ORIGINAL RESEARCH PAPER
Volume-9 | Issue-1 | January-2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

CONGENITAL GRANULAR CELL TUMOUR – A RARE CASE REPORT AND REVIEW OF LITERATURE

Pathology

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ABSTRACT

Congenital granular cell tumour (CGCT), an unusual benign mucosal lesion occurring in the oral cavity in infants, was first described in 1871. CGCT presents at birth as a mass over the gingival mucosa of the mandible or maxilla, with a marked female preponderance. Although benign, they usually interfere with respiration and feeding, necessitating early intervention. A two-day-old male child presented with a mass protruding from his mouth. It was surgically resected and diagnosed as CGCT on histopathology. Recommended management for CGCT is early surgical resection, which can be performed under local anaesthesia. Removal of the tumour facilitates complete mouth closure leading to proper feeding and respiration.

KEYWORDS

Congenital Lesion, Granular Cell Tumour, Paediatric Surgery

INTRODUCTION

Congenital granular cell tumour (CGCT), an unusual benign mucosal lesion occurring in the oral cavity in infants, was first described by Neumann in 1871.[1] It is also known as congenital epulis, the word 'epulis' is derived from a Greek work which translates to 'on the gum'. It is a rare tumour, with less than 200 cases reported in literature.

Over the years, many theories have been proposed to explain its histogenesis and to determine its lineage. However, the definitive etiology and natural history have remained obscure, making it an intriguing tumour.

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CASE REPORT

A two-day-old male child was noted to have a mass protruding from his mouth. The infant was otherwise healthy and was normally delivered to a healthy mother at full term, with no antenatal or intrapartum complications. Intraoral examination revealed a 2.5x2 cm pedun culated soft tissue lesion over the left upper gingiva that was non-tender and non-fluctuant. It was firm in consistency with colour similar to the adjacent mucosa. The infant was unable to take feeds properly, however, there was no breathing difficulty. Based on the clinical findings, differential diagnosis of CGCT, haemangioma, and teratoma were made. The lesion was surgically resected and tissue submitted for histopathological examination.

The tumour was capsulated, irregular and grey white in colour. The cut section was yellowish white to brown in colour and solid in consistency. Light microscopy showed tumour comprising of round to polygonal cells arranged in sheets, having well defined cell borders, round eccentrically placed nucleus and abundant amount of granular cytoplasm (Figure 1 and 2). Immunohistochemistry showed negativity for S-100 protein. The diagnosis of CGCT was confirmed.

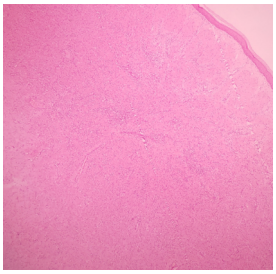

Figure 1 – Photomicrograph showing arrangement of tumour cells in diffuse sheets (4x)

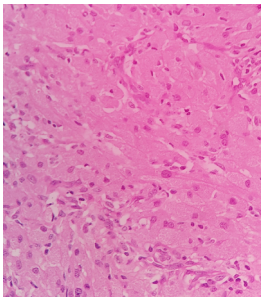

Figure 2 – Photomicrograph showing tumour comprising of round to polygonal cells arranged in sheets, having well defined cell borders, round eccentrically placed nucleus and abundant amount of granular cytoplasm (40x)

The child was followed up for a period of one year after surgery. There was no recurrence and the child had no other complications.

DISCUSSION

CGCT is a rare tumour, typically noted in infants just after birth. It usually arises from the mucosa over the alveolar ridge and interferes with mouth closure. This leads to difficulty in feeding and breathing.l Majority of the lesions are solitary, occurring on the maxillary alveolar ridge. This is probably due to the fact that this is the common site for supernumerary teeth.

Females are affected more frequently, with a ratio of almost 8:1. The female preponderance cannot be explained by hormonal receptor studies, as these tumours lack expression of estrogen and progesterone receptors.

The diagnosis is usually made clinically, however, the diagnosis may me missed in case of very small lesions or when the index of suspicion is low. The differential diagnoses include dermoid cysts or teratomas, haemangiomas, malformations of the lymphatics, melanotic neuroectodermal tumours of infancy, and rhabdomyosarcoma. Radiological investigations may be necessary, with magnetic resonance imaging (MRI) being most useful. The gingival origin of the tumour and absence of local extension can be visualized.[2]

Large lesions may completely obstruct the oral cavity and lead to a neonatal emergency on the first day of life itself. Occurrence of CGCT in a child with tetralogy of Fallot has also been documented.[3]

Prenatal diagnosis of larger lesions can be made during ultrasonographic (USG) screening after 26 weeks of gestation. Poor

swallowing usually leads to associated polyhydramnios, which may raise the index of suspicion.4 As the histogenesis and clinical course of the tumour are not established, it is imperative to make the diagnosis at the earliest possible stage and initiate intervention.

There are several differences between CGCT and the granular cell tumours (GCT) seen in adults. GCTs occur in all age groups and very rarely involve the gingiva. They usually exhibit pseudoepit heliomatous hyperplasia of the overlying epithelium. GCT exhibits immunoreactivity for S-100 protein, whereas CGCT is immunon egative for the same.

Recommended management for CGCT is early surgical resection, which can be performed under local anaesthesia. Removal of the tumour facilitates complete mouth closure leading to proper feeding and respiration. Few cases have undergone reduction is size, advocating the use of observation and spontaneous regression.5 CGCT is a rare tumour, typically noted in infants just after birth. It usually arises from the mucosa over the alveolar ridge and interferes with mouth closure. This leads to difficulty in feeding and breathing.[1] Majority of the lesions are solitary, occurring on the maxillary alveolar ridge. This is probably due to the fact that this is the common site for supernumerary teeth.

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