ISSN: 0975-833X

Availableonlineathttp://www.journalcra.com

International Journal of Current Research
Vol. 11, Issue, 10, pp.7552-7555, October, 2019

DOI: https://doi.org/10.24941/ijcr.36753.10.2019

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

CASE REPORT

GRANULAR CELL TUMOR: A CASE REPORT AND REVIEW OF LITERATURE

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ARTICLE INFO

Article History:
Received 04th July, 2019
Received in revised form
19th August, 2019
Accepted 15th September, 2019
Published online 30th October, 2019

Key Words:
Granular cell Tumor,
Abrikossoff's tumor,
Granular cell myoblastoma.

ABSTRACT

Granular cell tumor (GCT) is a common neoplasm that can occur in any part of the body, including the orofacial region with controversial origin. The tumor is usually benign, but there are case reports in which the tumor shows a locally aggressive behavior, malignancy and distant metastases. The most widely accepted hypothesis is that granular cell tumor arises from the altered metabolism of Schwann cells. The tumor is typically asymptomatic and appears as a nodule that does not exceed 3cm. This article brings forth a case of GCT occurring in a 42 years old male patient on the lateral border of the tongue.

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***Citation: Arun B.S., Rekha Krishna Pillai, Priya Thomas and Bindhu, P.R.* 2019.** "Granular Cell Tumor: A Case Report and Review of Literature", *International Journal of Current Research*, 11, (10), 7552-7555.

INTRODUCTION

Granular cell tumour (GCT) is a rare benign neoplasm, first described by Abrikossoff in 1926 as myoblastomas, because of its origin from smooth muscle (Rejas *et al.,* 2011). Since then numerous hypotheses have been laid down on its origin and for a long time believed to be of neural origin. Hence various terminologies have been put forth for GCT as granular cell neurofibroma, granular cell neuroma and granular cell schwannoma (Garin *et al.,* 1992). Recently the ubiquitous nature of the tumor has been recognized with reports of GCT arising in various sites like skin, breast, lung, nervous system, gastrointestinal tract, urinary bladder, male and female reproductive tracts, as well as other sites in the head and neck region including the orbit, larynx, parotid gland and peripheral and cranial nerves (Sposto, 2006; Collin, 1995). In Head and Neck region GCT accounts for about 45 -65% all involved sites with tongue being the most common site of occurrence (Sposto *et al.,* 2006). Though the lesion can present in any age group, occurs most frequently during fourth to sixth decades of life, but very rarely as congenital disease and affect both sexes, although most common being in females probably due to hormonal influence. Clinically GCTs grow slowly and insidiously and is characterized by the presence of a large amount of dense cytoplasmic lysosomes in different fragmentation stages, giving it a granular lesion under microscopy (Garin L *et al.,* 1992).

Here we report a case of an asymptomatic mass on the tongue of a 42 - year old male patient.

CASE REPORT

A 42 - year old male patient reported with a chief complaint of painless mass on the tongue of 1 year duration. The swelling was slow growing and attained the present size. There was no other relevant medical history. Intraoral examination revealed an asymptomatic pale pink sessile nodule of 2cm in diameter on the left lateral border of the tongue. The mucosa overlying the swelling was intact and normal in colour (Figure1). On palpation the nodular solitary lesion was non tender and firm in consistency with no evidence of palpable lymph nodes. A provisional diagnosis of Neurofibroma/ fibroma was made and an excisional biopsy was advised (Figure 2). Histopathologically the lesion showed proliferation of numerous large polygonal cells with abundant pale eosinophilic granular cytoplasm arranged in sheets and islands in a fibro cellular connective tissue (Figure 2A). Neoplastic cells were also seen between the muscle fibers with evidence of neurovascular bundles in the deeper layer (Figure 2B). Periodic acid-Schiff (PAS)-stain positive granules were detected in the cytoplasm (Fig 2C) and IHC analysis with S -100 showed positive granules (Fig 3D). Based on the histopathological findings a diagnosis of granular cell tumor was given. The patient is under regular follow-up for the last 8 months with no signs of recurrence.

