



Case Report

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Oral Rhabdomyoma in a newborn male presenting as difficulty in feeding: A very rare case report.

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Abstract

Rhabdomyoma, by definition is a benign muscle tumour. Rhabdomyomas constitute 2% of all myogenous neoplasms. This tumour is in incongruence with other benign soft tissue tumours, in that it is rarer than its malignant counterpart. The peak incidence of rhabdomyosarcoma in children between 0 years and 4 years is 4 cases per 1million children. They are broadly categorised as cardiac and extra-cardiac. Three different subtypes exists as 1) the adult type, 2) the fetal type and 3) the genital type, the adult type being the most common.[1] AR (Adult Rhabdomyoma) generally occurs in the 4th and 5th decade with a male predilection.[2] There have been very few presentations of this lesion in the paediatric age group. Here we present a case of congenital lingual fetal rhabdomyoma in a newborn baby boy presenting as difficulty to feed since birth.

Keywords: fetal rhabdomyoma, muscle tumor, tongue.

Introduction

Rhabdomyoma, by definition, is a benign muscle tumor. Rhabdomyomas constitute 2% of all myogenous neoplasms. This tumor is in incongruence with other benign soft tissue tumors, in that it is rarer than its malignant counterpart. They are broadly categorized as cardiac and extracardiac. Three different subtypes exist as: (1) the adult type, (2) the fetal type, and

(3) the genital type, the adult type being the most common.[1]Solitary tongue lesions are a diagnostic challenge as they have a wide range of differential diagnosis. The diagnosis of rhabdomyoma is further difficult as it is mainly dependant on its characteristic histopathological and immunohistochemistry findings. The purpose of this article is to present our findings of the case with an appraisal of literature on the incidence of such cases in children.

Case Report

A newborn baby was referred to the emergency/casualty of TMC as a case of oral mass obstructing the foodpipe. The baby was admitted in SNCU of TMC and pediatric surgery consultation was taken. The baby was initially stabilised, thoroughly investigated and was planned for excision biopsy. Endotracheal intubation was really a challenging task for the anaesthesiologist after induction, as the oral aperture was very much restricted by b/l huge sublingual swelling. After endotracheal intubation, under general anaesthesia, the baby was painted and draped. The tongue was exposed and pulled out of the oral cavity by two stay sutures on each side. B/L sublingual swellings were meticulously dissected and excised safeguarding the lingual

nerve one after the other. Proper hemostasis was maintained and the surgical wound on each undersurface side of the tongue was repaired. The baby was given orogastric feed (expressed breast milk) on post operative day 2. The baby was on full oral breast feed from post operative day 4. The baby was discharged successfully on post operative day 6. The excisional biopsy shows an unencapsulated tumour mass with peripheral skeletal component as bands and interlacing fascicles. The tumour cells are bland, plump, spindle shaped with rounded poles. Cytoplasm is eosinophilic and moderate. Mitotic activity is very low. Immunohistochemistry was done & it showed the spindle cells were positive for DESMIN, suggesting it to be a case of fetal rhabdomyoma.



Fig1- B/L sublingual swelling



Fig 2- intra operative picture



Fig 3-HPE showing DESMIN positive diffuse

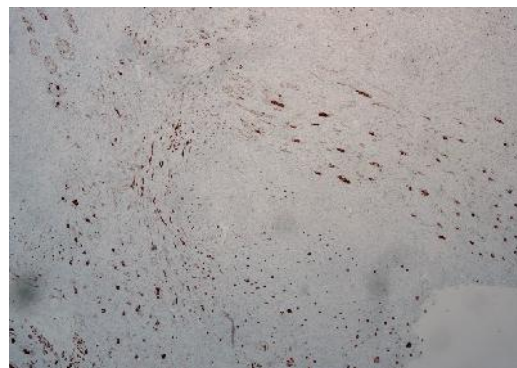


Fig4- HPE showing smooth muscle actin focal positive

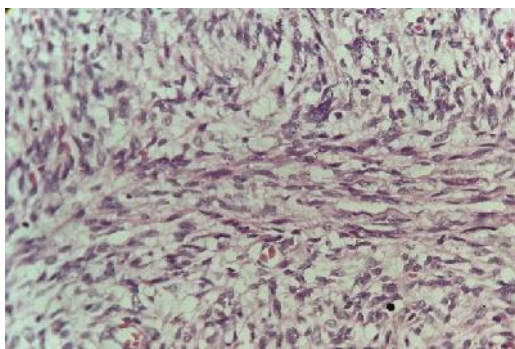


Fig 5: HPE showing H&Estain(10X and 40X)

Discussion

Extra-cardiac rhabdomyomas are extremely rare benign tumor of the skeletal muscle. The fetal type of rhabdomyoma usually presents at or below 3 years of age. AR mostly presents in the adult age group of 40 years and above. The occurrence is mainly in the head-and-neck region particularly floor of the mouth, soft palate, tongue, and buccal mucosa. The tumor is postulated to originate from the muscle component of the third and fourth brachial arches. Its cellular genesis is not definite but is most likely thought to be from primitive mesenchymal stem cells that undergo striated muscle differentiation.[2] There are very few cases of solitary lingual ARs of childhood reported in the literature.[3-18] A comprehensive search in PubMed/MEDLINE database was done using

MeSH terms such as “Adult Rhabdomyoma,” “Muscle tumor,” “Tongue,” “Benign” using various Boolean operators such as “AND” and “OR”. To the author’s knowledge, till date, 18 cases of AR have been found in the younger age group. Only 11 cases of this tumor have been described in infants and children up to 16 years reported in literature by Cacciari in 2001.[3] The age group in our review ranged from 0 weeks to 16 years. The most common site of involvement is the head-and-neck region. Seven out of 18 cases occurred in the tongue showing a predilection toward the oral cavity. Male preponderance can be seen with 13 out of 17 cases occurring in males. They were all solitary lesions. Most were asymptomatic and were treated by simple excision [Table 1].

