Rhabdomyosarcoma in a pediatric patient: A rare case report

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Abstract

Rhabdomyosarcoma (RMS), a tumor of skeletal muscle origin, is the second most common soft tissue sarcoma encountered in childhood after osteosarcoma. The common sites of occurrence are the head and neck region, genitourinary tract, retroperitoneum, and to a lesser extent, the extremities. Site predilections in the oral cavity are a soft palate, maxillary sinus and alveolus, posterior mandibular region, cheek and lip, and possibly tongue. RMS is a highly malignant tumor with extensive local invasions and early hemorrhagic and lymphatic dissemination. Despite aggressive approaches incorporating surgery, dose-intensive combination chemotherapy, and radiation therapy, the outcome for patients with metastatic disease remains poor. Here, we report a case of oral RMS in a 1-year-old child and describe the clinical, radiological, histopathological, and immunohistochemical findings.

Keywords: Chemotherapy, oral cavity, radiotherapy, rhabdomyosarcoma

Introduction

Rhabdomyosarcoma (RMS) was first described by Weber in 1854, is a malignant soft tissue neoplasm of the skeletal muscle origin.[1] The most common sites of involvement of RMS are head and neck, genitourinary tract, retroperitoneum, and extremities.[2]

Clinically, the manifestations of RMS may vary from a small cutaneous nodule on the face to an extensive fast-growing facial swelling, which may be painless or occasionally associated with pain, trismus, paresthesia, facial palsy, and nasal discharge.[3]

The histogenesis of RMS is still unclear, but the most widely accepted hypothesis is that RMS arises due to the proliferation of embryonic mesenchymal tissue.[4]

There is a slight predilection for disease in males have been reported.[5,6] The survival rate of patients with this tumor ranged from 20% to 35% in reported series.[7]

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The purpose of this article was to report a case of oral RMS with mobile tooth present in upper left posterior maxillary ridge surrounded by large, firm, and tender swelling in a 1-year-old boy and discuss the clinical, radiological, histopathological, and immunohistochemical features.

Case Report

An 1-year-old boy was referred to our institution with painful swelling in his mouth. On history taking, his parents reported that the swelling was present for 6–7 months which was earlier small and painless but now has increased in size up to 5 cm × 6 cm. Initially, the patient was seen by a physician who prescribed antibiotic/anti-inflammatory therapy, however, the treatments were ineffective, and he was, therefore, referred to us. Written informed consent was obtained from her parent for further investigations.

Pain is intermittent which arises on eating and feeding and subsides on taking medication. Clinical examination showed severe facial asymmetry [Figure 1]. A mobile tooth is present in upper left posterior maxillary ridge surrounded by large, firm, and tender swelling. The skin had appeared stretched and inflamed. The opening of the mouth was partly restricted. Intra-oral examination showed a 5 cm × 6 cm, red, firm mass with grayish areas of central necrosis, covering the left side of the maxillary gingiva, from the canine to the second molar region [Figure 2].

Computed tomography confirmed the presence of an extensive infiltrative lesion accompanied by severe bone destruction and displacement of adjacent structures [Figure 3]. An incisional biopsy was made. Histopathological analysis of the hematoxylin and eosin stained material showed clusters of small round cells with hyper chromatic nuclei and eosinophilic cytoplasm separated by fibrovascular septae [Figure 4]. The neoplastic cells were strongly positive for vimentin, desmin, myoglobin, and muscle-specific actin. A diagnosis of oral

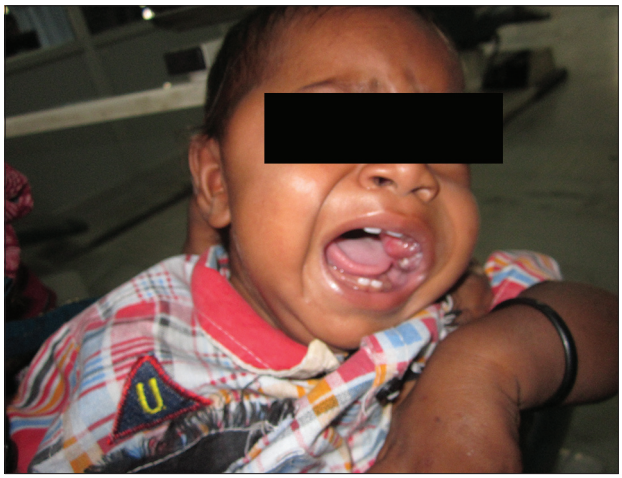

Figure 1: Extraoral photograph showing the lesion

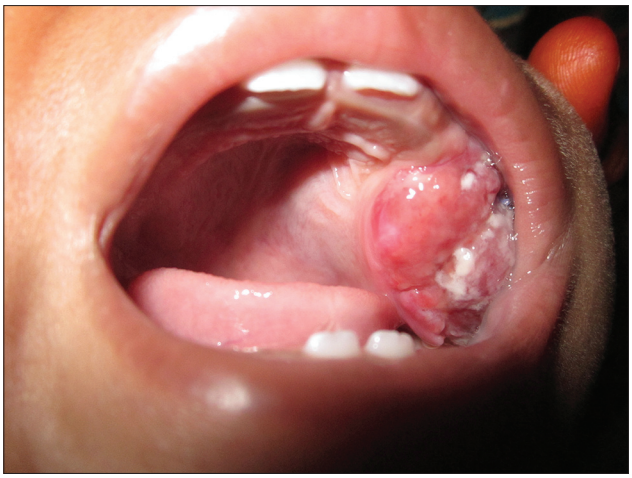

Figure 2: Intraoral aspect showing extensive mass involving the maxillary alveolar mucosa

