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Case Report

Pelvic Alveolar Rhabdomyosarcoma: A Diagnostic Challenge and Imaging Insights

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Abstract

Rhabdomyosarcoma is a primitive malignant soft tissue sarcoma of skeletal muscle phenotype that originates from a primitive mesenchymal cell. It is the most common soft tissue tumor in the young population. We present a case of a 22-year-old female patient with alveolar rhabdomyosarcoma in an unusual para-vesical and para-uterine location, mimicking an ovarian tumor. This case highlights the diagnostic challenges faced by clinicians and provides useful imaging insights to differentiate ovarian from non-ovarian tumors.

Keywords: *Pelvic Alveolar Rhabdomyosarcoma; Imaging Insights; Ovarian from Non-Ovarian Tumors*

Introduction

Rhabdomyosarcoma is a malignant soft tissue sarcoma of skeletal muscle phenotype originating from a primitive mesenchymal cell. It is the most frequent soft tissue tumor in the young population, accounting for 5% of all childhood cancers. In adults, it represents only 0.03% of malignancies [1]. Most cases are sporadic, though some are associated with familial syndromes. There are four recognized subtypes of rhabdomyosarcoma: embryonal (60%), alveolar (20%), pleomorphic (10%), and spindle/sclerosing (10%). While definitive diagnosis relies on pathological examination, MRI and CT scans can indicate distinctive characteristics that help differentiate rhabdomyosarcoma subtypes from other soft-tissue tumors. Despite advancements in treatment, overall survival remains poor.

Case Report

A 22-year-old female presented to our hospital with chronic pelvic pain persisting for three months. She exhibited minimal tenderness in the hypogastric region but had no associated symptoms such as fever, menstrual irregularities, or general health deterioration.

An initial ultrasound revealed a large pelvic mass with mixed echogenicity, comprising both cystic and solid components, with detectable blood flow on color Doppler. The mass appeared to be attached to the ovary, prompting further evaluation with MRI.

Contrast-enhanced pelvic MRI identified a right para-vesical and para-uterine mass (Figure 1) measuring 118x32x85 mm, exhibiting a triple composition: a cystic component hyperintense on both T1 and T2 sequences, a liquid component hypointense on T1 and hyperintense on T2 containing a 5 mm thick septum, and a solid component isointense to muscle on T1-weighted images, demonstrating enhancement post-gadolinium injection (Figure 2).

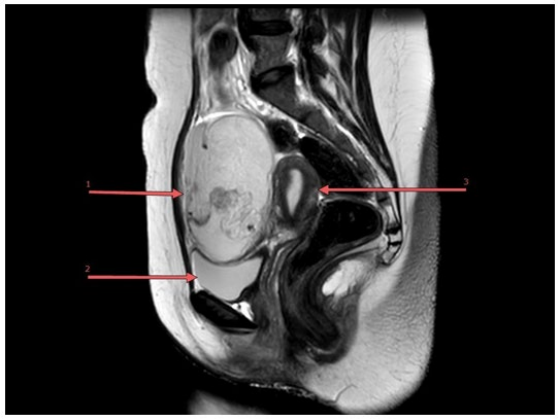

Figure 1: *Pelvic MRI, T2 sagittal sequence, displaying a pelvic mass (1), displacing without infiltrating the bladder (2) and uterus (3).*

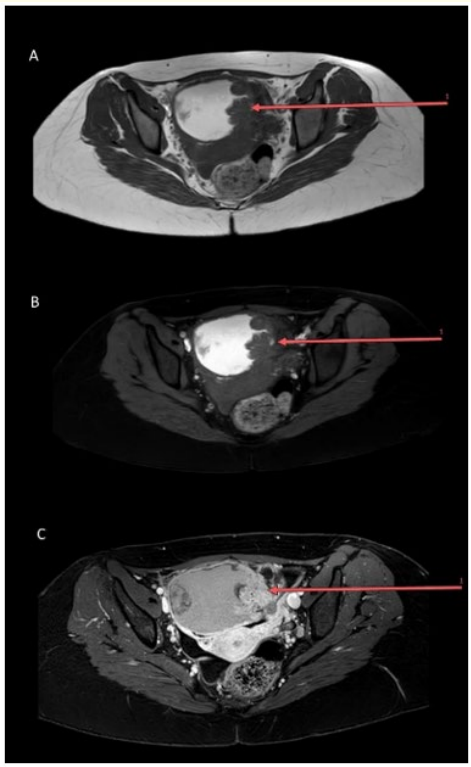

Figure 2: *Pelvic MRI, axial T1 (A), T1 FS (B) and T1 FS with contrast injection (C), showing a masse with solid component (1) which is isointense to muscle on T1 sequences, enhanced after gadolinium injection.*

Due to the close proximity between the ovaries and the tumor (Figure 3 and 4), diffusion-weighted imaging with a low B value was instrumental in confirming the non-ovarian origin of the mass. It revealed a hyperintense right ovary, displaced yet distinctly separate from the mass (Figure 5).

