Case Report

Angiomyofibroblastoma: Imaging and histopathology of a rare benign mesenchymal tumor

Kulkarni AR, Mohammed Ashfaque Tinmaswala, Shetkar SV, Kondekar S

Department of Radiodiagnosis and Imaging, Tapadia Diagnostic Centre, Aurangabad, Maharashtra, India

ABSTRACT

Angiomyofibroblastomas, aggressive angiomyxomas and cellular angiofibromas are rare mesenchymal tumours with many overlapping radiological, histopathological and immunohistochemical features. Amongst these tumours angiomyofibroblastoma is relatively benign mesenchymal tumour with very low chances of recurrence. It is clinically confused with bartholin gland cyst due to its well demarcated and smooth appearance. Due to overlapping histopathological features its very difficult even for experienced pathologists to differentiate between these mesenchymal tumours. Earlier desmin reactivity was thought to be specific for angiomyofibroblastoma but recently many aggressive angiomyxomas have also been found to be positive for desmin. Ultrasound, computed tomography and magnetic resonance imaging may be useful in diagnosis and ruling out more sinister malignancies. A well demarcated lesion with characteristic histopathological appearance of alternating hypo and hypercellular edematous regions with abundant blood vessels and stromal cells with dispersed chromatin is usually seen in angiomyofibroblastoma. Immunohistochemistry may further help in diagnosis. We here report a case of vaginal angiomyofibroblastoma. The diagnosis was made on the basis of imaging and was confirmed by histopathology and immunohistochemistry.

Key words: Histopathology and immunohistochemistry; imaging; mesenchymal tumours; vaginal angiomyofibroblastoma.

Introduction

Angiomyofibroblastomas, cellular angiofibromas, and angiomyxomas are some of the rare mesenchymal tumors usually arising from genital tract of middle-aged females.[1] There is considerable histopathological and immunohistochemical overlap between these tumors and diagnosis of these lesion; apart from histopathology and immunohistochemistry, it depends on close morphological examination.[2] Angiomyofibroblastoma is a benign mesenchymal tumor first described by Fletcher *et al*. in 1992.[3] Histologically, it consists of vascular and stromal components and usually arises from vulva, vagina, perineum, and uterine cervix. In men, these tumors may involve inguinoscrotal region.[4] Unlike aggressive angiomyxoma and cellular angiofibroma, it is a less likely to recur after complete excision.[5] We report here a case of a 35-year-old female presenting with swelling involving vagina. Imaging by ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI) was done which showed features suggestive of mesenchymal tumor. Tumor was excised laparoscopically with a rim of healthy tissue. Gross appearance, imaging, histopathology, immunohistochemistry (reactive to desmin), and estrogen and progesterone receptor positivity proved it to be angiomyofibroblastoma.

Case Report

A 35-year-old female patient presented with complaints of sensation of heaviness and mass coming out of vagina since 6

Address for correspondence: Dr. Mohammed Ashfaque Tinmaswala, Department of Radiodiagnosis and Imaging, Tapadia Diagnostic Centre, Aurangabad, Maharashtra, India.
E-mail: dr.ashfaq.memon@gmail.com

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months. There was history of this swelling being painless and gradually increasing in size. On per speculum examination, there was evidence of a globular mass. The tumor was smooth, pinkish white, and compressible. There was no visible ulceration or bleeding over its surface [Figure 1].

Ultrasound showed an isoechoic mass lesion arising from right vaginal wall. On color Doppler imaging, there was evidence of vascular channels traversing the mass lesion [Figures 2 and 3]. There was no abdominal or pelvic lymphadenopathy on ultrasound examination.

Contrast-enhanced CT showed inhomogeneous mass in pouch of Douglas. This mass was seen arising out of right vaginal wall and was causing displacement of rectum. At the level of perineum, this mass was at right paravaginal region and was seen extending into gluteal fat [Figures 4-6].

An MRI was also done which showed hyperintense polypoidal lesion arising out of right vaginal wall. This lesion was seen protruding into presacral space [Figure 7].

Based on imaging features, the differential diagnoses considered were cellular angiofibroma, angimyxoma, and angiomyofibroblastoma. The patient was operated by laparoscopic excision of the mass with a rim of healthy tissue.

The histopathological examination revealed the lesion to be consisting of blood vessels and stromal cells with dispersed chromatin [Figure 8].

