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Rhabdomyosarcoma Of Oral Cavity – A Case Report With Imaging Perspective

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Abstract:

Rhabdomyosarcomas are among the most common soft tissue tumours in children. These tumours are derived from mesenchymal tissue with a tendency toward myogenic differentiation that probably originates from immature and highly invasive satellite cells associated with the embryogenesis of skeletal muscle. Head and neck is the principal location of RMS - 36% of these tumours. However, relatively uncommon in the oral cavity, with involvement of the jaws being extremely rare.Some of these tumours are associated with high rates of recurrence and metastasis. Hence an early diagnosis and proper treatment plan will be crucial for saving the life of these children. Here a case of intraoral RMS is discussed with emphasis on imaging features.

Introduction:

Rhabdomyosarcoma was first described by Weber in 1854 and they originate from the striated muscle and the common sites include extremities, head and neck, genitourinary tract and retroperitoneum. Rhabdomyosarcoma (RMS) arises from immature mesenchymal precursor cells committed to skeletal muscle lineage or embryonal muscular tissue origin displaced during early development. WHO defined RMS as a highly malignant tumor of rhabdomyoblasts in varying stages of differentiation with or without cross striation.[1, 2]

RMS commonly affects children below 7 years of age. Some authors found bimodal age distribution, the first peak in children aged 2 to 6 years and the second peak in adolescents. Head and neck is the principal location of RMS and accounts for 36% of these tumors. Based upon the primary site of involvement of tumor, RMS of head and neck has been subdivided into the parameningeal and non-parameningeal types. The tumors of oral cavity are included under non-orbital and non-parameningeal group, which accounts for 28% of head and neck RMSs.[3,4]

Case report:

A 10 year girl patient reported to the department with swelling & pain on right side of face since 7 months. Pain is severe, radiating to ear, forehead & neck on right side, increasing while speaking and taking food. Swelling initially started in relation to distal aspect of 47, increased in size intraorally and 3 months later extended extra orally and attained present size. Paraesthesia of lower lip on right side observed since 3 months and restricted mouth opening is seen.

Extra oral examination (FIGURE-1) revealed facial asymmetry with 5x6 cm swelling on right side of face extending anteroposteriorly from angle of mouth to 1cm crossing pinna of ear, superioinferiorly from zygomatic prominence to crossing inferior border of mandible with ill defined borders. Overlying skin is stretched and not pinchable over the swelling. Temperature is raised, tender and consistency is soft to firm and swelling is fixed to underlying structures.

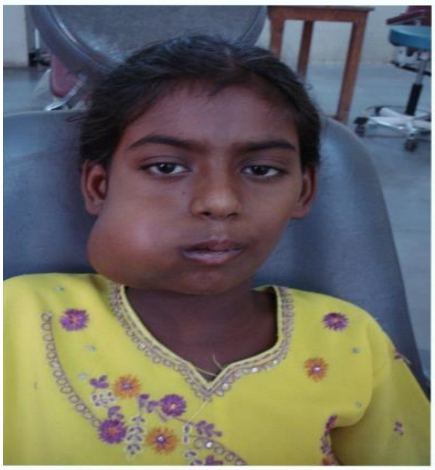
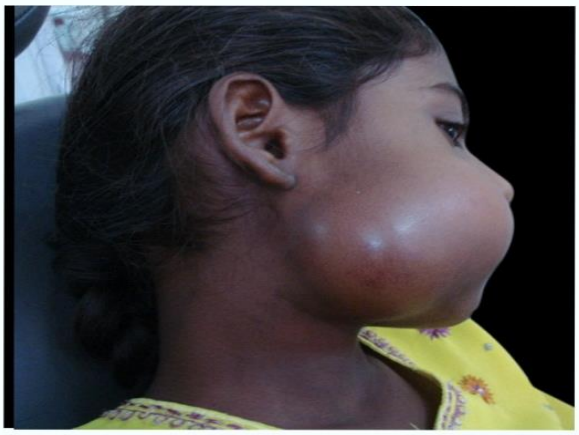



