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Case Report

"Look Both Ways When You Cross the Street" – A Reporting Sine Qua Non

Vishesh Dhawan[1], Shiv Pankaj Khanna[2], Ayushi Kediya[3*]

[1]Senior Resident, MD, Department of Pathology, Maharishi Markandeshwar Institute of Medical Sciences, Maharishi Markandeshwar (Deemed to be University), Mullana, Ambala -133207, Haryana, India
[2]Professor, MD, Department of Pathology, Maharishi Markandeshwar Institute of Medical Sciences, Maharishi Markandeshwar (Deemed to be University), Mullana, Ambala -133207, Haryana, India
[3]Senior Resident, MD, Department of Pathology, Maharishi Markandeshwar Institute of Medical Sciences, Maharishi Markandeshwar (Deemed to be University), Mullana, Ambala -133207, Haryana, India

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***Corresponding author:** Ayushi Kediya
Senior Resident, MD, Department of Pathology, Maharishi Markandeshwar Institute of Medical Sciences, Maharishi Markandeshwar (Deemed to be University), Mullana, Ambala -133207, Haryana, India

Abstract

We discuss an interesting case of a 67 years old female patient with an abdominal lump leading to a reporting dilemma among female genital tract lesions in which even though clinical acumen, radiological findings and gross examination of the resected specimen pointed towards a certain malignant provisional diagnosis, actually turned out to be a rare benign entity of Aggressive Angiomyxoma when seen under the microscope and proved on immunohistochemistry, pressing upon the importance of correlation of all the findings (clinical, radiological, gross and microscopy) while making a diagnosis. We also press upon the gravity of proper communication between the reporting pathologist and treating physician for the best possible patient management.

Keywords: Aggressive Angiomyxoma, Histopathology, Locally Aggressive.

INTRODUCTION

Since time indefinite, young pathologists have always drawn a straight line between benign and malignant neoplasms when it comes to histopathological specimens, former having smooth contours with non-infiltrating borders and absence of necrosis in comparison to what is seen in later. However this should never be a thumb rule. Certain benign neoplasms of lower female genital tract (FGT) such as aggressive angiomyxoma (AAM) can result in conundrum amongst the reporting pathologists. We discuss one such case in which what appeared as a malignant, infiltrating tumor of FGT both clinically and radiologically, even on gross appearance, turned out to be a locally aggressive benign neoplasm.

CASE REPORT

A 67 years old female presented to surgery OPD with complaints of lump over lower abdomen for 4 months associated with pain, on and off burning micturition and constipation. Her menstrual history comprised of irregular menstrual cycle. On per abdominal examination, a soft, non-distended, non-tender, hard mass was palpable over hypogastrium with audible bowel sounds. Contrast enhanced computed topography showed an ill-defined soft tissue lesion measuring 15x13 m seen in middle of pelvis deviated toward left side. This was followed by abdominal hysterectomy showing a growth over posterior wall of uterus involving left fallopian tube and ovary following which the resected specimen was sent for histopathological examination.

We received an already cut open specimen of hysterectomy with left salpingo-oophorectomy. Uterus with cervix measured 10x9x8 cm with hypertrophy of cervical lips and cervical canal measuring 2.8 cm in length. An exophytic friable growth (Fig 1), measuring 10x8.5x6.5 cm, was identified at the posterior vaginal cuff, extending medially towards the ectocervix, reaching at 3 and 6 o'clock cervical position, enclosing but not involving the cervical canal. Grossly the growth was seen involving isthmus, lower uterine segment and paracervical tissue. Cut surface of growth was greyish white and showed extensive areas of hemorrhage. The

endo-myometrium was unremarkable and measured 0.2+3.3 cm respectively. The attached left adnexa were unremarkable grossly.

