angiomyxoma is a rare mesenchymal tumor. Aggressive angiomyxoma specially affects the vulvovaginal region in females of a reproductive period. The mass is termed aggressive because slow growing with a high rate of local recurrence. Clinical diagnosis could also be difficult; but magnetic resonance imaging (MRI) often shows the swirled strands of this neoplasm. The “swirled appearance” is a diagnostic finding of aggressive angiomyxoma.¹⁻⁴ In present article, we report the MRI and histopathological findings of a 51-year-old woman who diagnosed aggressive angiomyxoma.

CASE REPORT

A 51-year-old female patient presented with a palpable mass in the perineal region. There was painless swelling in the perineum. The lesion was considered as a perineal hernia at the first examination. There was no other pathology in pelvic examination and ultrasonographic evaluation of the patient. Tumor

markers were normal limits. Other laboratory tests were also normal. Pelvic MRI was taken to the patient. MRI showed a large, well-circumscribed mass lesion in the left pelvic and perineal regions. The mass was isointense to muscle on T1-weighted (T1W) and hyperintense on T2-weighted (T2W) images. Even, T2W, and postcontrast T1W images had a “swirled appearance”. The mass was extending into the perirectal area. The lesion had infiltrative extensions to the perineal fat planes in the inferior part. On diffusion-weighted imaging (DWI), the mass showed high signal intensity on apparent diffusion coefficient (ADC) series. MRI features suggested aggressive angiomyxoma (Figure 1A-D). The patient was operated on with these findings. The patient was discharged without any postoperative problems.

On gross examination of the mass, tumor was soft-elastic, gray-yellow, irregularly, unencapsulated mass. The cut surface was gray-white, homogenous, glistening with mild bleeding areas. Histopathologic

Correspondence: Mehmet Haydar ATALAR

Department of Radiology, Sivas Cumhuriyet University Faculty of Medicine, Sivas, Türkiye

E-mail: mhatar@gmail.com

Peer review under responsibility of Türkiye Klinikleri Journal of Case Reports.

Received: 17 Feb 2022

Received in revised form: 24 Jun 2022

Accepted: 15 Jul 2022

Available online: 11 Aug 2022

2147-9291 / Copyright © 2022 by Türkiye Klinikleri. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).



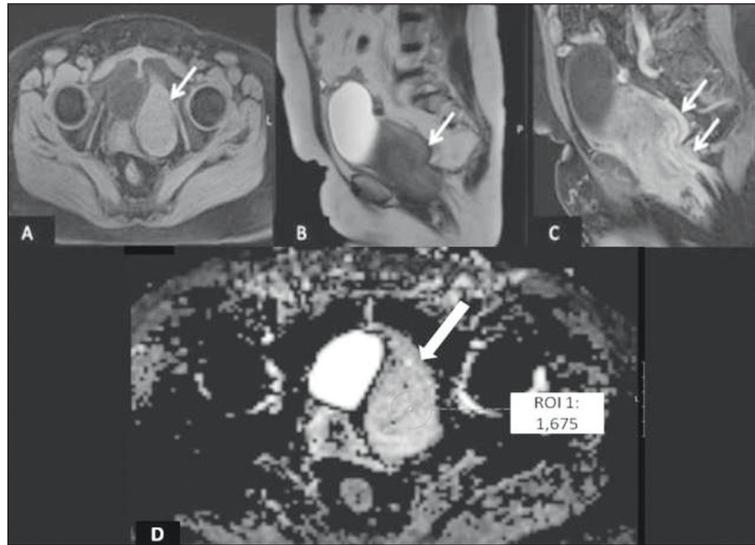


FIGURE 1: A) Axial T1-weighted magnetic resonance image demonstrates an isointense mass lesion with adjacent soft tissues in the left pelvic region. B) Sagittal T2-weighted magnetic resonance image shows a left perineal mass extending across the pelvic diaphragm, compressing the uterus, and a “swirled appearance” within the mass. C) Sagittal postcontrast fat-suppressed T1-weighted image shows the characteristic “swirled appearance”. D) On axial apparent diffusion coefficient image ($b=800 \text{ mm}^2/\text{s}$), the mass shows a high signal intensity (apparent diffusion coefficient value: $1.67 \times 10^{-3} \text{ mm}^2/\text{s}$).