FIGURE 1- EXTRAORAL VIEW SHOWING LARGE SWELLING OVER RIGHT SIDE OF ANGLE OF MANDIBLE

Intraorally, (FIGURE-2) 8x6 cm swelling in right buccal vestibule from angle of mouth to pterygomandibular raphe and superioinferiorly from upper buccal sulcus to lower buccal sulcus with ill defined edges and inflamed surrounding tissue. Swelling is completely occupying the vestibular space, coming in between maxillary and mandibular teeth with surface ulceration and interfering with occlusion with an interincisal distance of 1.8 cms. Swelling is tender, having soft to firm consistency, fixed to underlying structures.

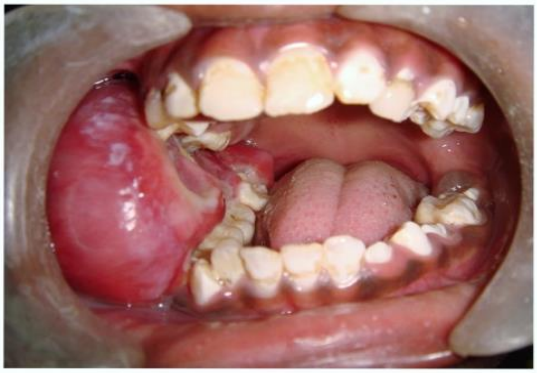


FIGURE 2- INTRAORAL VIEW SHOWING SOFT TISSUE MASS IN BUCCAL SULCUS

Based on the above findings a provisional diagnosis of Ewings sarcoma of mandible has been given with a list of differential diagnosis of Osteogenic sarcoma, Chodrosarcoma, Burkitts lymphoma, Fibrosarcoma, Ameloblasticfibrodentinoma and Rhabdomyosarcoma.

On radiological investigation OPG (FIGURE-3) showed mixed radiolucent- radiopaque lesion irt 46, 47 and 48 and CT scan (FIGURE-4) showed expansile osteolytic lesion involving ramus and body of mandible on right side embedding 46 and 47 within the lesion with cortical destruction buccally & lingually with associated soft tissue mass showing some bony spicules, with well circumscribed hypodense lesion within the mass suggestive of hemorrhage.

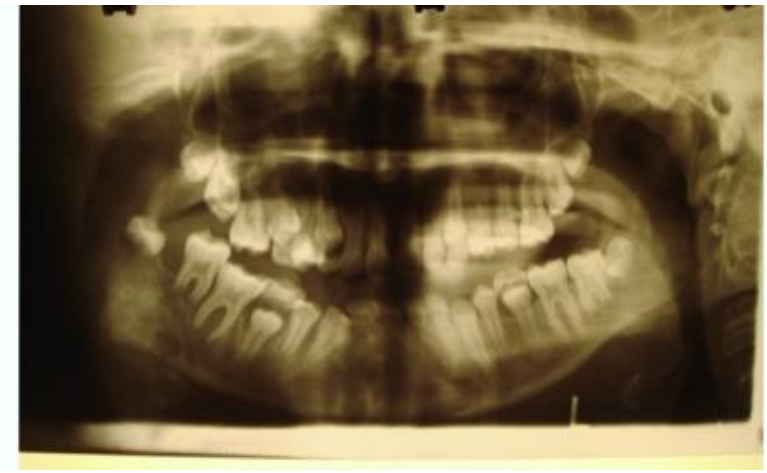
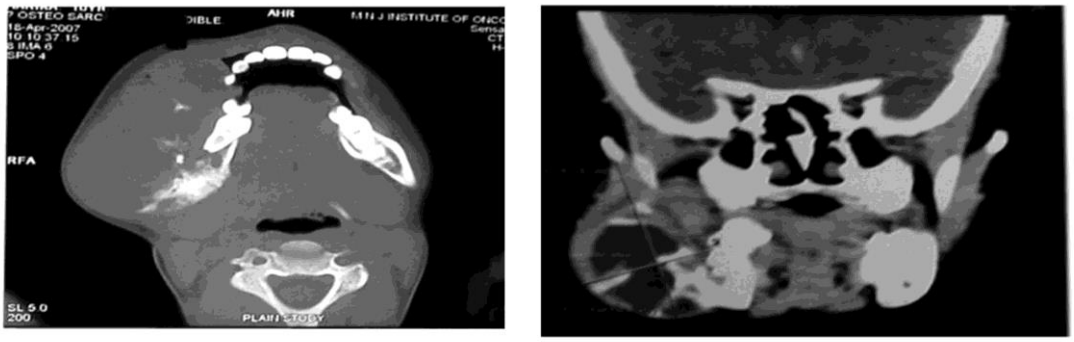


