

Case Report

CONGENITAL GRANULAR CELL EPULIS: REPORT OF 2 CASES

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إن خلايا الورم الليفي اللثوي الحبيبية الخلقية في حديثي الولادة نادراً ما تكون سرطانية في اللثة المغطية للارتفاع السنخي الأمامي لل فك . وقد ذكرها نيومان في عام ١٨٧١ وقد عرفت أيضاً بأنها سرطان الخلايا الحبيبية اللثوية الخلقى .
هذه الآفة هي ورم مرجلي ذات قوام متماسك وسطح أملس أو مفضص حجم الورم ذات قطر يتراوح بين بضعة ملليمترات إلى سنتيمترات . وحدوثه عند الإناث أكثر بعشرة مرات من الذكور . ويشاهد الورم في الفك العلوي أكثر من السفلي بمرتين وغالباً يكون في منطقة الأنثياب والأسنان الأمامية . ولا ينمو أو يكبر بعد الولادة وعادة غير ناكس بعد الاستئصال الجراحي .
يظهر الفحص المجهرى للورم كتلة مركزية ذات خلايا حبيبية متراسة .
ويغطي الورم نسيج ظهاري قشري مصفغ مع نسيج ضام ظهاري أملس .

Congenital granular cell epulis of the newborn is a rare lesion whose histogenesis and natural clinical history have remained obscure. It is important that new cases of this lesion are reported from different populations so that its occurrence and frequency may be ascertained more accurately. Furthermore, the lesion is important in the differential diagnosis of the more aggressive lesions seen in early life. The lesion occurs more often in female infants, but in our cases, one was a male and the other was a female.

Introduction

The congenital granular cell epulis of the newborn is a rare benign tumor that occurs on the gingiva in the anterior alveolar ridge of the jaws.¹ Originally described by Neumann² in 1871, it is also known as congenital gingival granular cell tumor.

This lesion is a pedunculated tumor of firm consistency with a smooth or lobulated surface. The size of the tumor varies from several millimeters to centimeters in diameter.³ Females are affected ten times more often than males. The tumor is seen twice as often in the maxilla than in the mandible and usually in the incisor canine region. It does not grow bigger after birth and normally there is no recurrence after surgical excision.¹

On microscopic examination, a central mass of closely packed granular cells is seen. The tumor is covered by a stratified squamous epithelium with a flat epithelium-connective tissue junction. A prominent arborizing fibrovascular network in the thin connective tissue septa is usually noted throughout the tumor.⁴

Since the light microscopic appearance of the granular cells is similar to that seen in

granular cell myoblastomas, some investigators have described congenital epulis as congenital gingival granular cell myoblastoma.⁵ In contrast with congenital epulis, granular cell myoblastoma occurs anywhere in the skin or mucous membranes and in any age group.⁶ It is rarely congenital, and may recur when incompletely excised. Histomorphologic features such as the prominent fibrovascular network and the absence of pseudoepitheliomatous hyperplasia of the overlying epithelium present in congenital epulis help to differentiate these two entities.⁷ Case 1: An otherwise normal twenty-day old male infant presented with a grape-sized growth of the mandibular alveolar process. Clinical examination showed a pedunculated mass measuring 1.5x1x0.5 cm in the incisive region arising from the gingiva of the mandibular alveolar ridge and covered with the normal mucosa (Fig. 1). The tumor was surgically excised.

Macroscopic examination showed a lesion approximately 1.5x0.8x0.5 cm; the cut surface was white-gray and smooth. Histologic examination of the tumor revealed a surface border of typical squamous epithelium and a narrow zone of subepithelial connective tissue.

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There was no pseudoepitheliomatous hyperplasia of this covering epithelium (Fig. 2). The tumor consisted of closely packed relatively large cells with eosinophilic, granular cytoplasm and small, basophilic nuclei (Fig. 2). There was

mass interfered with normal breast-feeding. Under a local anaesthetic, the lesion was completely removed surgically.

Macroscopically, the excised lesion measured 2x1.8x0.8 cm and its cut surface was

white-gray and smooth. The histologic report was similar to that of Case 1 (Fig. 3).

Discussion

Congenital epulis is a rare tumor of the newborn. One hundred and sixty-seven cases (195 lesions) have been reported since 1871.'

Females are affected 8 to 10 times more often than males,^{1,8,9} and the lesion was reported to be seen usually 2-4 times more often in the anterior maxilla than in the

mandible.^{1,9,10}

Despite the characteristic anatomical location of congenital epulis on the anterior alveolar ridge of the maxilla and mandible, correct preoperative clinical diagnosis was seldom made.^{9,10} Only three of 21 cases after reported by Lack et al¹¹ were correctly diagnosed clinically as congenital epulis. Diagnoses such as haemangioma, fibroma and granuloma were made."

