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Aggressive Angiomyxoma - A Bizarre Tumor of Pelvis

Case Report

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Introduction

Aggressive Angiomyxomas (AA) are rare infiltrative mesenchymal neoplasms that show local recurrence. They are benign tumors with strong preponderance in the reproductive age women in the pelvic and perineal regions. Due to its location, examination findings and rarity it can be misdiagnosed as other gynecological malignancies/ groin hernias. The term aggressive is a misnomer due to its nature of local recurrence and local infiltration. Wide local excision of the tumor is the treatment of choice.

This case report presents a rare infiltrative mesenchymal neoplasm of reproductive age group women with complaints of mass per vagina.

Case Report

A 32year old female came with complaints of low back pain, mass descending per-vagina, difficulty in micturition and dyspareunia. On examination a non-tender, reducible mass arising from the posterior urethral wall extending up to the left labia-major a with positive cough impulse was noted. Patient underwent MRI pelvis showed a well-defined T2, PDFS iso-hyperintense heterogeneous signal intensity lesion, T1 hypo intense lesion of size 6X4X3. 7 cm with characteristic swirled pattern in the left hemi pelvis extending inferiorly along the left lateral wall of vagina (Figure 1 and 2). Patient underwent wide local excision of lesion. Histopathological examination revealed myxoid stroma with interspersed medium to large sized blood vessels suggestive of Aggressive Angiomyxoma (Figure 3).

Discussion

Aggressive angiomyxomas are rare infiltrative mesenchymal neoplasms that commonly-recur locally. The term aggressive is a misnomer suggesting its high local recurrence after resection and not of any malignant characteristics of the lesion [1,2].

The female-to-male ratio has been reported as 6.6/1 [3]. Aggressive angiomyxomas mainly affect women of reproductive age with a peak incidence in the fourth to fifth decades of life [1,4]. It was first described in 1983 by Steeper and Rosai [5]. Aggressive angiomyxoma is an uncommon mesenchymal tumor which is mostly derived from the pelvic and perineal regions including vulva, vagina, bladder, and rectum [6,7]. However uncommon locations have also been reported like lung, liver, larynx, and orbit [8].

The main pathogenesis proposed by Nucci and Fletcher suggested that a translocation at the level of chromosome 12 is responsible where the high mobility group protein HMGA2 is located [9].

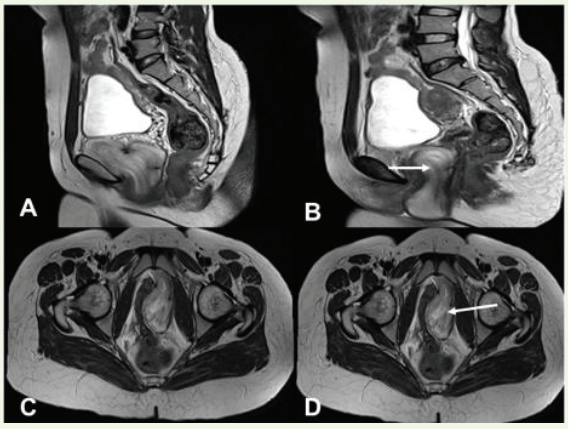

Figure 1: Sagittal T2(A,B) and Axial T2(C,D) showing swirled pattern of linear hypointense strands (arrow)within the hyperintense lesion in the left hemi-pelvis extending inferiorly along left lateral wall of vagina

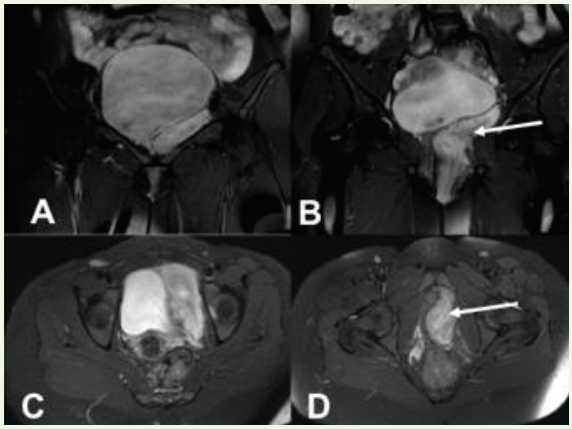

Figure 2: Coronal PDFS (A,B) showing long oval shaped hyperintense lesion (arrow) in the pelvis, Axial PDFS (C,D) showing swirled pattern of hypointense strands within the hyperintense lesion (arrow).