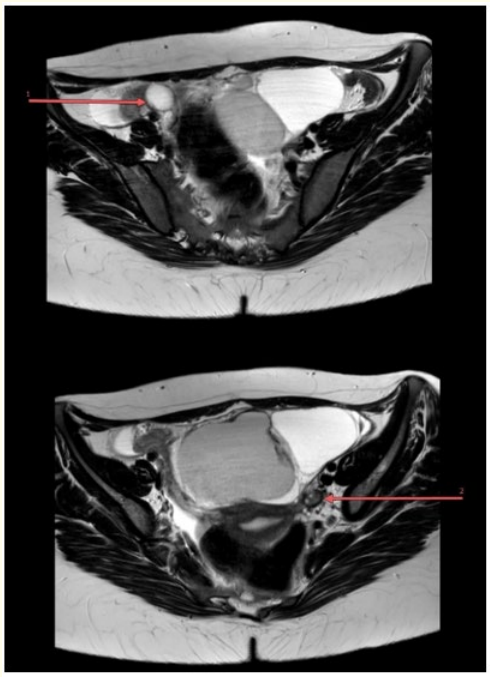

Figure 3: *Pelvic MRI, axial T2 sequence, showing the right (1) and left (2) ovaries.*

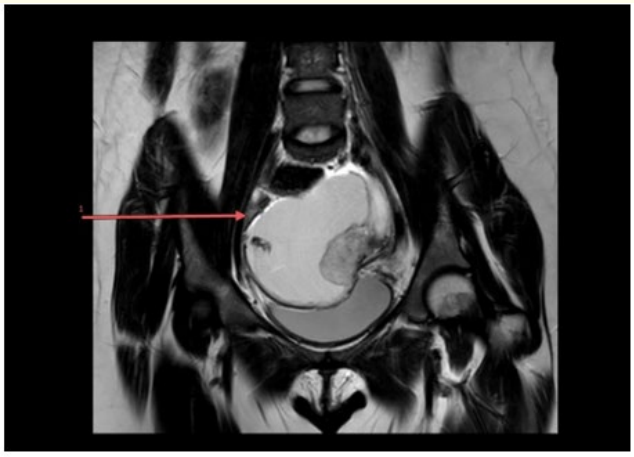

Figure 4: *Pelvic MRI, coronal T2 sequence, showing the right ovary with a pseudo claw sign appearance with the mass.*

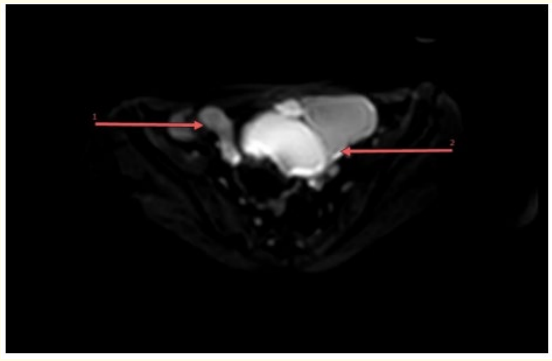

Figure 5: *Pelvic MRI, axial DWI sequence, showing the ovary which is hyperintense, separated from the mass, confirming the extra ovarian origin of the tumor.*

The patient underwent complete surgical resection of the mass. Histopathological analysis revealed cavities lined by cells with atypical nuclei, consistent with alveolar rhabdomyosarcoma (Figure 6), with strong desmin immunostaining (Figure 7). Postoperative surveillance MRI confirmed the integrity of both ovaries and the preservation of the uterus and bladder, confirming the extra genital origin of the tumor (Figure 8).

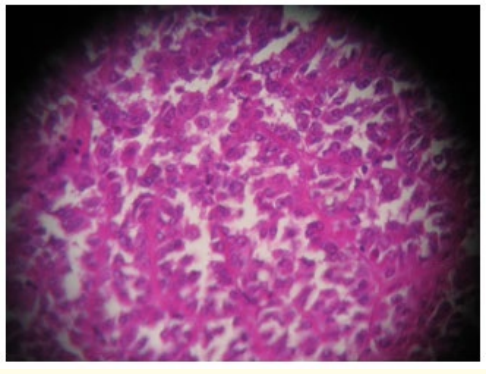

Figure 6: *Histopathological finding in alveolar rhabdomyosarcoma: cavities lined by cells with atypical nuclei, suggestive of alveolar rhabdomyosarcoma.*