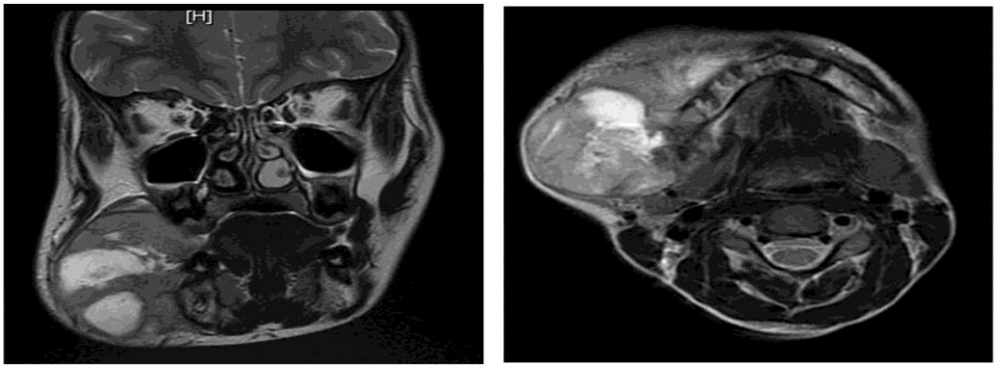
FIGURE 3- OPG SHOWING MIXED RADIOOPAQUE & RADIOLUCNET LESION



CT AXIAL AND CORONAL IMAGES - EXPANSILE OSTEOLYTIC LESION INVOLVING RAMUS & BODY OF MANDIBLE ON RIGHT SIDE, EMBEDDING 46 & 47 WITH ASSOCIATED SOFT TISSUE MASS SHOWING BONY SPICULES

FIGURE 4- CT SCAN

Magnetic resonance imaging (FIGURE-5) shows evidence of large soft tissue heterogenous intensity mass lesion over the right mandibular region, measuring 73x43 mm with hypo and hyperintense foci within the centre of the lesion having heterogeneously hyperintense on T2 & FLAIR sequences with multiple speculated hypointense foci in all sequences, suggestive of calcification. Bony sclerosis is present. There is large area of hyperintense signal within the lesion in T1 & T2W1 suggestive of hemorrhage. The lesion shows erosion of mandible with extenson into oral cavity invading the mucosa. Adjacent musculature is displaced and adjacent neurovascular bundle is stretched over the mass (trigeminal and facial). The lesion is encasing the premolar. No lymphadenopathy. Neck vessels are normal. Submandibular soft tissues are normal suggestive of malignant bone lesion – Ewing's sarcoma.



MRI T2 CORONAL AND AXIAL IMAGES - LARGE SOFT TISSUE HETEROGENOUS INTENSITYMASS LESION OVER THE RIGHT SIDE OF MANDIBLE , MEDIALLY EXTENDING INTO ORAL CAVITY

FIGURE 5- MRI IMAGES

Histopathology section shows stratified squamous epithelium with underlying sub epithelial tissue. A small fragment of tissue showing tumour cells with pleomorphic, hyper chromatic nuclei & scanty cytoplasm, suggestive of small round cell tumour, possibility of embryonal rhabdomyosarcoma..

