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CONGENITAL RHABDOMYOSARCOMA OF TONGUE IN A NEWBORN

Rhabdomyosarcoma, though the most common sarcoma in paediatric population rarely involves the tongue. Its treatment involves triple therapy i.e. surgery, radiotherapy and chemotherapy. We present a case of rhabdomyosarcoma on the tongue in a newborn and discuss its clinical presentation and management.

Keywords: rhabdomyosarcoma,tongue, triple therapy

INTRODUCTION:

Rhabdomyosarcoma (RMS) is the most common sarcoma in paediatric population accounting for up to 60%, the common site being the head and neck region which ranges from 35% to 40%.[1,2] Though 25% of head and neck RMS arises from non-orbital, non-parameningeal sites i.e. mouth, neck, face, scalp, larynx,[3] its origin in the tongue is very rare. In the large series of the Intergroup Rhabdomyosarcoma Studies (IRS) I,II and III, only seven cases of RMS of the tongue were documented.[4] We present a case of congenital rhabdomyosarcoma of the tongue.

CASE REPORT

A full term newborn child weighing 3.7 kg, delivered by vacuum delivery for poor maternal effort, with no respiratory difficulty post natally was noted to have lobulated, firm, tongue masses, measuring 5X5 and 5X3 cm, which were smooth, sessile, non tender,non compressible, non reducible, arising from the dorsal surface of anterior 2/3rd and left border of tongue. The tip and the right border of the tongue were spared (Fig1). Ultrasonography with doppler showed heterogeneously echoic vascular mass in tongue with multiple thin parallel linear trabeculations within. Fine needle aspiration was inconclusive. Excisional biopsy with primary repair was done (Fig 2). Grossly the specimen was reddish in color and on cut section, muscle fibres traversing the mass could be seen. Microscopic examination revealed predominantly spindle shaped tumour cells with moderate amount of eosinophilic cytoplasm with central to eccentrically placed hyperchromatic nucleus. Some of the cells showed cytoplasmic extension, forming strap like configuration. Extensive myxoid change was present in the stroma. The cells were infiltrating the skeletal muscle of the tongue (Fig 3,4). These features were suggestive of embroynal rhabdomyosarcoma-botryoid variant. The patient was then considered for triple therapy and so was referred to a medical oncologist.

Fig. 1: Showing lobulated firm mass arising from from dorsal surface of anterior 2/3rd of tongue and left border of tongue.

Fig. 2: Showing post excision and primary repair of the tongue mass.

Fig. 3: Spindle cells seen infiltrating skeletal muscles. Magnification:20x H&E stain

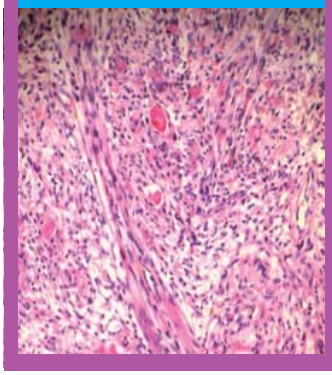
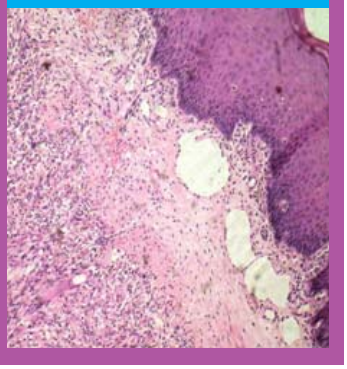


Fig. 4: Botryoid type embryonal rhabdomyosarcoma with the characteristic cambium layer ,and submucosal zone of markedly increased cellularity. Magnification:20x H&E stain.



