Magnetic Resonance Findings in Adolescent Vaginal Rhabdomyosarcoma: A Rare Case Report

Trifonia Pingkan 1*, Fitri Juniarta 2, Hartono Tjahjadi 3, Meliyana 1
1 Department of Radiology, Doctor Cipto Mangunkusumo National General Hospital, Faculty of Medicine, Universitas Indonesia, Jakarta, Indonesia
2 Department of Radiology Residency Program, Doctor Cipto Mangunkusumo National General Hospital, Faculty of Medicine, Universitas Indonesia, Jakarta, Indonesia.
3 Department of Pathological Anatomy, Doctor Cipto Mangunkusumo National General Hospital, Faculty of Medicine, Universitas Indonesia, Jakarta, Indonesia

*Corresponding author:
Trifonia Pingkan
Department of Radiology, Doctor Cipto Mangunkusumo National General Hospital, Faculty of Medicine, Universitas Indonesia, Jakarta, Indonesia
trifonia.pingkan01@ui.ac.id

INTRODUCTION

Vaginal (RMS) is the most common type of all genitourinary RMS that occurs most frequently in the infants and early childhood period rather than adolescents and young adults. Most cases are diagnosed in children less than 6 years of age. The presentation of vaginal RMS is a rapidly growing tumor that presents as vaginal bleeding or as a polypoid fleshy mass filling and protruding through the vagina [1–3].

CASE PRESENTATION

A 15-year-old female was treated at Cipto Mangunkusumo Hospital with a rapidly growing mass on the vagina and vaginal bleeding for seven months accompanied by pain in the lower abdomen region. On general examination, the patient appeared pale and moderately ill. We found a solid mass measuring 8 x 8 x 5 cm protruding from the vagina.

The laboratory tests revealed normocytic normochromic anemia, hypoalbuminemia, hyponatremia, and urinary tract infection. The chest radiograph was normal. Pelvic magnetic resonance imaging (MRI) was performed and showed a bulky solid tumor of 10.5 x 10.5 x 12.2 cm in the vagina, posterior to the bladder, with a heterogeneous low signal intensity in T1-weighted (T1 W) sequences (Figure 1A), high signal intensity in T2-weighted (T2 W) sequences (Figure 1B and 1D), high signal intensity in Diffusion-Weighted Imaging (DWI) sequences with b value = 1.000 s/mm2 (Figure 1E), and low signal intensity in Apparent Diffusion Coefficient Map (ADC map) sequences (Figure 1F). There was also a strong and heterogeneous enhancement after gadolinium contrast administration (Figure 1C). This bulky tumor displaced the cervix and uterine corpus to the superior, displaced bladder to the cranial part, and compressed the rectal lumen without any infiltration to those pelvic organs. Histopathological examination of the detached tissue from the vagina showed that...
the cell nucleus was coiled and had mild to moderate pleomorphism (Figure 2A). Hyperchromatic medium to large tumor cells with eosinophilic cytoplasm were found (Figure 2B). Immunohistochemical staining results were positive for myogenin, Myo-D1, and desmin (Figure 2C).

DISCUSSION

Genitourinary RMS is the second-most-common site of all RMS. This is a fast-growing, primitive, high-grade, malignant mesenchymal tumor. In adolescents, the cervix and uterine are the most common sides of RMS, but in this paper, we found a rare case of vaginal RMS that occurs in adolescents. The vagina is the most common organ location in infant and young-age genitourinary RMS. The diagnosis of this tumor is very challenging because the rapidly growing mass can affect the other pelvic organs [2,4].

Radiology plays an important role in diagnosis to describe the tumor origin, tumor characterization, tumor effect on the other pelvic organs, lymph node metastasis, and distant metastasis. MRI is the gold standard imaging in vaginal RMS that shows aggressive locally vaginal tumor, high signal intensity on T2WI, and heterogeneous low signal intensity on T1WI with strong enhancement after gadolinium administration. MRI is the best imaging modality to identify the effect or infiltration to the other pelvic organs (uterus, bladder, or rectosigmoid colon). In some cases, we can find hemorrhage transformation in

![Figure 1. Pelvic magnetic resonance imaging (MRI) of (A) Sagittal T1W; (B) Sagittal T2W; (C) Sagittal T1W Fat Saturation with Contrast (T1 W FS + C); (D) Axial T2W; (E) Axial DWI with b value 1,000 s/mm²; (F) Axial ADC Map, show a bulky heterogeneous solid mass with highly restriction and heterogeneous enhancement protruding from the vagina.](image1)

![Figure 2. (A) Macroscopic specimen of the detached tissue from the vagina shows irregular tissue with blood clots in several parts; (B) The tumor consists of subepithelial mesenchymal cells with moderate to high cellularity; (C) There is immunohistochemical evidence of skeletal muscle differentiation with focal positivity to myogenin, Myo-D1, and desmin.](image2)
this bulky mass that appears as hyperintensity foci on T1WI [2,6].

Histopathological examination is the gold standard diagnostic modality in RMS. In this case, the pathology finding is a botryoid sarcoma as a differential diagnosis. The immunohistochemical staining result was rhabdomyosarcoma not otherwise specified (NOS) which, based on epidemiology, is more common in adult patients. Based on clinical manifestations, radiological features, histopathological and immunohistochemical examinations, the diagnosis was rhabdomyosarcoma NOS [5,6].

Treatment of genitourinary RMS is necessarily multimodality, combining chemotherapy with locoregional treatments, essentially surgery and/or radiotherapy. The current paradigm of surgical treatment of RMS is complete wide excision of the primary tumor with a margin of uninvolved tissue whenever possible, but the patient, in this case, was not considered suitable for surgery. The standard chemotheraphy regimen for patients with rhabdomyosarcoma is the combination of vincristine, actinomycin, and cyclophosphamide/ifosfamide. Because of poor medication adherence, the patient only received two cycles of vincristine, actinomycin D, and ifosfamide chemotherapy. She was also treated with palliative hemostatic external beam radiation therapy. Some prognostic factors for RMS are organ invasion and lymph node metastasis. Several factors are beneficial for the prognosis in this patient, such as the absence of lymph node and distant metastases at the time of diagnosis and the successful complete tumor excision. However, there are unfavorable prognostic factors, such as histologically non-embryonal variants, tumor size more than 5 cm, and age more than 10 years at the time of diagnosis [2,4,6].

CONCLUSIONS

Adolescent vaginal RMS is a rare disease. Its management requires a multidisciplinary approach for diagnosis and treatment. In imaging workup for diagnosis, MRI is the best modality of choice in vaginal RMS. MRI provides superior characterization for tumor size, location, and association with other pelvic organs and identifies lymph node metastasis. Its management requires a multidisciplinary combination of several therapeutic modalities to improve prognosis.

DECLARATIONS

Competing of Interest
The authors declare no competing interest in this study.

Acknowledgment
The author would like to thank all those who have been involved in writing this paper so that it can be completed and hopefully will be useful for clinicians in their daily practice.

REFERENCES