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**Case Report**

# Embryonal rhabdomyosarcoma of hard palate: a case report and review of literature

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## ABSTRACT

Rhabdomyosarcomas are the most common malignant soft tissue tumors in children and adolescents. Around 40% of cases are diagnosed in head and neck region with orbit and face being the commonly involved areas. Oral lesions account for less than 10%. We report a case of embryonal RMS in a 4-year-old boy who presented with a polypoid mass in the hard palate. Though majority of the oral lesions encountered in children are benign, a small but significant percentage may be representative of an aggressive malignant tumor. Differentiating benign and malignant oral lesions is mandatory for optimal diagnostic strategies.

**Keywords:** Child, Oral cavity, Polypoid swelling, RMS

## INTRODUCTION

Rhabdomyosarcoma (RMS), arising from the primitive mesenchymal tissue is the most common malignant soft tissue tumor in children under 15 years of age. Head and neck (40%), genitourinary tract (25%), and extremities (20%) are the commonly involved sites.[1] Parameningeal and orbit are the most common sites of head and neck region and oral lesions accounts for only 10-12% of cases. Other less commonly involved sites are nasal cavity, nasopharynx, ear, paranasal sinuses.[2] Although exceedingly uncommon, persistent oral masses that are rapidly enlarging, painful, with easy friability and bleeding needs extensive evaluation to rule out malignant tumors.

## CASE REPORT

A 4-year-old boy first born out of non- consanguineous marriage, was found to have insidious onset of a small fleshy swelling in the roof of the mouth. There was no preceding history of trauma. The child was taken to a nearby physician and was reassured and advised observation. Six months later, as the swelling was progressively increasing in size and was associated with minimal intermittent bleeding episodes, he was taken to a dentist and evaluated for the same. Intra oral examination revealed, a well circumscribed lobulated growth in the para median region of the hard and soft palate junction in the posterior part of hard palate. It was non tender verrucous/pedunculated growth of 2×3 cm (Figure 1).

Blood investigations and peripheral smear revealed no abnormalities. A CT paranasal sinus was done, which revealed a well-defined enhancing lobulated lesion of 22×18×3.7 mm at the junction of the hard and soft palate, more towards left side projecting into the oral cavity. The differentials considered were verrucous papilloma, verruca vulgaris and lymphangioma. Child underwent surgical excision with good clinical clearance under general anesthesia by oral and maxillo facial surgeon.

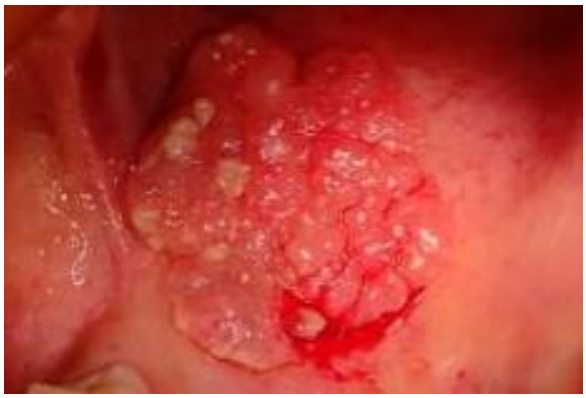

**Figure 1: A well circumscribed lobulated growth in the para median region of the hard and soft palate junction.**

Histopathology showed polypoid tumor lined with stratified squamous surface epithelium with subepithelial zone of loose stroma with condensation of round/spindle shaped tumor cells underneath, exhibiting vesicular nuclei, moderate pleomorphism and abnormal and increased mitoses (Figure 2).