DISCUSSION:

RMS is a malignant tumor of mesenchymal origin and is categorized as a small blue round cell tumor of childhood.[2] Though it is commonly described as a tumor of muscle origin, it is not derived from mature skeletal muscle and most probably arises from primitive or undifferentiated tissue with capacity for rhabdomyoblastic differentiation.[5]

World Health Organization classification of soft tissue tumors in 1969, has divided RMS into four histological subtypes: embryonal, alveolar, pleomorphic and botryoid.[5] Botryoid is a variant of embryonal subtype.[2,3] Embryonal subtype carries a good prognosis while alveolar carries a poor prognosis.[3]

According to site of origin, the head and neck RMS has been categorized as orbital, non-orbital parameningeal (nasopharyngeal, paranasal sinus, middle ear/mastoid, pterygoid-infratemporal fossa) and non-orbital non-parameningeal sites (mouth, neck, face, scalp, larynx). Oral RMS falls within the non-orbital non-parameningeal group of tumors, which bears a better prognosis and has less tendency to invade the central nervous system. The five year survival rate is approximately 85% for this type of RMS subtype.[2] Oral RMS can cause dysphagia, dyspnea, cough and respiratory obstruction. Trauma to the tumor from sucking or swallowing may cause bleeding. The tumour expands and infiltrates the muscle from which it arises and can initially present as nodule or polypoidal lesion with soft or gummy consistency.[5] In our case, the child had difficulty in feeding so was fed using a nasogastric tube but the child did not have respiratory distress. Oral RMS can be mistaken for hemangioma, when the overlying surface is erythematous and exhibits telangiectasia. However, RMS is more firm, anchored deeply and so immobile.[6] Other differential diagnosis of tongue lesions includes teratoma, thyroglossal duct cyst, lingual thyroid, lymphangioma, hemangioma, dermoid cyst, granular cell myoblastoma and heterotopic gastric mucosal cyst.[7]

Magnetic resonance imaging (MRI) helps in differentiating the various tongue lesions. It gives information regarding the lesion, its relationship with the surrounding soft tissue structure.[6] RMS in MRI appear as large heterogenous mass with a variable amount of associated necrosis.[8]

Treatment of RMS requires multimodal therapy, with the rationale that such patients are presumed to have micrometastasis at diagnosis.[9] The multimodal therapy includes judicious surgery, multiagent chemotherapy and radiotherapy.[3] Surgery is critical in patients with resectable RMS. Complete removal of tumor wards off the necessity of radiotherapy and thus avoids its long term complications hence improving the clinical outcome.[2] In unresectable tumor, role of surgery will be for biopsy purpose or debulking.[1]

The IRSC grouping classification has four surgical±pathologic grouping system, which categorizes patients according to the extent of disease remaining after the initial surgical procedure(s) but before beginning chemotherapy and radiation therapy.[2]

Table 1: Intergroup Rhabdomyosarcoma Study Committee classification System

IRSC Group	Definition
I	Localized disease, complete resection
II	Microscopic residual disease
III	Gross residual disease
IV	Distant metastasis

All patients with RMS receive chemotherapy as it is chemosensitive. The chemotherapeutic agents consist of vincristine, actinomycin D, cyclophosphamide, etoposide, ifosfamide, doxorubicin, cispatin and carboplatin. The intensity of treatment depends on the stage of disease, site and the histology of the disease.[3] The standard treatment regimen consisting of vincristine,actinomycin-D and cyclophosphamide (VAC).[9] The use of radiotherapy is to achieve control of local and regional disease. With modern multidisciplinary therapy, cure rates have improved significantly in the past three decades, at least 70% children and adolescents of RMS are cured.[9]

The literature shows various treatment modalities being tried for RMS of tongue i.e. combined brachytherapy and chemotherapy without surgery, only surgery, chemotherapy with surgery and triple therapy consisting of chemotherapy, surgery and radiotherapy depending on the size of the tongue lesion, the morbidity anticipated due to radical surgery (total glossectomy), the response to surgery and chemotherapy.[2,5]

CONCLUSION:

Rhabdomyosarcoma of tongue is a rare condition.Treatment involves triple therapy consisting of surgical excision, radiotherapy and chemotherapy.

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