Case Report

Paediatric Parameningeal Alveolar Rhabdomyosarcoma in the Nasal Cavity

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ABSTRACT

Introduction

Rhabdomyosarcoma is the second most common malignant soft tissue tumor, which is of skeletal muscle in origin. Alveolar subtype of rhabdomyosarcoma is commonly seen in 10-25 years of age. We present a rare case of alveolar rhabdomyosarcoma in a one-year-old child.

Case Report

A one-year-old boy presented with swelling and nasal discharge in the left nasal cavity of two months duration. Examination showed a smooth pink-coloured mass in the left nasal cavity arising from the lateral wall. Fine needle aspiration cytology, biopsy, imaging studies and immunohistochemistry were done and a diagnosis of alveolar rhabdomyosarcoma confirmed. Treatment was done by chemoradiation.

Discussion

Biopsy and immunohistochemistry are the gold standard investigations for rhabdomyosarcoma. It requires a multimodal treatment approach comprising surgery, chemotherapy, and radiotherapy.

Keywords

Rhabdomyosarcoma; Paediatric Malignancy; Para Meningeal; Alveolar; Nasal Cavity

Weber first described Rhabdomyosarcoma (RMS) in 1854. It is primarily a paediatric soft tissue malignancy that derives from mesenchymal cells associated with skeletal muscle differentiation.[1] Until 1946 histologic definition was not available, later Stout recognized the distinct morphology of rhabdomyoblasts.[2] It is a rare tumour with an overall incidence of 0.034 cases per 100000 population.[3] The most common sites of involvement of RMS are the head and neck, genitourinary tract, retroperitoneum, and extremities.[4] Regional lymphatic disease is seen in 8% of the affected patients and metastatic disease is present in 13%, the most common sites being the bone marrow, cerebrospinal fluid, peritoneal fluid, and lung.[5]

These tumours are categorized by the Intergroup Rhabdomyosarcoma Study (IRS) into embryonal, alveolar, pleomorphic, and mixed types. Embryonal type represents the most common in both children and adults, the alveolar type occurs primarily in the teenage and young adult populations.[6] We present a rare case of parameningeal alveolar rhabdomyosarcoma in the nasal cavity presenting in a one-year-old child.

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Case Report

A one-year-old boy presented with swelling and nasal discharge in the left nasal cavity of two months duration. On examination, a smooth pink-colored polypoidal mass was seen in the left nasal cavity possibly from the lateral wall pushing the inferior turbinate and septum to the opposite side. There was fullness in the left cheek area causing facial asymmetry [Figure1]. The left side gingivolabial sulcus was obliterated. No lymph nodes were palpable in the neck region.

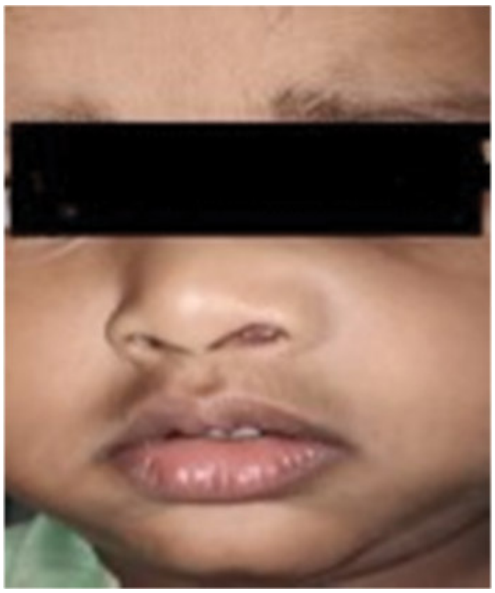

Fig. 1. Mass seen in left nasal cavity and obliteration of left nasofacial fold.

Contrast-enhanced Computed Tomography (CECT) showed a well-defined lobulated lesion of 2.7*2.4*2.5cm in the left nasal cavity showing moderate heterogeneous central and rim enhancement, causing bony erosion of frontal process of the left maxilla, alveolar process of the maxilla at the level of left central and lateral incisors and also the left inferior turbinate. [Figure 2].