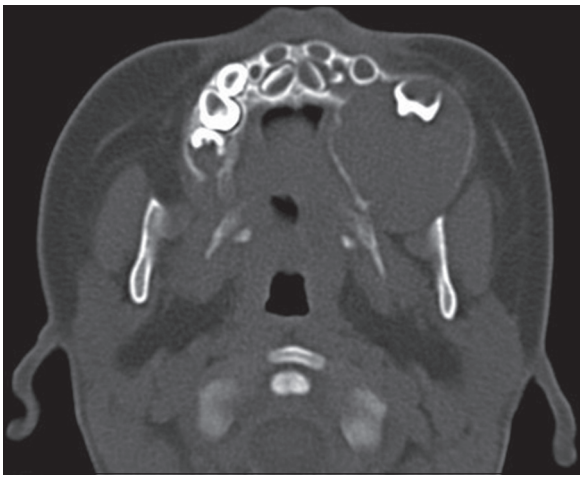

Figure 3: Computed tomography scan showing extensive infiltrative lesion with displacement of adjacent structures

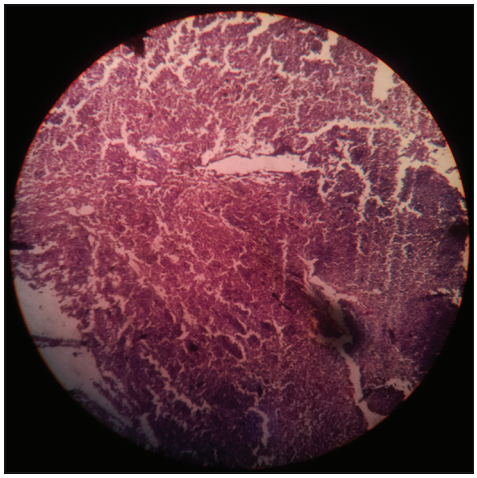

Figure 4: Photomicrograph showing a sheet of mesenchymal cells in a myxoid stroma (H and E, ×200)

RMS was established on the basis of the history, clinical, radiographic and histopathological findings.

After performing the standard diagnostic workup, the tumor was diagnosed as alveolar RMS. He was referred to the Pediatric Oncology Department, and the proposed treatment plan was a combination of chemotherapy, including vincristine, actinomycine, cyclophosphamide, and dexamethasone radiotherapy. The patient's parents refused external beam radiation therapy. Subsequently, patient developed metastatic lesions in bone and lungs. Despite the treatment, the tumor continued to increase in size, and the patient died from lung metastases 6 months after the treatment.

Discussion

The incidence of RMS is the highest in children aged 1–4 years, lower in children aged 10–14 years, and lowest in those aged 15–19 years. Our patient was 1-year-old fall in a very high-risk category. These tumors exhibit a fast and aggressive growth, reaching large dimensions, and are generally painless associated with high rates of recurrence and generalized metastases through the hematogenic and/or lymphatic routes.[8,9]

The head and neck region is the most common site for RMS, with the orbit being the most frequent primary site. The most common site of involvement in the oral cavity is the tongue followed by the soft palate, hard palate, and buccal mucosa.[10] In our patient, the maxillary alveolar ridge and hard palate were involved.

A careful histological examination is required to differentiate such lesions from other more frequent and aggressive lesions affecting the concerned site. In our case, the marked pleomorphism noted was critical for differentiating RMS from Ewing's sarcoma. The presence of an alveolar pattern, pleomorphism, cohesive nature of the cells, and the absence

of lymphadenopathy ruled out the diagnosis of lymphoma. In this respect, the most critical differential diagnosis is neuroblastoma. However, neuroblastoma, another small cell tumor characterized by a diffuse pattern of small round cells and the presence of rosettes/pseudorosettes with pale eosinophilic material is quite similar to the alveolar variant of RMS. The frequently elevated level of urinary catecholamines in neuroblastoma aids in the differential diagnosis.

The differential diagnosis also includes vascular malformations, within which the most common affecting the pediatric airway is the lymphatic or lymphatic-venous malformation.[11]

Prognosis of RMS is relatively poor compared to that of other oral soft tissue malignant lesions and depends on the clinical staging and the anatomic site of the tumor.[12,13]

Unfortunately, in the present case, the lack of cooperation of the patient's guardians and the lack of institution of adjuvant chemotherapy and/or radiotherapy at first admission may have favored the rapid progression of the tumor and subsequently aggravated the severity of the condition, resulting in the child's death.

Patients with RMS may present signs and symptoms such as pain, paresthesia, loss of teeth, and trismus as a result of factors such as advanced tumor stage, infiltrative growth, and tumor location.[14] In the present case, involvement of teeth was observed, and the patient complained of painful symptoms associated with the tumor.

Treatment, therefore, is by a multidisciplinary approach. It consists of surgical removal of the tumor followed by multiagent chemotherapy with or without radiotherapy since RMS tends to metastasize to bone marrow. Bone marrow aspiration should be a part of the staging procedure.[15]

Finally, we conclude that in children, any swelling should be carefully examined, and treatment outcomes should be regularly followed up. High degree of suspicion, early diagnosis, and a multidisciplinary treatment approach would be of great importance in such cases.

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