sections showed an unencapsulated tumor comprised of sparsely populated benign spindle shaped cells in a myxoid background in most of the fields. The tumor cells were cytologically bland and had a spindled, ovoid, or stellate appearance with ill-defined cytoplasmic borders. Tumor was composed variable size from thin or thick walled vascular channels in a myxoid stroma. In addition, it contained foci of hemorrhage. The tumor was infiltrating adipose tissue. Any pleomorphism, hyperchromasia, increased mitosis and necrosis could not be detected in the lesion. In immunohistochemical examination, the spindle cells were positive for estrogen receptor (strongly diffuse, >50%), progesterone receptor, smooth muscle actin, vimentin, CD34 and desmin (focally), and negative for DOG1, S100, CD117. Ki67 index was 1-2%. In the light of immunohistochemical findings, typical histomorphological features were compatible with aggressive angiomyxoma (Figure 2, Figure 3, Figure 4). However, local recurrence occurred during the six-month follow-up period after the initial excision. Written informed consent was obtained from the patient for publication of this report.

DISCUSSION

In aggressive angiomyxoma, the deep soft tissues of pelvis and perineum in women are frequently af-

ected. Aggressive angiomyxoma is generally seen in the form of large masses. When they diagnosed, they are larger than 10 cm. These tumors present either as a painless mass or as a mass causing local pressure effect.¹ Aggressive angiomyxomas are not malignant neoplasms; but they can show locally aggressive features. This tumors often tend to infiltrate contiguous tissues.² Often, the deep pelvic component of aggressive angiomyxoma relative to the pelvic diaphragm is not evaluated clinically.¹ The clinical findings can sometimes be confused with the benign diseases such as vaginal cysts, pelvic lipoma, pelvic hernia or abscess. For these reasons, preoperative diagnosis is usually not performed and the tumor is often misdiagnosed during the presentation.³

The computed tomography (CT) features are not typical. On unenhanced CT, there are hypodense or isodense mass in the pelvis and perineum. These tumors may rarely show the typical “swirled appearance” on postcontrast CT.⁴ Outwater et al. reported that only three of six patients have the classical “swirled appearance” on CT.⁵ Surabhi et al. reported that only six of 14 patients have the classical “swirled appearance” on CT.¹ MRI is more valuable due to the soft tissue resolution and “swirled appearance” in the diagnosis of aggressive angiomyxoma.³ The masses are usually isointense to muscle and relatively

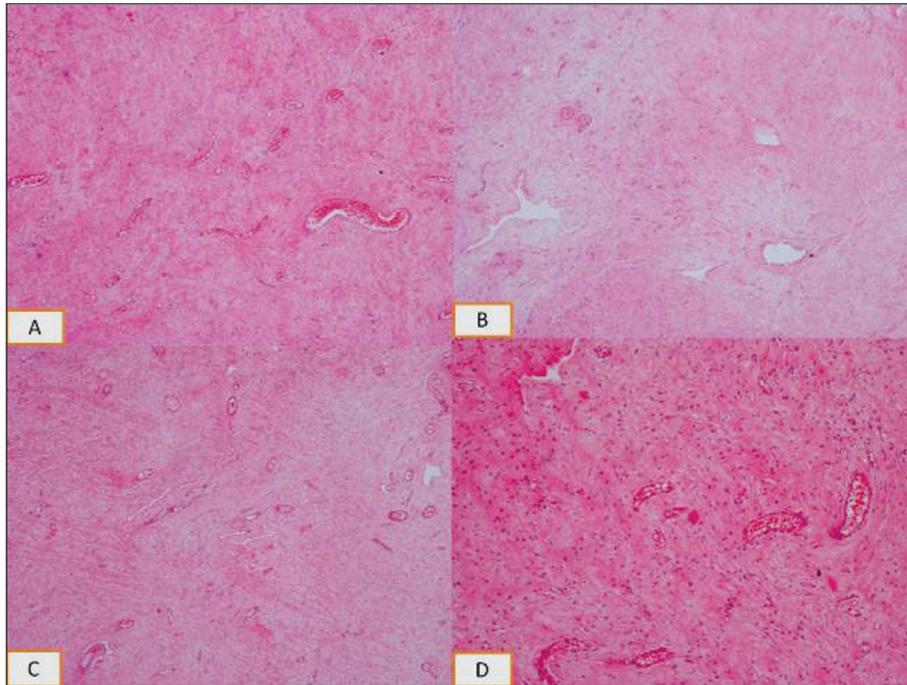


FIGURE 2: This tumor is composed of spindle cells and medium-size to large vessels within an abundant myxoid matrix in the H&E (x100) staining sections.