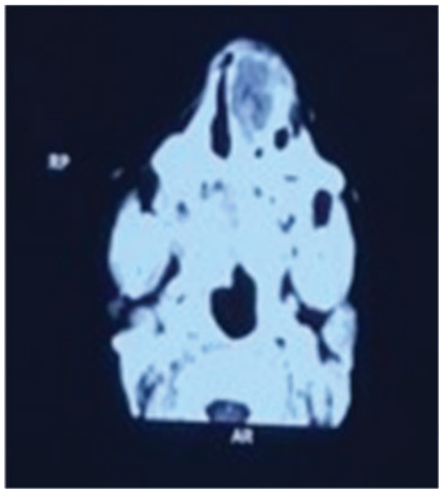

Fig. 2. CECT (axial cut) showed well-defined lobulation in the anterior aspect of the left nasal cavity

Fine needle aspiration cytology (FNAC) of the lesion showed small round cells arranged as clusters with occasional rosette formation [figure 3]. Later an incisional biopsy of the mass was done and it showed sheets of small round blue cells with hyperchromatic nuclei and cytoplasm with varied morphology including nests, trabeculae, and stroma showing fibro myxoid areas giving alveolar pattern [figure 4]. The histopathology report was suggestive of rhabdomyosarcoma.

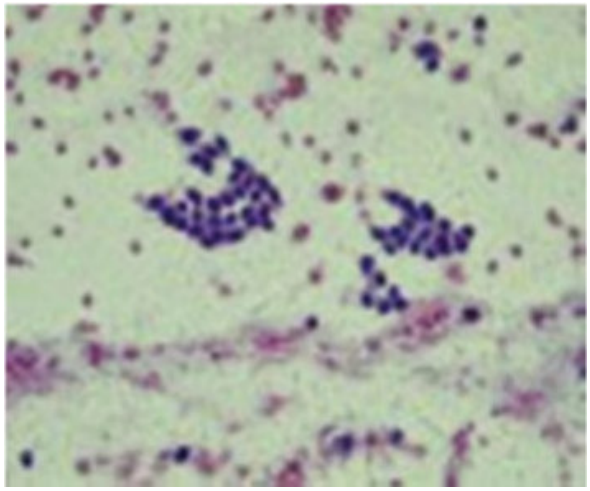

Fig. 3. FNAC using Papanicolaou stain (100X magnification) showed small round cells arranged as clusters with occasional rosette formation.

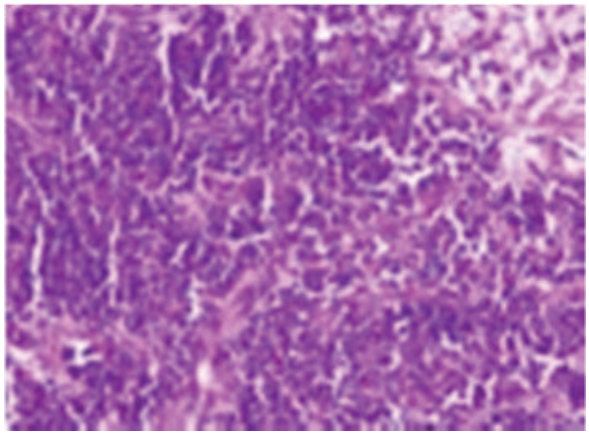

Fig. 4. Histopathological analysis of the hematoxylin and eosin-stained material showed clusters of small round blue cells [100X magnification]

Diagnosis of parameningeal alveolar rhabdomyosarcoma was confirmed with immunohistochemistry (IHC)which was strongly positive for tumor markers like Desmin, Myogenin, and CD99. A metastatic workup done by Technetium-99 Methylene diphosphate bone scan showed no metastasis.