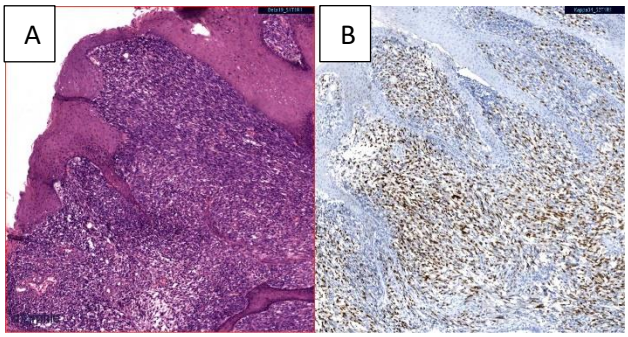


**Figure 2: A) Hematoxylin and B) Eosin 100X squamous epithelium with underlying subepithelium showing sheets of small round cells with scant cytoplasm IHC Myogenin 100X diffuse nuclear positive.**

In few areas, the tumor cells were arranged in irregular fascicular pattern. Immunohistochemistry was positive for desmin, myogenin and Ki 67-60%, and a final diagnosis of embryonal palatal RMS was made. Whole body PET CT done as part of metastatic evaluation did not reveal any residual lesion at the primary site and there was no FDG avid lesions elsewhere. RT PCR for PAX3/PAX7-FKHR translocation was negative. Child was risk stratified as low risk as per Children oncology group (COG) and completed chemotherapy. Child is currently on follow up and doing well.

## DISCUSSION

RMS accounts for 60% of soft tissue sarcomas in children. There is a bimodal distribution with one peak in the first decade of life and the other peak occurring during adolescence. A review of all pediatric rhabdomyosarcoma cases over a 20- year period at Texas Children's Hospital identified 50 head and neck RMS cases. The primary tumor sites identified were; face (18%), orbit/periorbital (16%), nasal cavity (14%), lymph nodes (12%), paranasal sinuses (10%), parameningeal (10%) and palatal (2%).[3] Oral rhabdomyosarcomas usually presents as a rapidly enlarging painless mass and is generally larger than 1 cm at initial presentation. Involvement of adjacent nerves, might cause paresthesia and pain. The tongue is the most common site involved in oral rhabdomyosarcomas, followed by palate and buccal mucosa.[3] Palatal lesions most commonly involve the soft palate and uvula in few cases. Congenital RMS of lip and tongue has been reported but lesions of floor of the mouth are extremely rare.[4-6] The differentials of soft palate masses in children are benign and malignant salivary gland tumors, hemangiomas, granular cell tumor, hematolymphoid tumors and metastatic lesions.[7] Histologically, RMS is classified into embryonal, alveolar, botryoid and pleomorphic subtypes. The embryonal type accounts for most cases and occurs mainly in the genitourinary tract and head and neck. It contains immature striated muscle-like cells (rhabdomyoblasts). The alveolar type typically presents in the extremities and is characterized by clusters of small round cells separated by fibrovascular septae. Embryonal type is more common in younger children and alveolar type in adolescents. Though the botryoid type has the classic polypoid appearance, due to its relative rarity and non-specific presentation, diagnostic delays are common. Translocations t (2;13) (q35; q14) or t (1; 13) (p36; q14) are the common cytogenetic abnormalities reported in alveolar RMS.[8] Loss of heterozygosity or loss of imprinting at a specific locus on the short arm of chromosome 11 (11p15) has been identified in embryonal RMS.[9] The treatment involves a multidisciplinary approach involving chemotherapy, surgery and radiation therapy. Prognosis depends on the age, site of lesion, clinical staging and histopathology type. Orbital and periorbital tumors have a 5-year survival rates of 95%.[3] Other favourable factors include gross total excision of tumor, smaller tumor size at presentation (<5 cms) younger age (<10 years) and embryonal histology. The primary location of the disease significantly influences the outcomes. According to the site of involvement, head and neck RMS can be divided into three subtypes: parameningeal (nasal cavity, paranasal sinuses, mastoid area, and infratemporal fossa), non-parameningeal (oral cavity, oropharynx, face, cheek, parotid region, and soft

tissues of the neck) and orbital. Parameningeal areas have poor prognosis than non-parameningeal cases due to the impossibility of obtaining a complete resection of the lesion, due to their proximity of the intracranial area.[10]

## CONCLUSION

Though oral cavity RMS is rare in pediatric population, they should be differentiated from the common benign/reactive mesenchymal lesions and periodontal inflammatory lesions to plan optimal diagnostic strategies and there is a need for multidisciplinary approach in case of proven malignancy.

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