Although fibro-blastic, histiocytic, myogenic, neurogenic, odontogenic and endocrine origins have been discussed,¹² immunohistochemical and ultrastructural investigations point to an origin from primitive mesenchymal cells with differentiation toward myofibroblasts.^{13,14}

The preponderant female occurrence suggests an in utero hormonal stimulus. The lesion is also known to regress

spontaneously by birth.⁸ Estrogen receptor studies of epulis tissue, however.



Figure 1: Congenital epulis situated on the mandibular alveolar process of one of the newborns.

no sign of cellular pleomorphism or mitotic activity. Many capillaries were present. Immunohistochemically, S-100 protein was negative. A diagnosis of congenital epulis was made.

Case 2: A ten-day old female infant, product of a normal full-term pregnancy was observed at birth to have a tumor attached to the anterior ridge of the maxilla. The oral mass was a well encapsulated, lobulated and pedunculated tumor measuring 2x2x1 cm. The

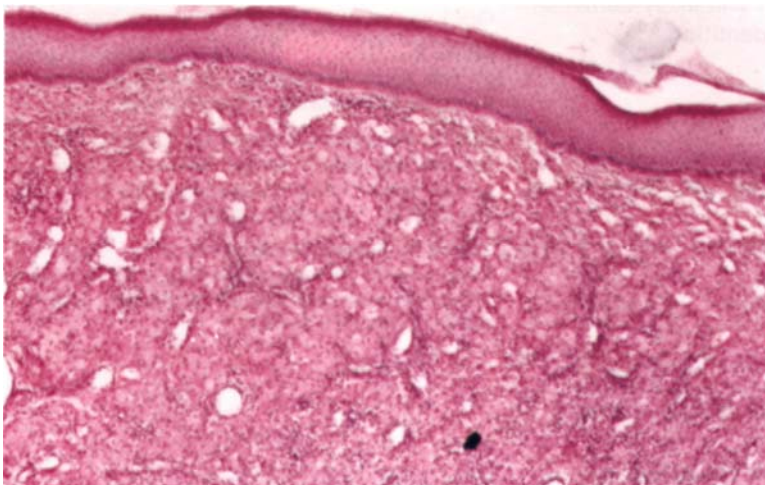


Figure 2: Flattened stratified squamous epithelium covers a mass of granular cells and thin capillary blood vessels. (HE. X40)

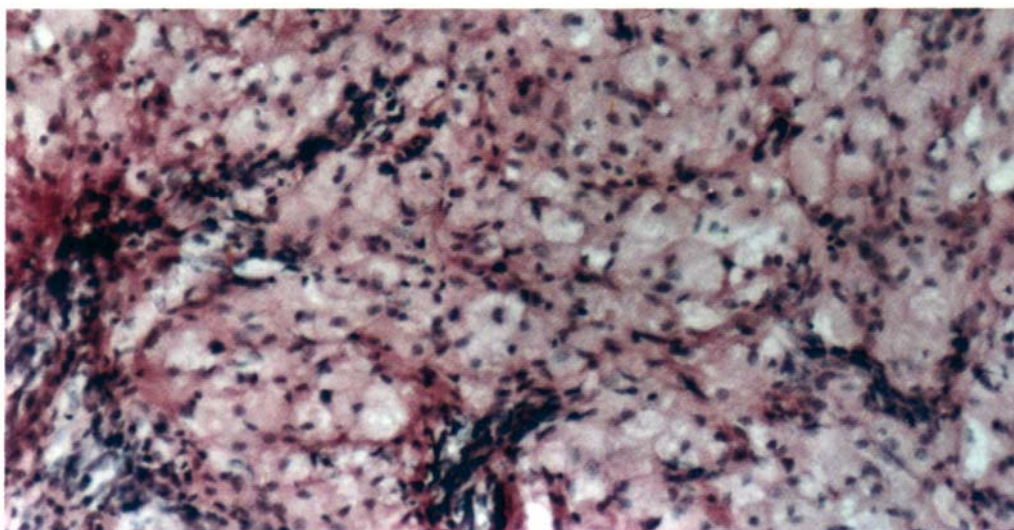


Figure 3: Large tumor cells with eosinophilic granular cytoplasm and basophilic nuclei. (H.E. X200).

have proved negative⁵ and exogenous maternal steroid used during pregnancy has not been associated with occurrence of congenital epulis.⁹

A similar congenital tumor which does not show granular cells microscopically although densely collagenized has been described.⁶

When congenitality, anatomical site location, female gender preponderance, absence of epithelial pseudoepitheliomatous hyperplasia and S-100 protein are considered, the differential diagnosis of congenital epulis is not difficult.⁸

The preferred treatment for congenital epulis remains surgical resection at the level of the alveolus. Deeper resections will likely damage the underlying unerupted dentition.⁹

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