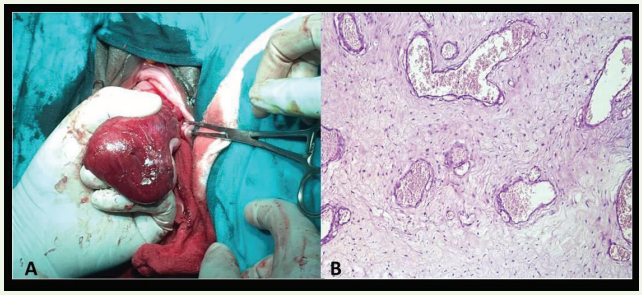

Figure 3: Intraoperative image. (A) showing fleshy mass and histopathology showing myxoid stroma with interspersed medium to large sized blood vessels (B).

Aggressive angiomyxoma is regarded as an aggressive tumor due to neoplastic nature of blood vessels, its high tendency of local infiltration and local recurrence. It is a benign tumor, but a few cases with metastasis to lungs causing death have been reported. It can be distinguished from the other lesions by its immunohistological findings. AA is derived from myofibroblasts as a phenotypic variant of the basic fibroblast with a prominent vascular component. Immunohistochemical staining of the tumor reveals high positivity for desmin, vimentin, ER, and PR receptor; however, it usually reveals negativity for S-100 protein [6,8].

The tumor tends to grow around the structures of the pelvic floor without penetrating the muscular is of the vagina or rectum [10]. Angiomyxoma remains asymptomatic until the tumor reaches large size. Urinary, gynecologic and gastro-intestinal symptoms like dysuria, dysmenorrhea, constipation, and chronic abdominal/ pelvicpain occur when the tumor begins to compress the adjacent organs including bladder, rectum, ureter, and uterus. . It is presented as painless mass in genito-femoral region or as a mass causing local pressure effect. For this reason, it is often misdiagnosed as vaginal prolapse, Bartholin's gland cyst, vulvar abscess, gynecological malignancy, femoral/groin hernia which leads to unnecessary surgical interventions.

Pre-operative imaging plays an important role in the diagnosis of aggressive angiomyxoma. On USG it appears as a hypoechoic cystic mass and is not of diagnostic importance. On CT it appears as a well-defined, hypoattenuated enhancing mass with swirling appearance in only 83% of patients [11]. MRI is more helpful than any other imaging modalities to characterize and to determine the extent of lesion. On T1W images, it appears isointense, on T2 it appears hyperintense due to high myxoid matrix and high water content [11]. Imaging not only helps in diagnosis but also helps in planning for surgery. Dynamiccontrast-enhanced MRI may be beneficial and helps to better understand tumor hemodynamics. Other non-specific findings may be internal cystic changes, background laminated appearance, large internal vessels.

In our case, the lesion was is to hypointense to the adjacent muscle on T1W, characteristic swirled pattern of linear hypointense strands within hyperintense lesion on T2W with similar appearance on PDFS. Reproductive age group woman, with painless mass descending per-vagina with characteristic swirled appearance on MRI helped in the diagnosis of Aggressive Angiomyxoma.

The characteristic MR imaging appearance of Angiomyxoma may aid in the differential diagnosis. The differential diagnosis of a pelvic or perineal softtissue mass in an adult female patient includes angiomyofibroblastoma, myxoma, infiltrating angiolipoma and myxoid lipoma. On DWI and FDG PET/CT reflects the low mitotic activity of Aggressive Angiomyxoma.

The main choice of treatment remains complete surgical excision alone with the tumor free margins. The rate of recurrence varies from 33% to 83%. Recurrence has been mostly occurred within 3 years of post-operative period3 .The patients having positive margins have more chances of recurrence than those with negative margins. Incomplete/partial excision, distant metastasis in some cases also have possibility of recurrence. In our case, wide local excision of the lesion was done with no recurrence till date. Few of the surgical complications may be present like infertility, colostomy.

Chemotherapy has no beneficial results for adjuvant therapy. Embolization remains insufficient due to vascular network of tumor. Pertaining to the reproductive organ of origin, positive ER and PR status, considering it as hormone responsive neoplasm, Tamoxifen or gonadotropin releasing hormone agonist had some beneficial effect [6,12].

Conclusion

The reproductive age of the women with painless swelling in the genito-femoral region, pressure symptoms must raise the suspicion of Aggressive Angiomyxoma. The clinical symptoms and examination may mislead the diagnosis as gynecological malignant lesions or groin/femoral hernias. Imaging with MRI plays a major role in confirming the diagnosis by its characteristic swirled pattern on T2W. Conservative en-bloc resection with microscopic positive margins achieves good local control. Histopathological correlation confirms the diagnosis.

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