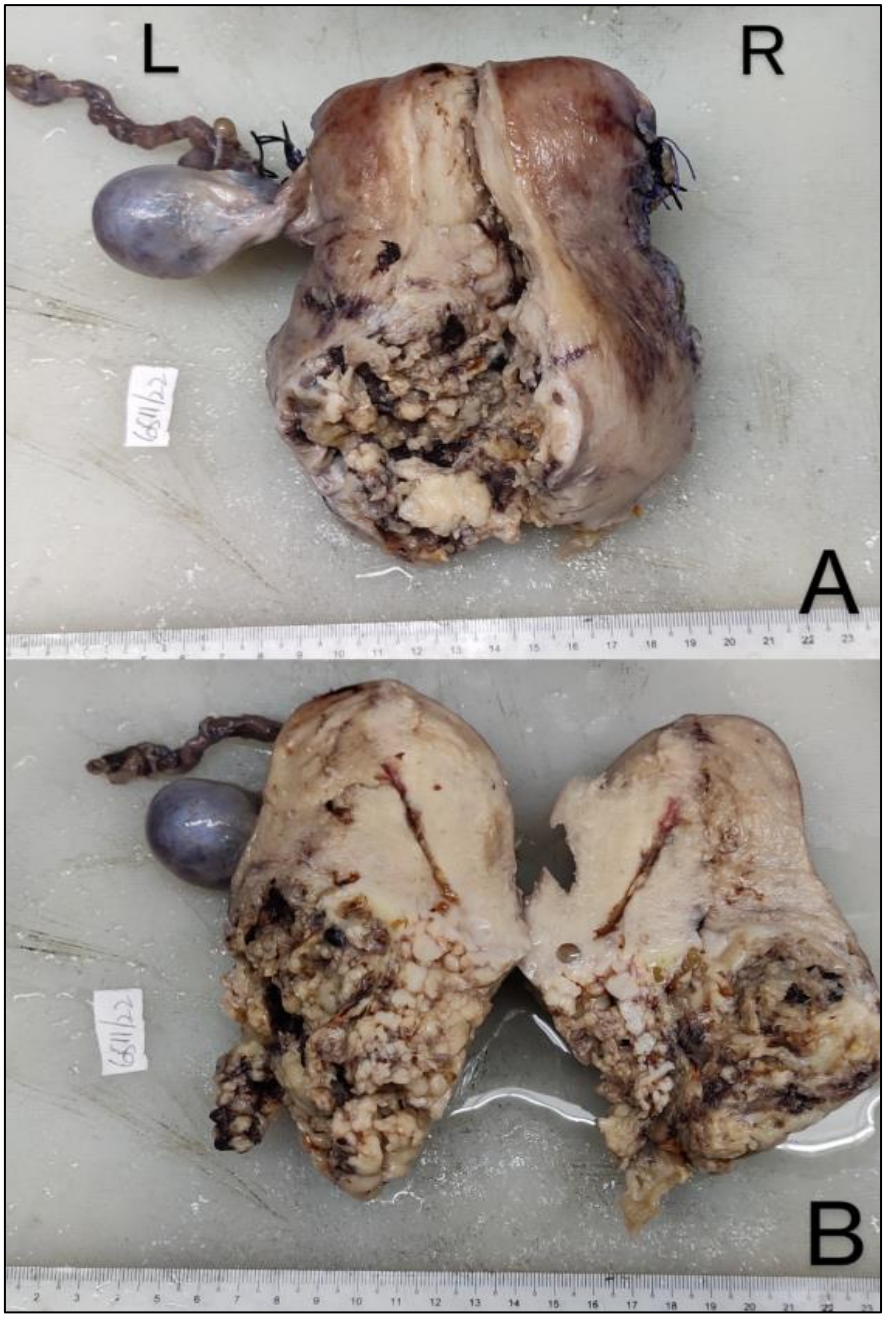

Fig 1: Gross photographs (A & B) show exophytic friable growth in the posterior vaginal area involving isthmus, lower uterine segment and paracervical tissue, reaching at 3 and 6 o'clock cervical position, enclosing but not involving the cervical canal

Histopathological sections (Fig 2) from growth showed uniform, short, spindle shaped cells in an edematous to fibrous stroma, containing short bundles of delicate collagen fibres and numerous medium sized thick walled blood vessels. These cells had short, oval to fusiform nuclei, inconspicuous nucleoli, scant eosinophilic cytoplasm and ill-defined borders. Occasional mitotic figures were seen with stroma consisting of wispy collagen with foci of dense eosinophilic collagen fibres. The tumor also showed stromal edema, hyalinization, perivascular lymphoid aggregates and myxoid change at places. No necrosis was seen in any section.