Table 1: Summary of cases of adult rhabdomyoma in children

Author	Age/sex	Location
Pai <i>et al.</i> ^[5]	8/male	Esophagus
Firdevs <i>et al.</i> ^[6]	8/female	Tongue
Solomon and Tolete-Velcek ^[7]	11/male	Tongue
Huang <i>et al.</i> ^[8]	4/female	Tongue
Cacciari <i>et al.</i> ^[3]	9/male	Mediastinum
Corio and Lewis ^[9]	13/male	Floor of mouth
Shapiro <i>et al.</i> ^[10]	22.5 months/male	Parapharyngeal
Pendl <i>et al.</i> ^[10]	8 weeks/male	Tongue
Rutz <i>et al.</i> ^[10]	5 months/male	Tongue
Nicory <i>et al.</i> ^[11]	5/female	Uvula
Pownell <i>et al.</i> ^[12]	15 months/male	Cricopharynx
Knowles and Jakobiec ^[13]	8/male	Orbit
Willis <i>et al.</i> ^[14]	12/male	Finger
Reitter ^[15]	11/female	Nose
De and Tribedi ^[16]	13/male	Axilla
	8/female	Labia majora
Kleinsasser and Glanz ^[17]	16/male	Larynx
Zwick <i>et al.</i> ^[18]	29 months/male	Intracranial
Present case	11/female	Tongue

In this case, the child presented as a mass in the tongue causing difficulty while feeding. It presented as bilateral huge mass on the lateral surface of the tongue. Clinically, this tumor is naïve in behavior unless its large size becomes a problem. This usually occurs in multifocal type. There have been reports of dyspnea due to the tumor.[2] They are solitary lesions but have been multifocal in 15% of cases. Recognition of the tumor is demanding as the diagnosis is based on its distinctive histopathological appearance. Its presentation as a swelling of lateral border of the tongue should raise a high degree of suspicion. Its differential diagnosis includes granular cell tumor, hibernoma, reticulohistiocytoma, lymphoma, and most importantly rhabdomyosarcoma as the latter being more common. The most common site of rhabdomyosarcoma is in the peripheral skeletal musculature, and histologically, they display considerable polymorphism with atypical mitoses. Macroscopically, it presents as a well-defined, rounded, unencapsulated intramuscular mass that shows characteristic texture and color of muscle.

Histopathologically, it presents as striated muscle cells in various stages of differentiation and maturity.[1] It is composed of tightly packed, large, round, ovoid, or polygonal cell. A large number of which are vacuolated.[4] Occasionally, the granular appearance can lead to a misdiagnosis of granular cell tumor. Positive staining with desmin and myoglobin demonstrates immunochemically that the tumor cells are derived from muscle tissue[1] [Figure 3]. Negative staining with S-100 further substantiates exclusion of granular cellular tumor. No reports of anaplastic changes have been seen in any case.[3] No spontaneous regression of the lesion is seen. Recurrences reported in literature were after a period of 1 month to 35 years with overall frequency of 16% and ascribed to incomplete removal of the tumor. The distinction between rhabdomyoma and malignant neoplasms, i.e. rhabdomyosarcoma, is of great significance to avoid aggressive excision and is not always very easy. A residual tumor may not only be a source of benign recurrences but also a source of cells with malignant potential.[5]

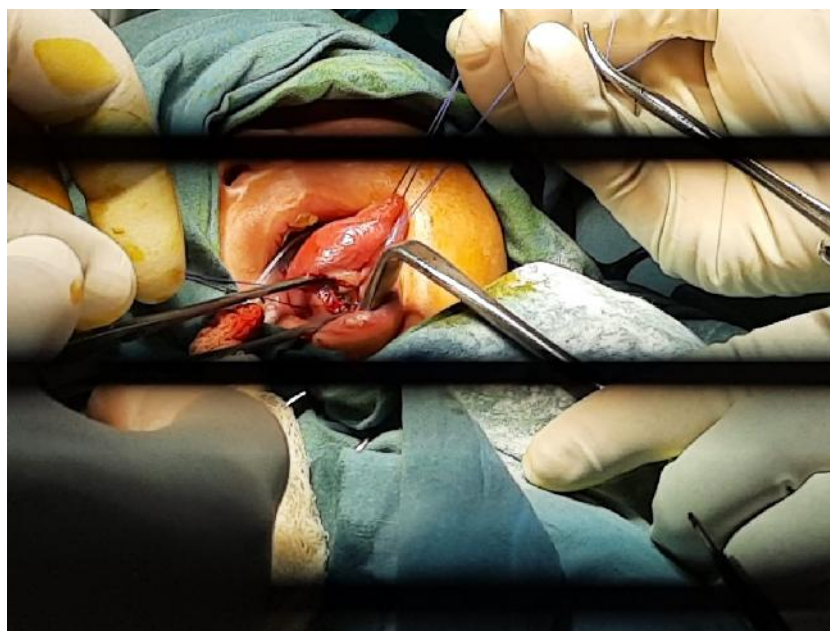


Fig 6: intraop dissection of oral rhabdomyoma

Conclusion

Rhabdomyoma is extremely rare condition of tongue. Treatment involves wide surgical excision of the lesion with clear margin, because any remnant of it is notorious for recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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