Discussion:

Rhabdomyosarcoma (RMS) is a malignant neoplasm composed of neoplastic mesenchymal cells with varying degrees of striated muscle differentiation. It is the most common sarcoma in children, accounting for 4–8% of all cases of malignant disease less than 15 years of age.1 there is a pronounced predilection of RMS to children, most commonly affecting males.[6]

Head and neck RMS has a distinct prognosis and biologic behaviour. RMS has four distinct histopathological subtypes: embryonal, alveolar, pleomorphic and undifferentiated. The embryonal subtype is the most common subtype, tends to occur in young patients and frequently arises in the perioral region.7

In general, most patients have an advanced disease even at the stage of initial presentation because RMSs are known to show rapid growth and the patients generally tend to delay medical consultation. In our case, when the patient presented at our clinic, she had severe facial asymmetry and experienced difficulty in opening the mouth. In cases of RMS, radiographic examination reveals the size of the lesion, space relation, and the extent of bone destruction, while MRI reveals the affected areas such as the mandible, condyle, maxillary sinus, and infra-orbitary space.[8]

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. A diffuse pattern of small round cells and the presence of rosettes/pseudorosettes with pale eosinophilic material seen in cases of neuroblastoma may suggest alveolar RMS. High level of urinary catecholamines observed in neuroblastoma is critical to rule out this tumor.[9, 10]

CT gave valuable information about bony spicules or calcification within the lesion and bone destruction. MRI clearly depicted origin, contents, muscle displacement and neurovascular bundle involvement. CT and MRI not only provide essential information about the deep extension of clinically detected masses, they can also delineate additional clinically unsuspected lesions, by providing cross-sectional imaging. The combination of soft-tissue characterization and anatomical localization afforded by CT and MRI allows radiologists to make a substantial contribution to the preoperative assessment of the patient with a mass and appropriate treatment plan by the surgeon.[11] In rhabdomyosarcoma, CT gave valuable information about bony spicules or calcification within the lesion and bone destruction. MRI clearly depicted origin, contents, muscle displacement and neurovascular bundle involvement.

A.Hagiwara, Y.Inoue, T. Nakayama et al in 2001 investigated nine patients with rhabdomyosarcoma in the head and neck (6- 53 years of age) using CT and MRI. The histological subtype was embryonal in five, alveolar in three and pleomorphic in one case. The tumors enhanced markedly and heterogenous on CT and MRI. The masses were isointense or gave slightly higher signal than surrounding muscles on T1- and heterogeneously high signal on T2-weighted images. In four tumors multiple ring enhancement resembling bunches of grapes was seen. This appears to be characteristic of rhabdomyosarcoma and probably reflects a component of botryoid- type rhabdomyosarcoma in which mucoid rich stroma is covered with a thin layer of tumor cells.[12]

Rhabdomyosarcomas appear as muscle density masses on CT and as muscle signal intensity masses on MRI, although they often have higher T2 – weighted signal intensity than normal muscle. These tumors tend to infiltrate the surrounding structures. They may exhibit a variable amount of enhancement following contrast administration.[13]

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. An early and accurate diagnosis of the tumor and a combined therapeutic approach involving surgery, chemotherapy, and radiation therapy are known to dramatically improve the survival rates, as seen in cases recorded over the past 20 years. Effective surgical excision is challenging in cases of RMS of the head or neck region owing to the involvement of other crucial structures in these locations.[14]

CONCLUSION

Dental practitioners frequently encounter cases of children with facial swelling and pain; in most of these cases, dental abscess is considered as the cause and treatment is prescribed accordingly. Taken together, 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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