Original Research Article

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Congenital granular cell tumour - a case report

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ABSTRACT

Congenital granular cell tumour (epulis) is a rare benign tumour occurring in gingiva of neonates, of unknown histogenesis, predominantly occurring in girls. It can cause feeding and respiratory difficulty. We describe a case of a newborn male baby born with swelling in the gingiva of anterior segment of upper jaw. The lesion was excised, with baby doing well at follow up.

Keywords: Congenital granular cell tumour, Epulis, Benign neoplasm, Gingiva

INTRODUCTION

Congenital granular cell epulis was first described by Neumann in 1871 and, hence also called Neumann's tumour and approximately 250 cases have been reported in the literature. It is a rare benign soft tissue lesion occurring mainly in the mucosa of maxillary ridges, with female preponderance. The lesion may cause feeding and breathing difficulty based on the site of the lesion

CASE REPORT

A 18 days old child presented with swelling in the gingiva of anterior part of upper jaw, which was noted since birth. The swelling was not causing any feeding or breathing difficulty. The infant was born at 37 weeks of gestation through caesarean section and weighed 2.6kg with no other congenital anomaly or malformations. On examination, a 1×1 cm growth was seen over gingiva of maxillary ridge. Surgical excision of the lesion was done and grossly it was a single lesion measuring 1.2×1.2×0.5cm with smooth surface. The cut section appeared whitish and glistening and the whole lesion was submitted for histopathological examination. Microscopically, sections showed ulcerated epithelium with an underlying lesion composed of nests of large polygonal cells with eosinophilic granular cytoplasm and

central bland nucleus. The diagnosis was consistent with congenital granular cell tumour. Immunohistochemistry was done with markers S100, CD68, vimentin and HMB45, the neoplastic cells were positive for vimentin and negative for S100, CD68 and HMB45.

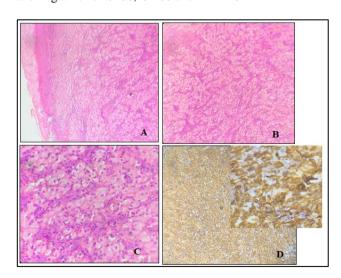


Figure 1: (A-C) Microscopy of the neoplasm showing ulcerated epithelium and nests of granular cells (D) IHC-vimentin showing positivity in granular cells.

DISCUSSION

Congenital granular cell tumour is a rare tumour arising in the gingiva of the incisor-canine region of the maxilla but not involving bone and teeth. It is grossly a pedunculated or lobulated swelling with smooth or ulcerated surface. The tumour typically ranges from 1-2 cm² and rarely size upto 9 cm is reported till date.³ There is a female preponderance of 8:1.3 prenatal diagnosis can be done by ultrasonography or MRI scan. Histogenesis of the tumour is uncertain and possible cell of origin proposed is odontogenic epithelium, undifferentiated mesenchymal cells, neuroendocrine progenitor cells and post degenerative or reactive changes.^{4,5} Microscopically, it is a fairly circumscribed lesion with nests and ribbons of large polygonal cells with granular eosinophilic cytoplasm and eccentric nuclei and small nucleoli. There is a prominent capillary network and thinning of overlying epithelium. Histological variants include lesions with prominent fibrosis and spindle cell morphology may be seen in older or traumatised lesions. Other variants have staghorn like vascular channels or odontogenic epithelium in between the granular cells.

Congenital granular cell show positivity for vimentin and NSE and characteristically lacks immunoreactivity to S100, CD31, CD34, CD68, keratins, SMA.

The differential diagnosis of congenital granular cell tumour is adult granular cell tumour which occurs in the age group of 30-60 years and rarely occurs in children. It presents as a painless nodule. Microscopy shows large granular cells, intermixed interstitial cells and cytoplasmic hyaline globules may be seen. Overlying epithelium shows pseudoepitheliomatous hyperplasia and frequent nerve involvement is seen. These tumour cell of adult granular cell tumour are positive for S100, NSE, laminin, myelin protein, PAS with diastase. However, the

cells of congenital granular cell tumour are negative for \$100 as in our case.

CONCLUSION

The treatment of congenital granular epulis is excision under anesthesia, although frequent monitoring for regression of the swelling is an alternative management as the swelling stops growing after birth and may spontaneously regress.

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