

Multimodality imaging of vaginal rhabdomyosarcoma

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Abstract

Rhabdomyosarcoma (RMS) is a malignant mesenchymal tumor arising from the embryonal muscle cells (rhabdomyoblasts), and is the most common soft tissue sarcoma in children and young adults accounting for 4–6% of all malignancies in this age group. Though rare overall, embryonal rhabdomyosarcoma is the most common malignancy arising in the pediatric female genitourinary tract with sarcoma botryoides being the most common variant of the tumor. In young and adolescent individuals, the cervix and uterus are affected; whereas in infants, vaginal lesions are more common. Imaging plays a crucial role not only in the initial diagnosis but also in long-term follow-up of genital RMS. We describe a rare case of embryonal rhabdomyosarcoma of the vagina occurring in a 23-year-old female who presented with abnormal vaginal bleeding ever since she was a child.

Key words: Embryonal rhabdomyosarcoma; sarcoma botryoides; vaginal tumor

Introduction

Soft tissue sarcomas (STS) are a group of malignant tumors that originate from primitive mesenchymal tissue and account for 7% of all childhood tumors. Rhabdomyosarcomas (RMS) and undifferentiated sarcomas account for approximately 50% of soft tissue sarcomas in children.^[1] Almost two-thirds of RMS cases are diagnosed in children less than 6 years of age, however, embryonal RMS of the vagina usually occurs in female infants and early childhood, in contrast with the corresponding tumor in the cervix which occurs at later ages.^[2] The former is a rare and rapidly growing tumor that presents as bleeding per vaginum or as a polypoid fleshy mass filling and protruding through the vagina.^[3] The tumor grows as a submucosal lesion giving the typical “grape-like” appearance. We report the imaging features in a rare case of sarcoma botryoides of the vagina presenting with abnormal vaginal bleeding in a

23-year-old female. To our knowledge, no case of vaginal RMS has been reported in the literature in an adolescent female.

Case Report

A 23-year-old nulliparous female presenting with menorrhagia and dysmenorrhea for 2 months was referred to our department for pelvic sonography. Associated urinary frequency was also noted, and on clinical examination, a reddish purple grape-like mass was seen extruding from the vagina. Labia majora, minora, clitoris, urethra, and the perineum appeared normal as well without any skin lesions. The clinical impression was that of a vaginal vascular malformation or a polypoidal myoma. On two-dimensional (2D) transabdominal scan, a large heteroechoic mass lesion was noted expanding the vagina [Figure 1A]. The lesion was also suspected of

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involving the cervix on 2D sonography with no ascites or pelvic adenopathy. On color Doppler examination, the lesion showed significant internal vascularity with both arterial and venous flow [Figure 1B]. The findings were confirmed on a transabdominal three-dimensional (3D) sonography [Figure 1C] whereby the lesion was seen lifting the cervix superiorly on coronal reconstruction images. The external aspect of the cervix was adherent to the mass while the suspicion of transmural infiltration was refuted. Preoperative pelvic magnetic resonance imaging (MRI) was done to have a global view of the lesion and for better assessment of pelvic anatomy and wall structures. The MRI confirmed the impression proposed on sonography. The mass was predominantly of low signal intensity on T1-weighted and of high signal on T2-weighted images [Figure 2A and B]. Multiple mildly thickened septations were seen which demonstrated moderate to intense enhancement after gadolinium administration [Figure 2C]. Few foci of internal necrosis/cystic change were also seen. Fat planes with the walls of adjacent organs as well as pelvic/perineal fat planes were well preserved. Uterus and cervix were normally visualized. No significant locoregional lymphadenopathy was seen. The patient underwent work-up for metastasis which was negative for any metastases. Punch biopsy of the lesion was performed which showed undifferentiated malignant cells having oval to spindle-shaped hyperchromatic nuclei with nuclear pleomorphism; mitotic figures were also seen. Tumor cells were arranged in short fascicles with alternating hypercellular and hypocellular myxoid areas. Foci of hemorrhage and necrosis were present. Rhabdomyoblastic differentiation of tumor cells and cross striations were not clearly visible. Imprint smears showed short fascicles and dissociated spindle cells with bipolar extension of

cytoplasm and plump vesicular nuclei with mild nuclear pleomorphism. Overall, the histological examination was consistent with embryonal rhabdomyosarcoma of low malignant grade [Figure 3]. The surgical findings were in concordance with the overall impression made on the basis of all imaging modalities put together [Figure 4]. Hysterectomy was performed in discussion with the patient and she was relieved of her symptoms. The patient was asymptomatic till last follow-up at 2 months.