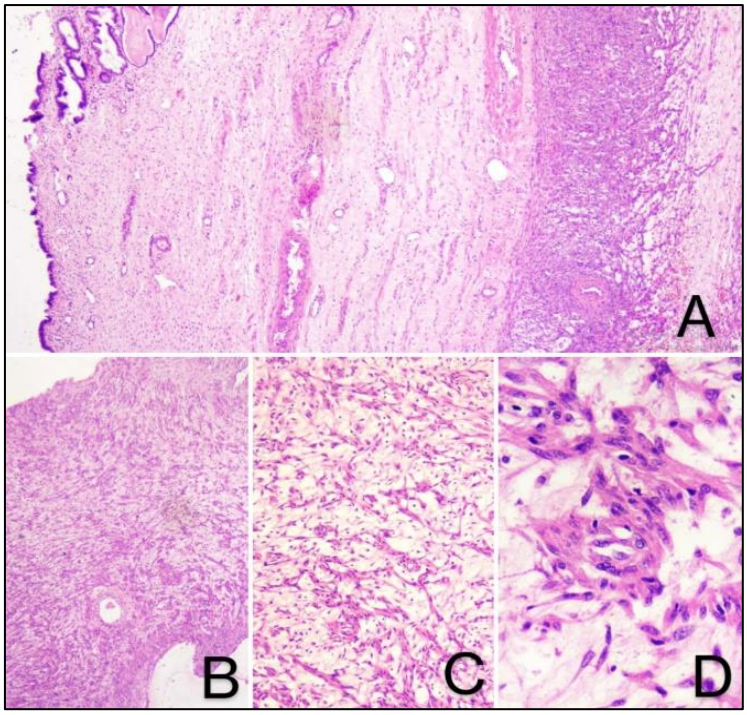

Fig 2: Histopathological examination shows (A) tumor in cervical stroma (H&E, x40), (B) swirling around thick walled blood vessels (H&E, x40), (C) in a myxoid, edematous stroma (H&E, x100), (D) spindle cells having oval to fusiform nuclei with eosinophilic cytoplasm. (H&E, x400)

Immunohistochemistry (IHC) (Fig 3) showed diffuse SMA positivity, positive ER, PR, desmin and negative CD34 pointing towards the final diagnosis of AAM. Ki-67 was also done which came out to be zero. Since the resection, patient had been on regular follow ups with no fresh complaints and had been improving symptomatically.

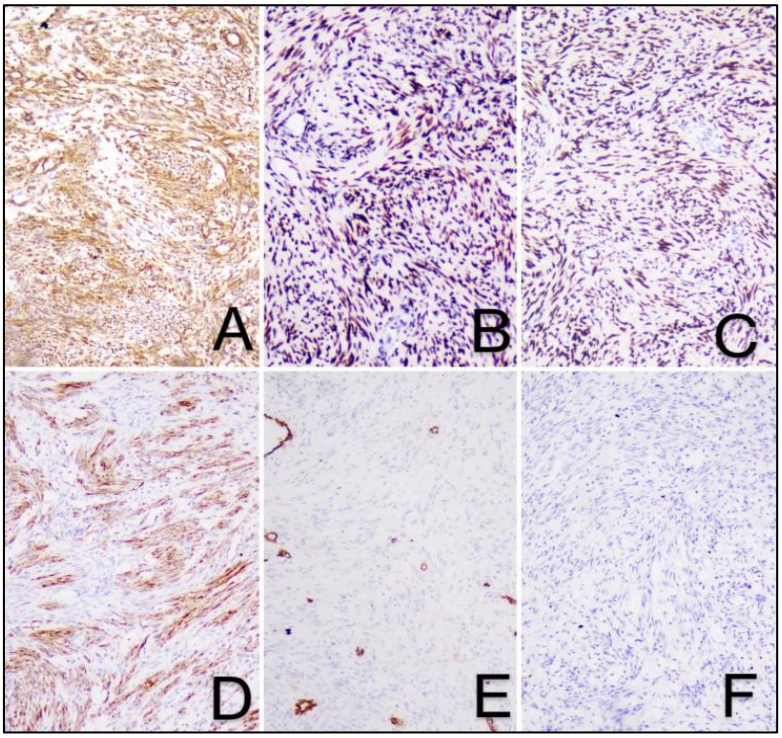

Fig 3: Immunohistochemistry shows (A) diffuse SMA positivity, (B) positive ER, (C) positive PR, (D) positive desmin, (E) negative CD34, (F) absent Ki-67. (IHC, 100X)

