Case report

Congenital Obstructive Granular Cell Tumour Of The Gingiva – A Rare Case Report

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ABSTRACT

Congenital gingival granular cell tumors (CGGCT) are rare lesions which have occasionally been reported in India. It is an uncommon benign soft tissue lesion that usually arises from alveolar mucosa of neonates and causes respiratory and feeding problems. Due to its clinical features and treatment approaches these cases have aroused interest of different disciplines. We reported a case of a 4 day old female child who presented with a mass attached to upper alveolar ridge which was protruding outside and obstructing the entire oral cavity leading to difficulty in feeding. The USG and histological findings of the mass makes it mandatory that CGGCT should be differentiated from the teratoma, hemangioma and lymphangioma.

1. Introduction

Congenital granular cell tumor of the gingiva also known as congenital epulis originate commonly from alveolar ridge in new borns. They are rare tumors benign in histology [1]. The exact incidence, etiology and histogenesis of the tumor is unknown because of its rarity. CGGCT is also known as Neumann tumor, Abrikosov tumor and granular cell myoblastoma in medical literature [2]. CGGCT has been first described in 1871 as "congenital epulis" by Neumann[3]. The word epulis is derived from the greek word meaning "gum boil". The size of the tumor varies from few millimeters to 9 cm in diameter [4]. Females are affected 8-10 times more frequently than males [5,6,7]. Usually, it is presents as a single lesion however, multiple cases have been reported [7-10]. Newborn CGGCT is a different entity than in the adult with different immunohistochemical features [11]. The histological and immunohistochemical study in most cases support a mesenchymal origin [12]. The tumor recurrences have not been reported and malignant counterpart is unknown [5,13].

2. CASE REPORT

A 4 day old female child was admitted in view of congenital soft tissue mass protruding from the mouth and causing obstruction. The baby was delivered normally per vaginum. Mother had no significant past history. The USG examination of the mass showed a vascular tumour with patent cavum septum pellucidum. The clinical impression was congenital epulis, teratoma, hemangioma or lymphangioma. All other investigations were within normal limits. On clinical examination, a large mass was seen protruding outside the oral cavity causing oral obstruction as shown in fig1. Surgical excision of the tumour was done under general anaesthesia to relieve the obstruction. After surgical excision, the gingival surface appeared oedematous but otherwise unremarkable as shown in fig 2. Gross examination of tumour revealed a pedunculated, unencapsulated soft to firm tissue mass measuring 4 x 3 x 3 cm. The external mucosal surface was congested. On cut section a well circumscribed soft to firm, homogenous whitish mass was seen. (fig 3 & 4)

On histopathological examination, (as shown in fig 5,6,7,8) it was a well circumscribed submucosal tumour mass. The tumour mass was arranged in homogenous solid sheets. The tumour was composed of polygonal cells with round to oval nuclei having abundant eosinophilic granular cytoplasm. Few of them showed prominent nucleoli. The tumour had increased vascularity. No pleomorphism, necrosis or mitosis was seen.

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The tumour was PAS positive and diastase resistant (fig 9), vimentin positive (fig 10) and S100 negative (fig 11). In view of age, clinical presentation, site and histological findings a diagnosis of congenital granular cell tumour was offered.

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Figure 1: a large mass protruding outside the oral cavity causing oral obstruction

Figure 2: post operative gingival swelling

Figure 3: pedunculated, unencapsulated soft to firm tissue mass

Figure 4: cut surface- well circumscribed soft to firm, homogenous whitish mass

Figure 5 & 6: low power view of the tumour showing cells arranged in homogenous solid sheets
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Congenital granular cell tumor can remain undetected during prenatal ultrasound as in this case. After birth, the size increases causing obstruction for feeding. On gross, these tumours are pedunculated, whitish and soft to firm in consistency with many congested areas. On histopathological examination the tumour is submucosal solid mass composed of large oval to round cells with abundant eosinophilic granular cytoplasm. The nucleus is located centrally with intact cell membrane. The tumour has increased vascularity. The cytoplasmic granules are PAS positive and diastase resistant (8), similar findings were found in our case.

CGGCT is known as a benign mesenchymal tumor of unknown origin (2). The immunohistochemical profile of the tumor is different in newborns and adults. This tumor is a variant of the granular cell tumor, is S-100 negative immunohistochemically and does not show any differentiation specific to any cell type (11,14). Our case was also negative the S-100 immune marker.

Usual treatment is complete surgical excision. These tumors do not recur after excision (12,15-18). Our case also did not show any recurrence in the follow-up period that was within 18 months.
after excision. The clinical differential diagnosis of CGGCT includes teratoma, hemangioma and lymphangioma. The absence of heterogenous elements rules out teratoma while hemangiomas and lymphangiomas are “spongy” on palpation and have a red or dark blue surface[2], while in our case tumor was soft to firm and cut surface did not show any hemorrhage.

5. Conclusion

A case of congenital obstructive granular cell tumor was reported which on reviewing the literature, exhibits many of the described clinical features. The correct recognition of this tumor is helpful because of its good prognosis.

6. Bibliography