Discussion

RMS is the most common pediatric soft-tissue sarcoma and constitutes 3–5% of all malignancies in childhood,^[4] most of which occur in the head and neck. RMS occurring outside the head and neck region comprise 40% of RMS, of which around 15% are genitourinary (GU) nonbladder prostate tumors (GU-NBP, i.e., paratesticular, vaginal and uterine tumors), 10% are bladder-prostate tumors (BP), 15% occur in the limbs, and 20% occur in other sites (i.e., thoracic or abdominal tumors). RMS is a fast-growing, primitive, high-grade, malignant mesenchymal tumor. Depending on their degree of differentiation, the tumor cells manifest features that more or less can be found in the cells of skeletal muscle. These features, essential for the diagnosis, are the presence of myofibrils and cross striations (on light and electron microscopy) and/or positive immunohistochemical staining for markers of muscle differentiation such as desmin and myoD1. Based on morphology, RMS is traditionally

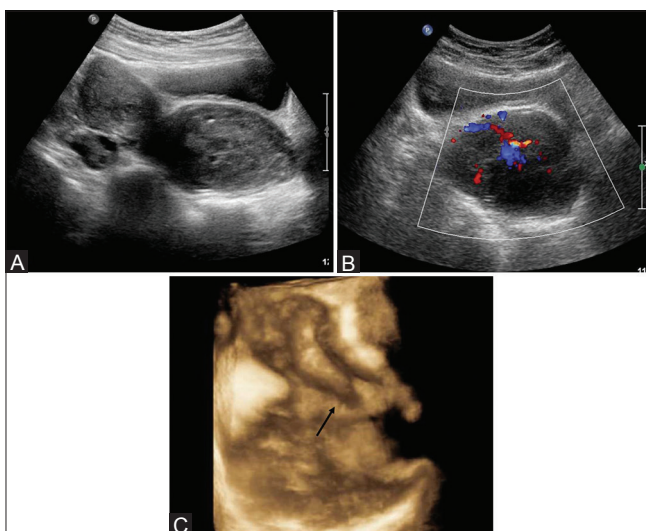


Figure 1 (A-C): (A) Transabdominal 2D USG sagittal image shows a large heterogeneous mass epi-centered in the vagina. (B) On color Doppler, the mass shows significant internal vascularity. (C) Transabdominal 3D USG confirms lifting up of the endocervix superiorly without any obvious cervical infiltration (straight arrow)

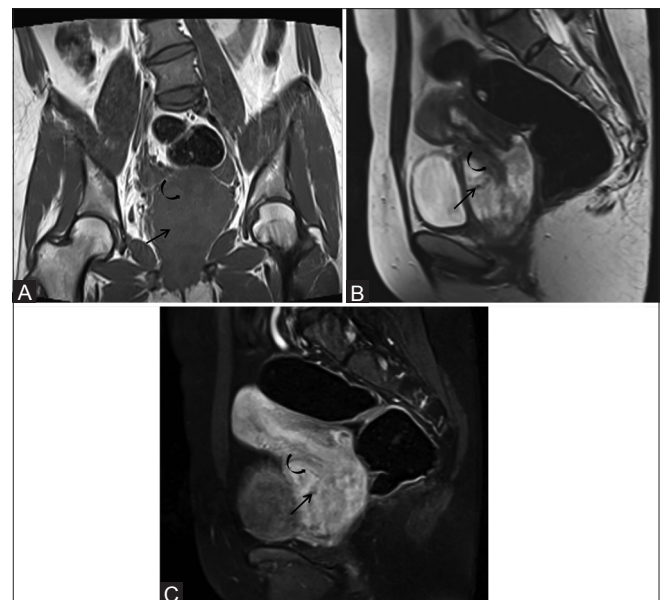


Figure 2 (A-C): MRI images of the patient showing the vaginal mass (straight arrow) and the cervical lip (curved arrow) on T1W pre-contrast coronal (A), Sagittal T2-weighted (B), and post-contrast fat suppressed T1W sagittal (C) images. Note the mass is entirely vaginal in location with lifting up of the cervical lip. The lesion shows homogenous post-contrast enhancement and different intensity than the myometrium on T2 weighted sequence, on T1W sequence however the lesion is isointense to myometrium

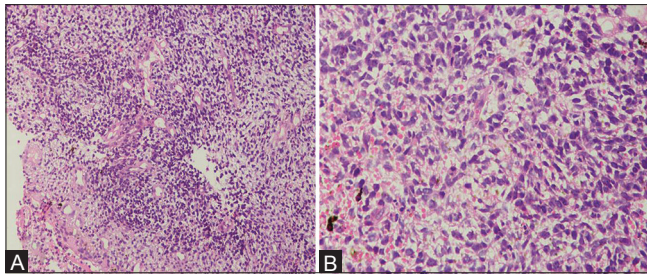


Figure 3 (A and B): (A) Intermediate-power photomicrograph shows tumor cells arranged in short fascicles with alternating hypercellular and hypocellular myxoid areas. (H and E stain). (B) High-power photomicrograph shows undifferentiated malignant cells having oval to spindle shaped hyperchromatic vesicular nuclei with nuclear pleomorphism and bipolar extension of cytoplasm. Mitotic figures are also seen. (H and E stain)

subdivided into embryonal, alveolar, and pleomorphic. Embryonal RMS is the most common type (60–70% of all RMS). The cells show a close resemblance to various stages in the embryogenesis of normal skeletal muscle. Subtypes are botryoid RMS and spindle-cell RMS. When arising in the submucosa, embryonal RMS may present as a fast-growing exophytic, polypoidal mass. This macroscopic variant is called botryoid RMS and, due to its growth pattern (primary exophytic and not invasive), has a better prognosis.^[5]