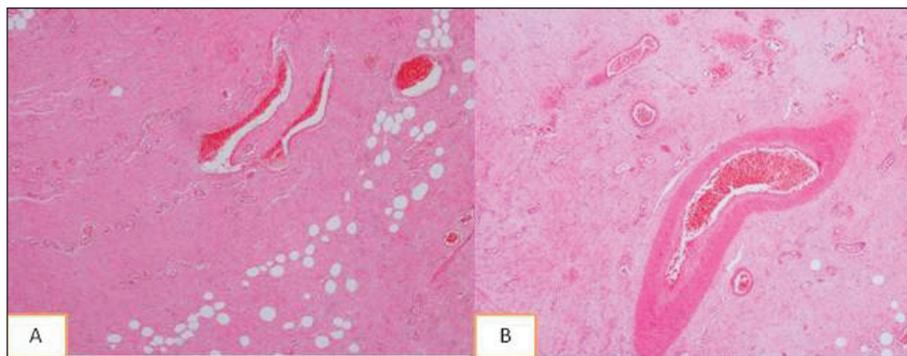


FIGURE 3: The vessel walls often hyalinized and contain perivascular concentric collagen fibres in the H&E (x100) staining sections.

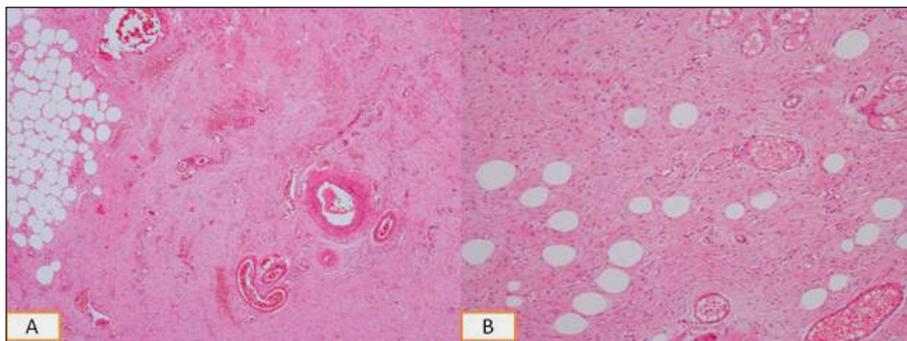


FIGURE 4: The tumor has infiltrative margins and non-neoplastic soft tissue in some microscopic areas in the H&E (x100) staining sections.

homogeneous on T1W images and hyperintense on T2W images, as in our cases.⁴ The hyperintense signal on T2W images is due to the myxomatous and

high water content of tumor.⁶ The “swirled appearance” is a diagnostic imaging finding of these tumors.⁵ The characteristic “swirled appearance” is

seen on T2W and post-contrast T1W images, as in our case.⁷ In aggressive angiomyxoma, an information about DWI is extremely low. In a recent study, high ADC values were determined in aggressive angiomyxomas, as in our case.⁸ Aggressive angiomyxoma characteristically do not have infiltrative features and tend to displace and grow around contiguous tissues.³

The imaging differential diagnosis of aggressive angiomyxoma should include myxoma, infiltrating angiolipoma, and infiltrating liposarcoma.⁷ Myxoma is a benign mesenchymal neoplasm. It mostly occurs in older patients. Myxoma is usually located in intramuscular area. It does not indicate marked vascularity.⁹ Infiltrating angiolipoma is hypervascular lesion similar to aggressive angiomyxoma. This tumor is usually found in the thigh area. It infiltrate muscle and subcutaneous tissue.¹⁰ The lack of high-fat content is an important feature in distinguishing of aggressive angiomyxoma.⁹ Infiltrative liposarcomas are deeply located and show infiltrative growth pattern. MRI findings varies depending on the degree and amount of adipose tissue.^{9,10}

The clinical differential diagnosis should be included vulvar masses, vaginal cysts, perineal lipoma, canal of Nuck hernia, hernias of pelvic origin, and pelvic organ prolapses in female patients.^{9,11} In particular, it should be kept in mind that it can easily be mixed with non-reducible pelvic floor hernias.

The main pathological differential diagnosis of aggressive angiomyxoma contains myxoma, various myxoid sarcomas and angiofibroblastoma. Myxoma does not have a vascular pattern of aggressive angiomyxoma. It does not usually occur in areas affected by aggressive angiomyxoma. Myxoid sarcomas generally have more cellularity and cytological atypia than aggressive angiomyxomas. Angiofibroblastoma occurs in regions similar to aggressive angiomyxoma, but is usually positioned more superficially.¹²

Surgical excision is the main treatment method. However, anatomical localization of the tumor makes it difficult to en bloc resection if the uterus is required to be preserved in reproductive age patients.⁸ Hormonal therapy can be reduced the tumor size in large tumors in the preoperative period. In addition, hormonal therapy can be used in postoperative recurrences.¹

