Swelling on the Upper Lip: A Diagnostic Challenge

Sir,

Rhabdomyosarcoma (RMS) is a malignant mesenchymal neoplasm that comprises of skeletal muscle cells with varying degrees of differentiation. In the head and neck, the frequently affected sites are orbit, and paranasal sinuses. Oral RMSs are rare, and more frequent in the soft palate.1 The differential diagnoses for RMS includes other small round cell tumors like lymphoma, neuroblastoma, Ewing's sarcoma, and poorly differentiated malignant melanoma. An 18-month boy was brought with a complaint of rapidly growing swelling over upper lip for 15-20 days. On clinical examination, a firm nodular swelling was present on the right lateral aspect of the upper lip. The swelling was approximately 1.5 x 1 cm in size, mildly tender with normal mucosal color and smooth surface. A provisional diagnosis of minor salivary gland pathology was established, and excisional biopsy was performed (Figures 1a-c). The specimen was submitted for histopathological examination, which revealed a submucosal cellular tumor divided by fibrous septae. The central cells of the aggregates were discohesive, giving an alveolar pattern. The cells were monotonous, small, round, with scanty faint eosinophilic cytoplasm; and hyperchromatic nuclei. A diagnosis of small round cell tumor was established. A second opinion was sought from Kidwai Memorial Institute of Oncology (KMIO), Bangalore, Karnataka, India.

Immunohisto-chemical panel was performed by KMIO, which was positive for desmin and myogenin (Figures 2a-b) and negative for CD 99, cytokeratin, S-100 and common leukocyte common antigen (LCA). They confirmed the diagnosis of alveolar RMS. A stimulus for such a sarcomatous proliferation in RMS is unknown but genetic etiology is widely accepted. RMS is the most common soft tissue sarcoma, accounting for 6% of all the malignancies in children under 15 years, accounting for 10 - 12% of head and neck tumors. These are characterised by rapid painless growth which are usually detected at an advanced stage of evolution.2-5 In the present case, the presenting age of the tumor (18 months) was unusual for the diagnosis of alveolar RMS. Embryonal RMS is common in the age group below 10 years and alveolar RMS is common in 15-20 years age group.4 Due to its clinical resemblance, RMS can be mistaken for mucocele or as any other salivary gland pathology by the general dental practitioner. It is recommended that tissue be submitted for histological evaluation and second opinion to be sought from general or oncopathologist, in case ancillary techniques such as immunohistochemistry are required.

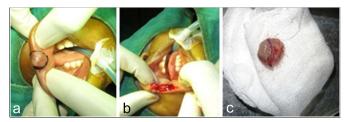


Figure 1: Clinical photograph showing a swelling on the upper lip, operated specimen.

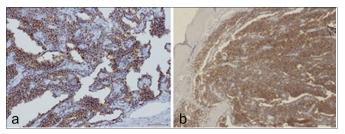


Figure 2: Photomicrograph showing desmin and myogenin positivity of the

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Anand S Tegginamani¹, Vanishree HS², Sonalika Ghate Wanjari³ and Gunjan Dubey4

- ¹ Department of Oral Pathology / Pediatric Dentistry², Faculty of Dentistry, SEGi University Kota Damansara Selangor,
- ³ Department of Oral Pathology / Oral Surgery⁴, Hitkarini Dental College and Hospital, Jabalpur, India

Correspondence: Dr. Anand S. Tegginamani, Department of Oral Pathology, Faculty of Dentistry, SEGi University Kota Damansara Selangor, Malaysia

E-mail: anandop2005@rediffmail.com

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