The word botryoid in Greek means a “bunch of grapes” which characteristically describes the clinical appearance of the tumor. Sarcoma botryoides is usually reported as a vaginal tumor in female reproductive tract of infants,^[6] and is the most common malignancy arising in the GU tract of the pediatric population prior to the age of 15.^[7] Vaginal bleeding is the most common presenting feature even though nonspecific. It may also present as a polypoid or fleshy mass in the vagina, or more classically projecting from the introitus. Other forms of presentations include urinary symptoms, especially when the tumor is anteriorly situated or tenesmus where there is posterior extension.^[8,9] The survival rate of vaginal and cervical lesions has been reported to be 60–96%, respectively.^[10] Botryoid morphology is characteristic, but not specific, for RMS within the vagina because yolk sac tumor may have a similar appearance. Invasion of adjacent structures by the primary tumor may make the precise anatomic origin of GU RMS difficult to determine on cross-sectional images. Recent refinements in multidisciplinary imaging modalities have dramatically improved the diagnosis of genitourinary RMS. Diagnostic imaging also plays an important role in monitoring response to therapy.

Imaging findings

As RMS is a soft-tissue tumor, conventional radiology plays an insignificant role in its diagnosis. USG is often the first imaging modality with soft-tissue masses because it is readily available, has high resolution, and can easily assess the extent and vascularity of a mass. As in the present case on USG, RMS presents as a well-defined,

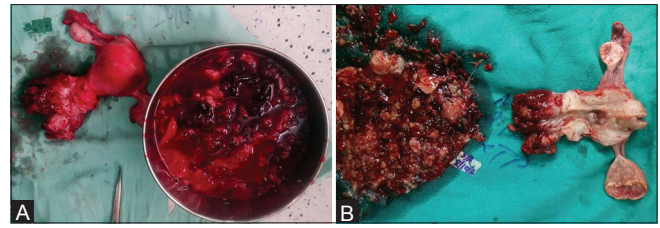


Figure 4 (A and B): Surgical specimens (A) gross and (B) cut section in coronal plane showing “grape-like” mass epicentered in the vaginal with involvement of the cervix

slightly hypoechoic inhomogeneous mass that can show significantly increased flow. USG can also demonstrate pelvic retroperitoneal lymphadenopathy. USG is also of use in image-guided biopsies. Further, a 3D sonography can be useful to demonstrate the relation of vaginal mass to the fornices and the cervical lip, as in the present case an initial suspicion of cervical invasion was there which was resolved on a 3D sonography. With its superior ability to depict soft-tissue changes, MRI is the primary imaging modality in RMS.^[11] Although imaging protocols should be tailored for individual patient, they should at least consist of axial T1-weighted and T2-weighted images (for anatomic detail and assessment of neurovascular structures), coronal T1-weighted images, and imaging after gadolinium administration. It is important that at least two series should be identical, one before and one after contrast agent administration, to be able to discern enhancement. Contrast-enhanced series are mandatory and ideally be performed with fat saturation. RMS show low to intermediate signal intensity on T1-weighted images and on T2-weighted images they tend to be of intermediate-to-high signal intensity. The high T2 signal due to abundant myxoid stroma within these tumors may give it a multiseptated cystic appearance and suggest the botryoid variant.^[7] On postcontrast imaging RMS demonstrate strong enhancement. In very rare instances, the tumor may show a predominantly cystic appearance. Positron emission tomography-Computed tomography (PET-CT) and CT studies should be done for metastatic work up. As proposed during our work-up of the case, the differential diagnosis includes vaginal vascular hamartomas and polypoidal myomas. While the former would show presence of extensive flow voids and altered blood at various phases of evolution on MRI, the latter shows a typical whorled pattern of stroma with intervening septations of dense fibrosis. Both lesions show variable contrast enhancement with the mature myomas usually showing more homogenous contrast uptake than a vascular hamartoma. On the basis of imaging work up in the present case we were able to rule out both these close differentials.

Sarcoma botryoides is a rapidly growing malignancy. We present a rare case of the botryoid variant of embryonal rhabdomyosarcoma arising from the vagina in an adult female and describe the imaging features with pathologic

analysis. Multidisciplinary imaging approach can help in accurate diagnosis and assessment of tumor extent. MRI provides superior delineation of tumor size, location, and relationship to other organs. It is the modality of choice in evaluating local disease.

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Conflicts of interest

There are no conflicts of interest.

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