In conclusion, aggressive angiomyxoma has a characteristic MRI appearance, such as a “swirled appearance”. MRI also guides the convenient surgical approach. Although the lesion is benign, it should be considered that the lesion may have recurrence.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Gökhan Yılmaz, Mehmet Haydar Atalar; **Design:** Gökhan Yılmaz, Mehmet Haydar Atalar, Nisa Başpınar; **Control/Supervision:** Gökhan Yılmaz, Mehmet Haydar Atalar, Neşe Yeldir; **Data Collection and/or Processing:** Gökhan Yılmaz, Mehmet Haydar Atalar, Neşe Yeldir; **Analysis and/or Interpretation:** Gökhan Yılmaz, Mehmet Haydar Atalar, Nisa Başpınar, Neşe Yeldir, Ali Cihan Yıldırım; **Literature Review:** Gökhan Yılmaz, Mehmet Haydar Atalar, Nisa Başpınar, Neşe Yeldir; **Writing the Article:** Gökhan Yılmaz, Mehmet Haydar Atalar, Neşe Yeldir; **Critical Review:** Ali Cihan Yıldırım, Neşe Yeldir; **References and Fundings:** Gökhan Yılmaz, Mehmet Haydar Atalar, Nisa Başpınar, Neşe Yeldir, Ali Cihan Yıldırım; **Materials:** Gökhan Yılmaz, Mehmet Haydar Atalar, Nisa Başpınar, Neşe Yeldir, Ali Cihan Yıldırım.

REFERENCES

1. Surabhi VR, Garg N, Frumovitz M, Bhosale P, Prasad SR, Meis JM. Aggressive angiomyxomas: a comprehensive imaging review with clinical and histopathologic correlation. *AJR Am J Roentgenol.* 2014;202(6):1171-8. [[Crossref](#)] [[PubMed](#)]
2. Srinivasan S, Krishnan V, Ali SZ, Chidambaranathan N. "Swirl sign" of aggressive angiomyxoma-a lesser known diagnostic sign. *Clin Imaging.* 2014;38(5):751-4. [[Crossref](#)] [[PubMed](#)]
3. Jeyadevan NN, Sohaib SA, Thomas JM, Jeyarajah A, Shepherd JH, Fisher C. Imaging features of aggressive angiomyxoma. *Clin Radiol.* 2003;58(2):157-62. [[Crossref](#)] [[PubMed](#)]
4. STATdx [Internet]. © 2022 Elsevier [Cited: July 2015]. Available from: [[Link](#)]
5. Outwater EK, Marchetto BE, Wagner BJ, Siegelman ES. Aggressive angiomyxoma: findings on CT and MR imaging. *AJR Am J Roentgenol.* 1999;172(2):435-8. [[Crossref](#)] [[PubMed](#)]
6. Sinha R, Verma R. Case 106: aggressive angiomyxoma. *Radiology.* 2007;242(2):625-7. [[Crossref](#)] [[PubMed](#)]
7. Tariq R, Hasnain S, Siddiqui MT, Ahmed R. Aggressive angiomyxoma: swirled configuration on ultrasound and MR imaging. *J Pak Med Assoc.* 2014;64(3):345-8.
8. Brunelle S, Bertucci F, Chetaille B, Lelong B, Piana G, Sarran A. Aggressive angiomyxoma with diffusion-weighted magnetic resonance imaging and dynamic contrast enhancement: a case report and review of the literature. *Case Rep Oncol.* 2013;6(2):373-81. [[Crossref](#)] [[PubMed](#)] [[PMC](#)]
9. Tappouni RF, Sarwani NI, Tice JG, Chamarthi S. Imaging of unusual perineal masses. *AJR Am J Roentgenol.* 2011;196(4):W412-20. [[Crossref](#)] [[PubMed](#)]
10. Stewart ST, McCarthy SM. Case 77: aggressive angiomyxoma. *Radiology.* 2004;233(3):697-700. [[Crossref](#)] [[PubMed](#)]
11. Elkattah R, Sarkodie O, Otteno H, Fletcher A. Aggressive angiomyxoma of the vulva: a précis for primary care providers. *Case Rep Obstet Gynecol.* 2013;2013:183725. [[Crossref](#)] [[PubMed](#)] [[PMC](#)]
12. Weiss LM. Soft tissues. In: Weidner N, Cote RJ, Suster S, Weiss LM, eds. *Modern Surgical Pathology.* 2nd ed. London: WB Saunders; 2009. p.1717-83. [[Crossref](#)]