Neumann's Tumor Vs Abrikossoff's Tumor: Uncommon Neoplasm with Controversial Histiogenesis (Granular Cell Tumor)

Dhiraj Nikumbh[1,*], Sudhir Singhari[2], Shirish R. Gondane[3]

[1]Associate Professor, [3]Assistant Lecturer, Dept. of Pathology, JMF'S ACPM, MC, Dhule, Maharashtra, [2]Consultant Pediatric Surgeon, Dhule, Maharashtra

***Corresponding Author:**
E-mail: drdhirajnikumbh@rediffmail.com

Abstract

Granular cell tumor is an uncommon soft tissue tumor with controversial histiogenesis that can affect any organ of the body including orofacial region. It is mostly benign neoplasm. Neumann's tumor and Abrikossoff's tumor are rare entities with different histiogenesis with varied clinico-pathological presentations. The etiology of the Granular cell tumor remains unclear due to its muscular, connective or neural tissue origin. It is a matter of long debate and diagnostic dilemma.

We are presenting this case report of congenital granular cell tumor in a 2 days old baby over an anterior maxillary alveolar ridge; diagnosed on histopathology and immunohistochemistry.

The aim of this report is to highlight the rarity of congenital granular cell tumor and its difference with other granular cell tumours.

Keywords: Congenital, Granular cell tumor, Newborn.

Introduction

In the newborns, soft tissue mass protruding from mouth and baby presented with difficulties in respiration and feeding due to mass, is a panic situation for the parents as well as clinicians. Granular cell tumor (GCT) is rare benign soft tissue tumor with controversial histiogenesis[(1)]. It is most commonly presents in 40-60 years of age and is rare in children[(1)]. Granular cell tumor may be congenital or acquired later in life. Congenital granular cell tumor also called as "congenital epulis" first described by Neumann in 1871, hence termed as Neumann's tumor[(2,3)]. Congenital GCT can be differentiated from adult GCTs. Congenital GCT is a rare clinical entity with pathological diagnosis whose origin remains unclear[(4)]. Long debate and controversial histiogenesis is associated with congenital GCTs, as myoblastic, histiocytic, neurogenic and endocrinological origin but not proved yet[(4)].

In 1926, GCTs were first described as myoblastomas by Abrikossoff and thought to be derived from smooth muscle[(5)]. GCTs are also known as Abrikossoff's tumor[(5)]. Later on various scientists named the tumours by various names. However, in 1999, Granular cell tumor (GCT) is the world nomenclature adopted by World Health Organisation (WHO)[(6)].

We are presenting this case with prime aim as to discuss varied histiogenesis, microscopical differences between Neumann's and Abrikossoff's tumor.

Case report

A 2 days female child presented with something protruding from mouth and with difficulty in feeding and respiration. The patient was referred to paediatric surgeon due to anxiety and panic situation. On local examination a soft lobulated mass arising from the upper jaw mainly from central and lateral incisor tooth and attached by narrow pedicle (Fig 1). No other congenital anomalies were noted. All haematological, biochemical and serological investigations were within normal limits. The mass was surgically excised in toto under general anaesthesia without any complications. We received the excised specimen for histopathology. Postoperative period was uneventful.

Grossly:- The mass was greyish white measuring 3x2x1 cms .E/S was well circumscribed and C/S showed firm ,whitish soft tissue mass (Fig 2) without areas of haemorrhages and necrosis. Multiple sections were taken and studied.

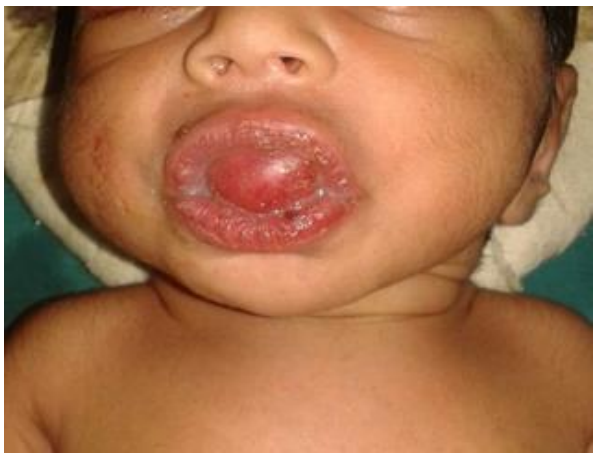

Fig. 1: Pre-operative image of lobulated mass from central and lateral incisor tooth and attached by narrow pedicle.