Immunohistochemistry revealed it to be strongly positive for estrogen and progesterone receptors. The lesion was found to be immune-reactive for desmin. Well-demarcated lesion, histopathological examination, and desmin positivity of the

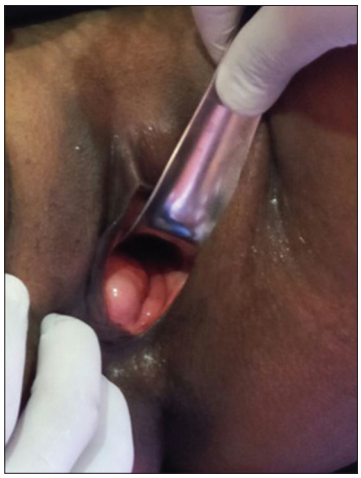

Figure 1: Per speculum examination showing paravaginal swelling which is grossly smooth without any evidence of ulceration or bleeding

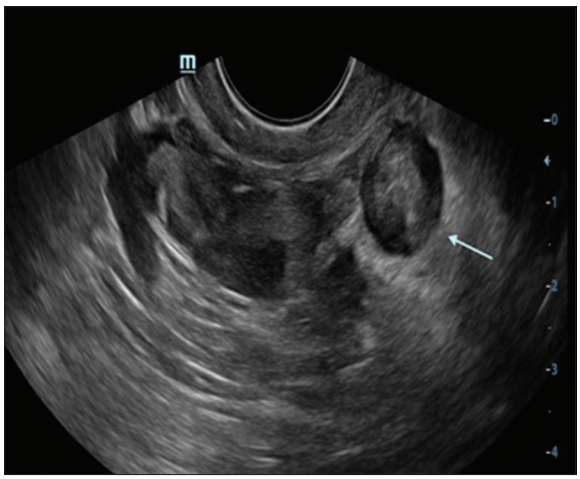

Figure 2: Transperineal ultrasound showing isoechoic mass lesion arising from right vaginal wall near anal sphincter (arrow)

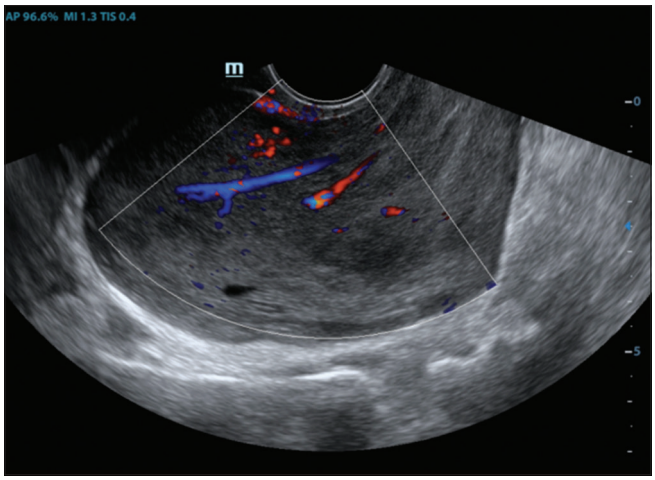

Figure 3: Transvaginal ultrasound showing well-defined homogeneous lesion with traversing vascular channels

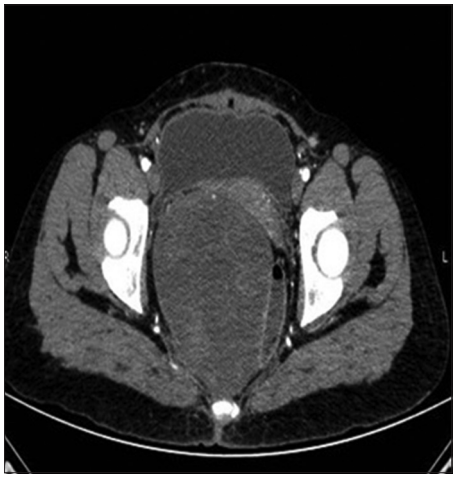

Figure 4: Contrast-enhanced CT: Axial image showing inhomogeneous mass in POD causing rectal displacement to right

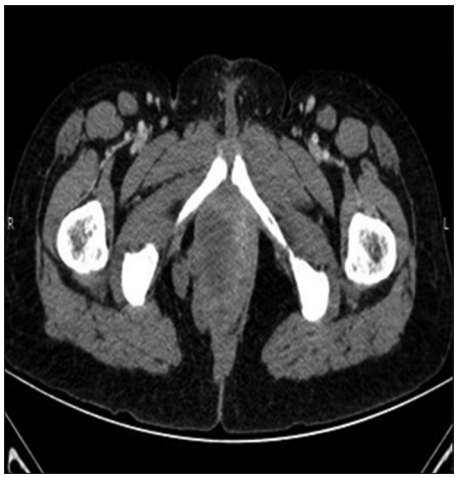

Figure 5: At the level of perineum, an enhancing mass is seen at right paravaginal region with extension into gluteal fat

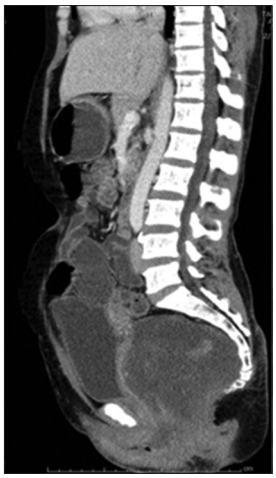

Figure 6: Contrast-enhanced CT right parasagittal image showing polypoidal soft tissue density mass with few enhancing components