DISCUSSION

Mesenchymal neoplasms of female genital tract pose a great diagnostic dilemma in day to day practice owing to their unique intersection between general soft tissue tumors and genital specific mesenchymal tumors. There diversity broadly include entities like superficial myofibroblastoma, cellular angiofibroma, angiomyofibroblastoma, and aggressive angiomyxoma [1].

AAM is a distinctive entity of pelvic and perineal soft tissue tumor, proposed for the first time by Steeper and Rosai [2]. It is a slow growing, low grade neoplasm which is locally infiltrative, displacing and infiltrating pelvic organs. Relapse has been reported in 30-40% of cases giving AAM its name [3]. This tumor can reach up to enormous sizes (up to 60 cm. in diameter). AAM can be clinically misdiagnosed leading towards an almost 80% of clinical error [4]. The differentiating features of AAM are the thick walled vessels which are less numerous than thin walled vessels in angiofibroblastoma. Microscopically, AAM comprises of small, spindle to stellate cells with small uniform nuclei, indistinct nucleoli and rare nuclear pseudoinclusions, embedded in a loose, myxoid background. Mitotic figures are rare or absent. Stroma comprises of extravasated RBCs, mast cells and dilated thin and thick walled blood vessels of varying sizes distributed haphazardly. IHC reveals positive vimentin, desmin (diffuse), SMA, ER, PR but a negative S100. High-mobility group protein isoform I-C (HMGI-C) is a potentially useful marker for microscopic residual disease and differentiating from rest of differentials [3].

Another well circumscribed differential is angiomyofibroblastoma, microscopy of which shows a well demarcated nature of lesion with alternating hyper and hypocellular edematous areas. These tumor cells are seen to swirl around small to medium sized blood vessels with minimal nuclear atypia and rare mitotic figures. IHC reveals positive vimentin, desmin (focal), SMA, ER, PR but a negative S100.[5] Literature search showed only one case of metastasizing AAM and a single case of malignant transformation of angiomyofibroblastoma [6, 7].

Cellular angiofibroma is one more differential of FGT lesions in which small monotonous spindle cells arrange around numerous small to medium-sized blood vessels with prominent hyalinization of their walls, and mature adipocytes. IHC shows positivity for vimentin but stain negative for CD34, S-100, actins, desmin, keratin, and epithelial membrane antigen [8].

Vulval lesions comprise the greatest bulk of FGT lesions. Amongst them, leiomyomas are the commonest, having variation in their sizes [9]. Epithelioid and myxoid variants of vulval leiomyoma are most frequently encountered with; however an eye has to be kept upon the infiltrating margins of the tumor which brings myxoid leiomyosarcoma (mLMS) in its differential. Myxoid LMS of the uterus is, in itself, a rare neoplasm which has a more aggressive feature and worse 5-year survival [10]. IHC confirms mLMS via positivity for atleast 1 smooth muscle markers like SMA, desmin & h-Caldesmon. These tumors have a variable proliferative index varying from absent to low Ki67 [4]. Distinction between typical leiomyoma and LMS is based on three or all of the four features: tumor size, infiltrative margins, mitotic activity and cytological atypia [9]. Myxoid smooth muscle tumor of uncertain malignant potential (STUMP) are rare tumors which exceed myxoid leiomyoma criterias but fall short of a diagnosis of mLMS.

CONCLUSION

Our case study on AAM proves that mesenchymal tumors of FGT have certain overlapping features and diagnosticians should keep them in mind while facing such dilemmas.

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Conflicts of Interest: None

The study was approved by institutional review board

Authors' Contributions

Conceptualization: VD, AK
Data curation: VD
Critical and intellectual evaluation: VD, AK, SPK
Drafting of manuscript: VD, SPK
Approval of final manuscript: All authors

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