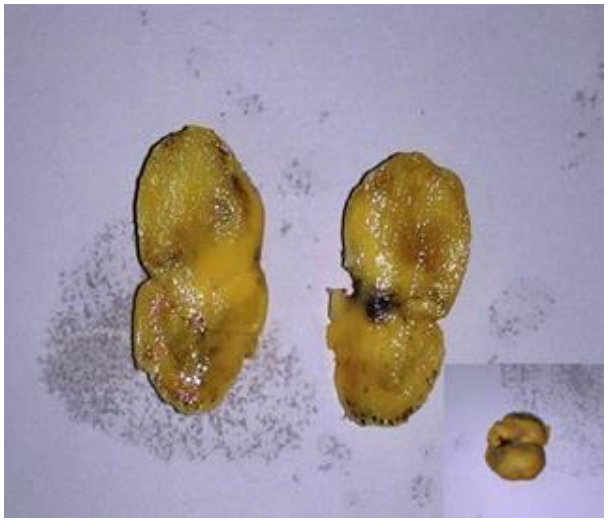

Fig. 2: Cut section of the excised lobulated whitish mass without haemorrhages and necrosis (Inset-External lobulated mass)

Light Microscopy:- Showed tissue bits lined by stratified squamous epithelium and beneath it shows a tumor (Fig 3). The overlying epithelium doesn't show any pseudoepitheliomatous hyperplasia. The tumor composed of sheets, nests and clusters of round to polygonal cells having abundant granular eosinophilic cytoplasm with monomorphic vesicular nuclei with inconspicuous nucleoli (Fig 4). The intervening fibrocollagenous septae showed network of blood vessels. Final histopathological diagnosis was given as congenital granular Cell tumor (Neumann's Tumor). Immunohistochemistry was performed on block showed diffuse negativity for S-100, confirmed the Neumann's tumor (Fig 5).

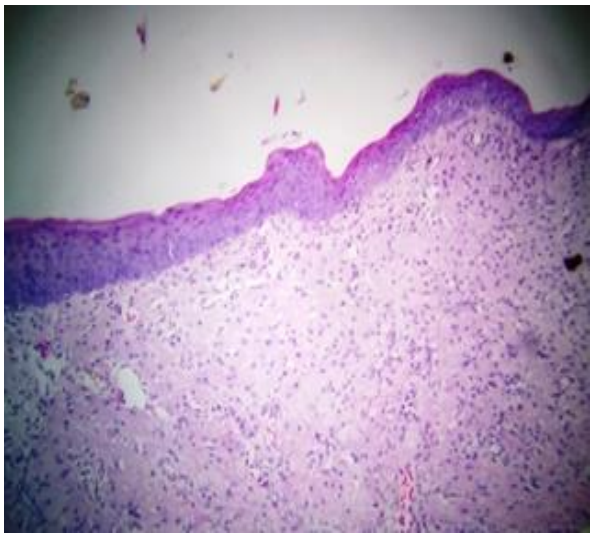

Fig. 3: Photomicrograph showing Tumor with overlying stratified squamous epithelium without pseudoepitheliomatous hyperplasia. (H&E, x100)

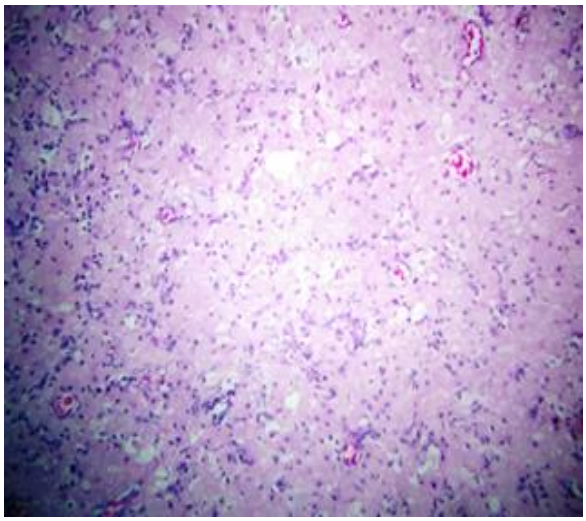

Fig. 4: The tumor cells are round to polygonal cells with abundant granular eosinophilic cytoplasm and monomorphic vesicular nuclei (H& E, x100 and x400)