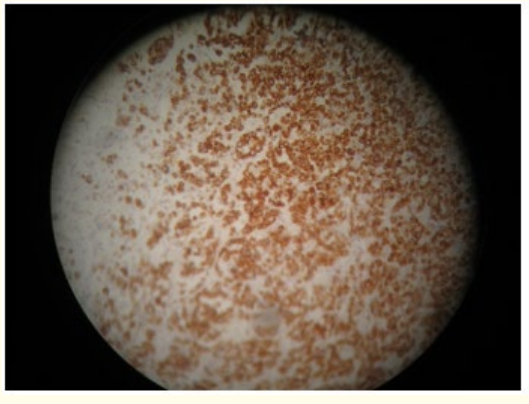

Figure 7: *Histopathological finding in alveolar rhabdomyosarcoma: strong staining with anti-desmin antibody.*

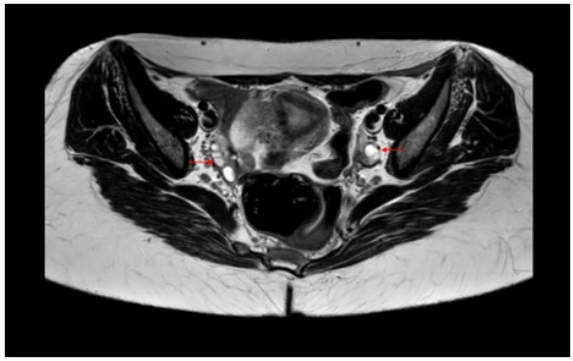

Figure 8: *Pelvic MRI, coronal T2 sequence, showing both the right and left ovaries (arrows).*

Discussion

Rhabdomyosarcoma is the most common soft tissue tumor in the young population, accounting for 19% of childhood soft tissue sarcomas and 5% of all childhood cancers [1,3]. However, it is rare in adults. Rhabdomyosarcomas originate from primitive muscle cells and can arise in various locations, from the orbits to the extremities. They are highly malignant in nature.

The latest edition of the World Health Organization (WHO) Classification of Tumors of Soft Tissue and Bone redefined rhabdomyosarcoma subtypes. Botryoid rhabdomyosarcoma is now categorized under embryonal RMS, while spindle cell/sclerosing RMS is recognized as a distinct subtype [4]. Alveolar and pleomorphic RMS remain largely unchanged in classification.

Embryonal rhabdomyosarcoma typically arises in the head and neck (47%) or genitourinary system (28%) and is associated with Beckwith-Wiedemann syndrome. Alveolar rhabdomyosarcoma is a highly aggressive neoplasm with a poor prognosis, characterized by primitive cells with round nuclei and arrested myogenesis. Prognosis depends on age, stage, and location, with extremity tumors having the worst outcomes. Pleomorphic rhabdomyosarcoma primarily affects adults and is characterized by bizarre polygonal, round, and spindle cells. It commonly occurs in deep soft tissues, particularly the thigh, and has a poor prognosis. Spindle cell/sclerosing rhabdomyosarcoma is a rare variant occurring in both children and adults, with children typically diagnosed at an early stage and achieving a 95% five-year survival rate [4].

Initial imaging typically involves an abdominal ultrasound, which identifies a heterogeneous, often well-vascularized mass within the abdominal cavity, distinct from solid organs. The mass is generally displacing rather than invading adjacent structures. On CT, rhabdomyosarcomas can appear iso- to hyperdense compared to skeletal muscle, with necrosis being a common feature. The presence of calcifications is atypical and argues against rhabdomyosarcoma. Bone invasion is seen in approximately 25% of cases, and metastases, usually lytic, may also be present. MRI findings typically show the tumor as isointense to muscle on T1-weighted images and heterogeneously hyperintense on T2-weighted images, often with necrotic areas. Alveolar rhabdomyosarcoma is particularly notable for its marked enhancement and high vascularity [1,5].

The standard treatment approach involves surgical excision when feasible, followed by radiation therapy and chemotherapy. Despite aggressive multimodal treatment, prognosis remains poor due to the high frequency of metastases at initial diagnosis and the high recurrence rate. Lymph node involvement is common at presentation, with the lungs being the primary site for metastases. The five-year survival rate remains below 50%, with pediatric cases generally having a better prognosis than adults [6].

Conclusion

Alveolar rhabdomyosarcoma is an aggressive malignancy that can pose a diagnostic challenge when occurring in atypical locations, such as the para-vesical and para-uterine regions. This case underscores the importance of advanced imaging techniques, particularly MRI, in distinguishing ovarian from non-ovarian tumors. Early recognition and accurate diagnosis are crucial in guiding appropriate surgical and therapeutic management, ultimately improving patient outcomes.

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