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Radiology

Aggressive Parameningeal Alveolar Rhabdomyosarcoma in A Child: A Case Report and Review of Imaging Features

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Abstract **Case Report**

Rhabdomyosarcoma is the most common soft tissue sarcoma in children and frequently involves the head and neck region. Parameningeal locations are associated with delayed diagnosis and poor prognosis due to their deep anatomical position and proximity to critical neurovascular structures. We report the case of a 12-year-old girl presenting with rapidly progressive craniofacial symptoms. Imaging revealed an aggressive skull-base mass with extensive sinonasal, orbital, and intracranial involvement. Histopathological analysis confirmed alveolar rhabdomyosarcoma. Despite initial response to chemotherapy, the disease progressed with leptomeningeal dissemination. This case highlights the essential role of imaging in early diagnosis, staging, and prognostic assessment.

Keywords: Rhabdomyosarcoma, Parameningeal, Pediatric tumor, MRI, Skull base.

INTRODUCTION

Rhabdomyosarcoma is a malignant tumor arising from primitive mesenchymal cells with skeletal muscle differentiation and represents approximately 5–8% of all pediatric malignancies. The head and neck region is one of the most common sites of involvement and includes orbital, non-parameningeal, and parameningeal locations. Among these, parameningeal rhabdomyosarcoma is associated with the worst prognosis due to its deep location, delayed clinical presentation, and early extension to the skull base and intracranial compartment.

Clinical manifestations are often nonspecific, which may lead to diagnostic delay. Imaging therefore plays a crucial role in the initial detection of the tumor, assessment of its local and intracranial extension, guidance for biopsy, and evaluation of therapeutic response. We report a case of aggressive parameningeal alveolar rhabdomyosarcoma in a child, emphasizing the key radiological findings and their prognostic implications.

CASE REPORT

A 12-year-old girl with no significant past medical history presented with a 20-day history of progressive headaches associated with left-sided exophthalmos. The clinical condition rapidly worsened, with bilateral decrease in visual acuity and deterioration of general status.

On physical examination, a non-axial left exophthalmos was noted, along with bilateral visual impairment. There was no diplopia or motor deficit. Nasal obstruction was present, and no cervical lymphadenopathy was detected. Laboratory investigations revealed an elevated inflammatory marker profile with otherwise unremarkable findings.

Radiological Findings
Computed Tomography

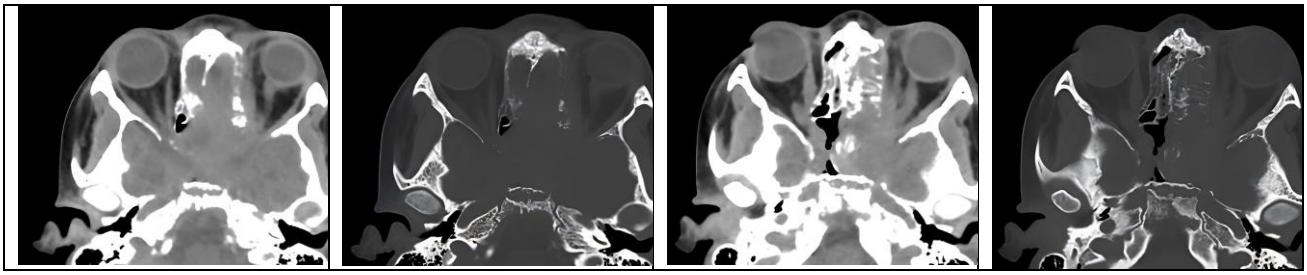

Fig. 1: Computed tomography of the craniofacial region demonstrated a large, aggressive soft tissue mass centered at the skull base. The lesion was associated with extensive bone destruction involving the ethmoid air cells and adjacent structures. The mass extended into both nasal cavities and caused erosion of the lamina papyracea bilaterally.