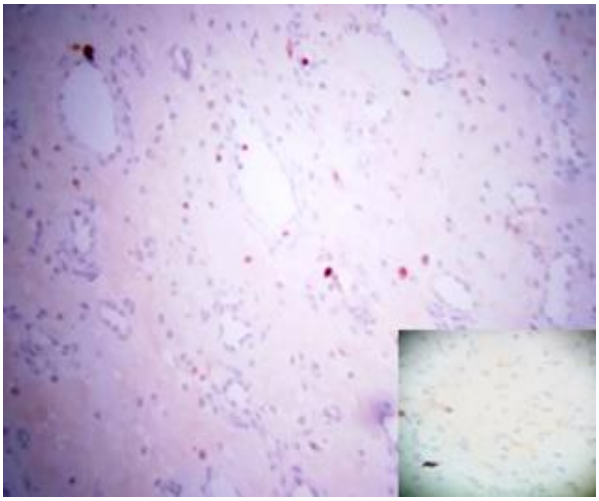

Fig. 5: IHC image Showed diffuse negativity of S-100 marker (Inset-high power view)

Discussion

Granular cell tumor (GCT) is an uncommon benign soft tissue neoplasm arising from mucosa of the gingiva and presented as mass protruding from the infants' mouth often interfering with the respiration and feeding[7].

Congenital GCT or congenital epulis/Granular cell epulis also referred as "Neumann's tumor"[2,3]. It is almost important to be differentiated from adult GCTs i.e. Abrikossoff's tumor; granular cell myoblastic or granular cell schwannoma[8]. The origin of these tumours remains unclear and controversial for its peculiar behaviour, variable localization and not completely clarified etiology[8]. GCTs can occur in any part of the body with more than half of the cases are

presented in head and neck areas. The most frequent orofacial localization is tongue[9].

GCT can develop at any age (4-6) and while some authors has observed a greater prevalence among women[10].

In majority of the cases, GCTs has benign behaviour. Occasionally, it is locally aggressive and 2% cases are malignant with distant involvement[11]. Clinically, the appearance of GCT is not distinguishable from that of other benign connective tissue tumours such as lipoma, fibroma and neural tumours such as schwannomas, neuromas and neurofibromas[8]. The lesion due to modification of mucosa, clinically appears as carcinoma[8]. The correct localization with excisional biopsy with safety margins and final histopathological study is obsolete necessity for the proper diagnosis and treatment of lesion[4.8].

Congenital GCT (Neumann's tumor) is different entity from other GCTs (Abrikossoff's tumor). Neumann's tumor has a marked female preponderance (8-10:1) and mostly occurs on maxillary alveolar ridge in neonates[12,13], presents at birth. Abrikossoff's tumor is rarely seen in first decade of life, it is most frequently diagnosed between the third and sixth decades of life, affects a wide variety of visceral and cutaneous sites and also has predilection for the females[14].

Histopathologically, Neumann's tumor consists of granular cells similar to other GCTs with some differences as marked vascularity, more nerve bundles, and absence of pseudoepitheliomatous hyperplasia[1] Abrikossoff's tumor has granular cells with pseudoepitheliomatous hyperplasia of overlying epithelium with less vascularity and less nerves[12,13].

Congenital GCTs thought to be originated from odontogenic epithelial cells, undifferentiated mesenchymal cells, fibroblasts, myofibroblasts or schwann cells or although the exact histiogenesis is not known[14]. However, it is currently believed that the GCTs are caused by alterations in the cellular metabolism of Schwann cells - a hypothesis that is reinforced by the constant presence of S-100 marker in immunohistochemistry (IHC)[14]. Regarding IHC, Abrikossoff's tumor showed diffuse positivity of S-100 proteins and Neumann's tumor showed diffuse negativity.

In the present case, histopathology suggestive of Neumann's tumor in view of granular cells, rich vascularity and neural bundles without severe pseudoepitheliomatous hyperplasia of overlying epithelium.

IHC findings of S-100 protein was diffusely negative in our case, confirms congenital GCT (Neumann's tumor) and differentiates it from Abrikossoff's tumor[12-15].

Granular cell tumor	Neumann's tumor	Abrikossoff's tumor
Histopathology	Granular cells with rich vascularity, nerve bundles and pseudoepithelioma tous hyperplasia absent	Granular cells with severe pseudoepitheliomat ous hyperplasia with less vascularity
Age	Congenital, Neonates	Adults
Sites	Anterior alveolar ridge of maxilla (upper jaw)	Head and neck most common
Histiogenesis	Odontogenic epithelium	Schwann cells
IHC: S-100	Diffuse negativity	Diffuse positivity

Conclusion

Congenital GCT/Neumann's tumor is a pathological diagnosis. It can cause extreme anxiety in parents as well as in clinicians. Surgical excision is the treatment of choice. Besides helping in correct diagnosis, IHC has allowed to improve the knowledge of controversial histiogenesis of Neumann's tumor from Abrikossoff's tumor. We highlighted these points in present case due to its rarity.

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