He was referred to the Pediatric Oncology Department, and the proposed treatment plan was a combination of chemotherapy and external beam radiotherapy. Surgery was refused by the parents, even though a multimodal treatment including surgery was advised. There was a considerable reduction in the tumour size after 4 cycles of chemotherapy and radiation therapy with improvement in the general well-being of the child.

Discussion

RMS more commonly occurs in children aged between 1-4 years and 10–14 years.[7] The incidence of RMS is 1.5 times higher in males than females and its incidence has risen over the past 3 decades by an average annual increase of 1.16%.[8]

Within the head and neck region, RMS is divided into three types based on anatomical distribution: orbital, parameningeal and nonorbital. Parameningeal type involves pterygopalatine, infratemporal fossa, paranasal sinuses and middle ear. These are associated with poor prognosis. The alveolar type occurs primarily in the teenage and young adult population with a prognosis worser than embryonal type.[9] Our patient was a one-year-old boy with left parameningeal alveolar type RMS in the nasal cavity which is rare in this age.

Incisional biopsy showed small round cells with scanty cytoplasm, clumped chromatin, arranged as small nests with fibrous septae involving alveolar pattern with a non-distinct histological presentation making it difficult to diagnose the subtypes. This diagnostic dilemma was overcome by IHC testing. So, it's a challenge to diagnose RMS, both clinically and histologically, due to its versatile appearances. IHC has provided diagnostic techniques that add value to the histologic diagnosis of RMS, with anti-desmin staining in 94%, 77% positive for desmin and 78% positive for muscle-specific actin.[8] Here the IHC was strongly positive for Desmin, Myogenin, and CD99, and got a confirmatory diagnosis of alveolar RMS. Most alveolar tumors are strongly positive for myogenin, whereas embryonal tumors often exhibit positivity to a lesser degree, which agrees with the IHC results of the present case.[10,11]

Staging of RMS is commonly based on the IRS classification, which incorporates the extent of disease with metastases and surgical results. It was also recommended by the Intergroup Rhabdomyosarcoma Study Group (IRSG) that staging systems for this disease require ongoing analysis to confirm prognostic correlation with stage. Risk stratification of children with RMS is done utilizing the tumor, node, metastasis (TNM) staging (I to IV) and clinical groupings based on embryonal versus alveolar subtypes. Further they are classified into low-risk (TNM stage I embryonal/clinical stage I–III, TNM stage II–III embryonal/clinical stage I–II), intermediate-risk (TNM stage II–III/clinical stage III, nonmetastatic alveolar) and high-risk (metastatic any subtype). This child is categorized as intermediate risk due to alveolar subtyping in histopathology.

A multidisciplinary approach to the management of RMS with surgery, radiotherapy, and chemotherapy has improved the prognosis in the current medical literature reporting a five-year survival rate of 74–77%.[1,2] Patients with alveolar RMS present with regional and distant metastases and have a higher recurrence rate and poorer survival than patients with the embryonal or botryoid subtype.[12] Infiltration of the skull base and the presence of a residual tumor after primary therapy have also been associated with an unfavorable clinical course.[13] This child got induction chemotherapy with vincristine, actinomycin and cyclophosphamide (VAC regime) followed by radiation therapy. Surgery is indicated to debulk the tumor and should be done when not associated with functional or cosmetic deformity.

Conclusion

Alveolar RMS is unusual in children of age less than 2 years and more common in teenagers. The involvement of parameningeal sites is clinically significant because it

carries the risk of involvement of the skull base and/or intracranial extension, which confers para meningeal RMS a poorer prognosis. Metastasis is a very poor prognostic factor in RMS, decreasing the survival rate from 40% to 5%, so it is of the utmost importance to detect metastasis at the earliest. In this report, we highlight the importance of maintaining a high index of suspicion for uncommon pediatric malignancy in children with unilateral sinus symptoms and the paramount importance of immunohistochemistry in fetching the final diagnosis, which helps in proper management of the tumor. An increased awareness among otorhinolarygologists will help in early detection and timely intervention for this condition.

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