Magnetic Resonance Imaging

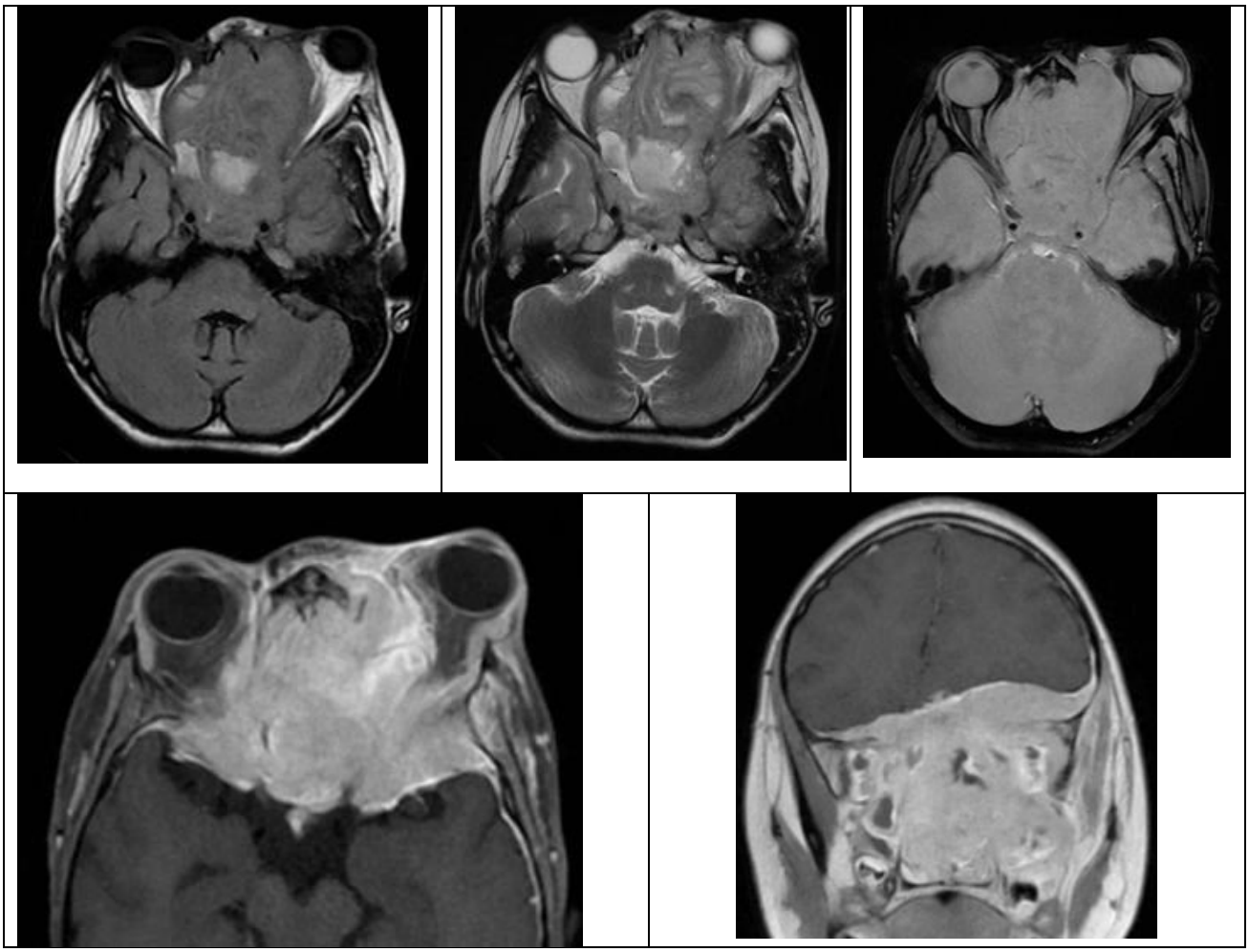

Fig. 2: Magnetic resonance imaging revealed a poorly defined, expansile lesion centered in the anterior skull base. The mass appeared isointense relative to muscle on T1-weighted sequences and heterogeneously hyperintense on T2-weighted images. Following contrast administration, intense and heterogeneous enhancement was observed. Areas of signal void on susceptibility-weighted sequences suggested intratumoral hemorrhage

The tumor extended into both nasal cavities and infiltrated the ethmoid air cells. It eroded the lamina papyracea and extended into both orbits, with more pronounced involvement on the left side. The lesion infiltrated the extraconal fat and was in close proximity to the extraocular muscles and optic nerves. Diffusion-weighted imaging demonstrated high signal intensity without significant restriction.

Histopathological Findings

A surgical biopsy was performed, and histopathological examination revealed a malignant

tumor composed of small round cells arranged in an alveolar pattern. Immunohistochemical analysis confirmed the diagnosis of alveolar rhabdomyosarcoma, with positive staining for desmin and myogenin.

Treatment and Outcome

Initial staging investigations, including thoraco-abdominopelvic imaging, showed no evidence of distant metastasis at the time of diagnosis. Following multidisciplinary discussion, the tumor was considered unresectable, and neoadjuvant chemotherapy was initiated.

The patient initially demonstrated partial radiological response with a reduction in tumor volume. However, after several cycles of treatment, disease progression occurred, with the development of leptomeningeal dissemination seven months after diagnosis.

DISCUSSION

Rhabdomyosarcoma is an aggressive malignancy of childhood, and the alveolar subtype is known to be associated with a poorer prognosis compared with the embryonal form. It typically affects older children and adolescents and demonstrates a higher propensity for metastatic spread.

Parameningeal tumors present a particular diagnostic challenge because of their deep location and nonspecific early symptoms. As a result, they are often diagnosed at an advanced stage, when significant local extension has already occurred. The proximity of these tumors to the skull base facilitates early intracranial spread, which is a major adverse prognostic factor.

Imaging plays a central role in the evaluation of these tumors. Computed tomography is useful for assessing bone destruction, while magnetic resonance imaging provides superior soft tissue contrast and allows precise evaluation of tumor extent, including orbital and intracranial involvement. MRI is also essential for treatment planning and follow-up.

Typical imaging features include an ill-defined, aggressive soft tissue mass with heterogeneous enhancement and associated bone destruction. Orbital invasion and contact with the optic nerves are frequently observed in advanced cases. Diffusion-weighted imaging may show high signal intensity, although restricted diffusion is not always present.

Several factors influence prognosis, including tumor location, histological subtype, and the presence of intracranial extension. Parameningeal location and alveolar histology are both associated with poorer outcomes, particularly when diagnosis is delayed.

CONCLUSION

Parameningeal alveolar rhabdomyosarcoma is a highly aggressive pediatric malignancy with a poor prognosis. Early recognition of imaging features is essential for accurate diagnosis, staging, and management. Radiologists play a key role in identifying patterns of tumor spread and guiding multidisciplinary treatment strategies.

Declarations

Ethical Approval: Not required for a single case report.

Consent: Informed consent was obtained from the patient's guardians.

Conflict of Interest: The authors declare no conflict of interest.

Funding: No funding was received for this study.

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