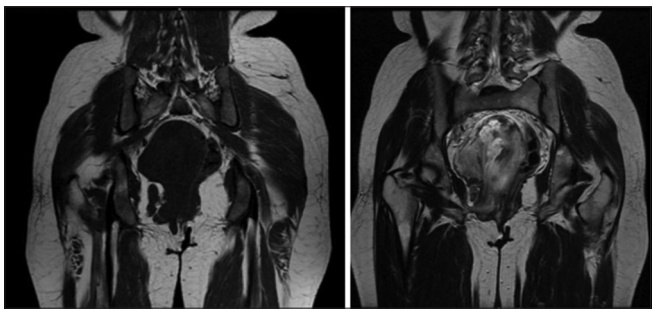

Figure 7: Coronal MRI T1 (left) and T2 (right)-weighted images showing polypoidal mass arising from right vaginal wall and showing signal intensity similar to muscles with few areas of flow void and necrosis

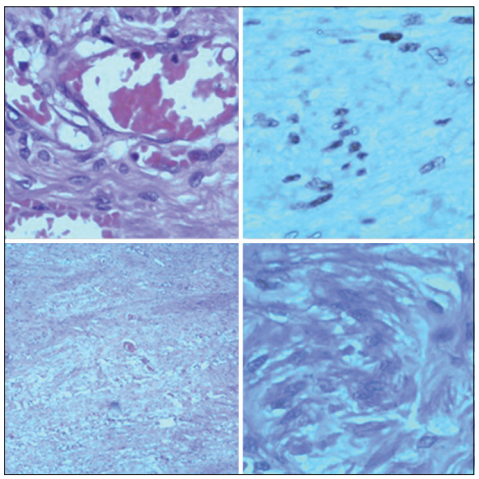

Figure 8: Histopathological examination showed spindle cells in fascicles interspersed with number of thin walled blood vessels. Individual cells were elongated with vesicular nucleus and dispersed chromatin. No significant mitosis was seen

lesion without any evidence of mitosis favored diagnosis of angiomyofibroblastoma. Post surgery, the patient is disease-free since 8 months.

Discussion

Fletcher *et al*. described 10 cases of benign tumors of vulva which mimicked aggressive angiomyxoma.[3] On gross examination, these tumors are typically well circumscribed, with rubbery consistency and pink in color. On ultrasound, these tumors usually present as mixed echoic soft tissue tumors sometimes containing cystic spaces within the mass. The correlation of ultrasound findings with pathological findings suggests that echogenic areas correspond to hypocellular myxoid stroma, hypercellularity presents as hypoechogenicity on ultrasound, and cystic spaces correspond to dilated glands in vulva. On, CT angiomyofibroblastoma may present as hypoattenuating mass with inhomogeneous central enhancement. Hypoenhancement most likely corresponds to hypocellular myxoid stroma and inhomogeneous enhancement may be related to hypercellular zones.[6] On MRI, these tumors appear isointense to skeletal muscles on T1- and hyperintense on T2-weighted images. Postcontrast images may show strong enhancement.[7] These imaging features are not exclusive to angiomyofibroblastoma, and it is important to distinguish this rare tumor from other equally rare mesenchymal tumors such as aggressive angiomyxomas and cellular angiofibromas.[8]

The histopathological examination of angiomyofibroblastoma may show well-demarkated lesion with alternating hypo- and hypercellular edematous regions with abundant blood vessels. Tumor cells tend to cluster around blood vessels and there is presence of epitheloid elements in angiomyofibroblastoma.[9] Immunohistochemical examination of these tumors show them to be positive for desmin. In fact, some authors have previously concluded that desmin can be used to differentiate between cellular angiofibromas and angiomyofibroblastomas.[10] But many cases of aggressive angiomyxomas have also been reported to be desmin positive, hence histopathological and

immunohistochemical diagnosis of angiomyofibroblastoma even today remains fairly complicated. Complicated as it may be, it is of utmost clinical importance to differentiate between angiomyofibroblastoma and other mesenchymal tumors such as aggressive angiomyxomas and cellular angifibromas because angiomyfibroblastoma runs a benign course and is less likely to recur than more aggressive lesions such as aggressive angiomyxoma.

Conclusion

Angiomyofibroblastoma is a rare mesenchymal tumor which usually arises from female genital tract. It needs to be differentiated from other equally rare but more aggressive mesenchymal tumors such as cellular angiofibroma and aggressive angiomyxoma. A well-demarcated lesion with characteristic radiological appearance, having immunoreactivity to desmin and estrogen and progesterone receptor positivity is more likely to be angiomyofibroblastoma than cellular angiofibroma or aggressive angiomyxoma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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