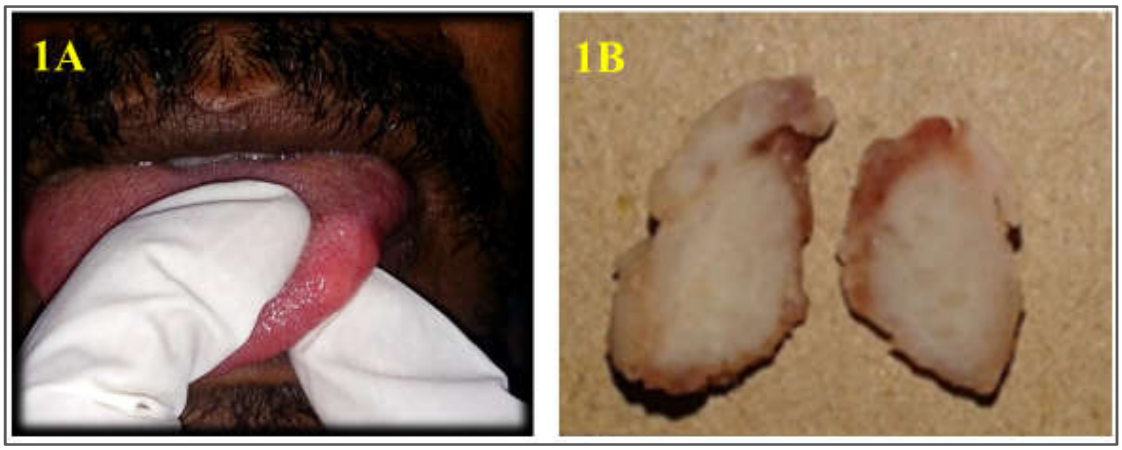

Fig 1. Clinical photograph of a Granular cell tumor in the anterolateral aspect of tongue. (A) & the gross specimen excisional biopsy (B)

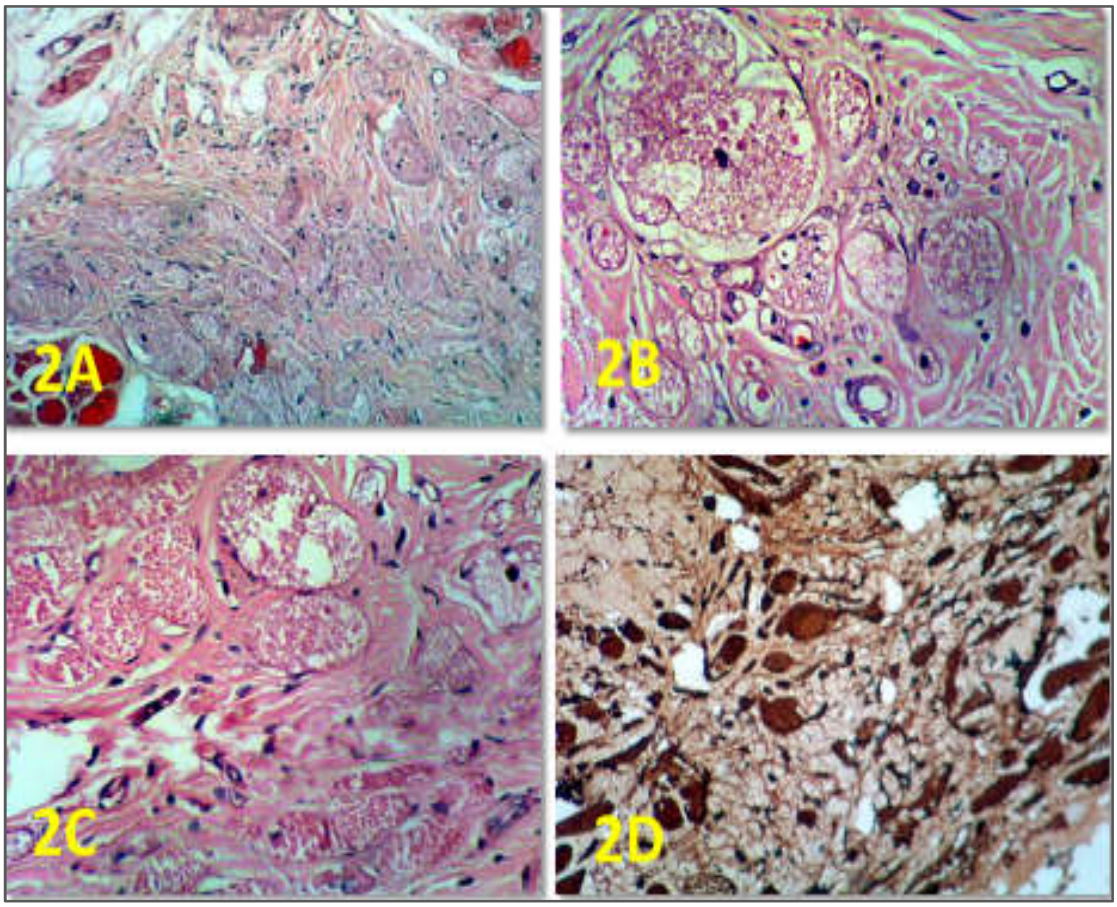

Fig 2. Photomicrographs demonstrating histopathologic features of GCT. A. Numerous large polygonal cells with abundant pale eosinophilc granular cytoplasm arranged in sheets and islands (H&E; 40x)B. Invasion of granular cells into muscles (H&E;100x) C.granular cells stained positive for periodic acid-Schiff positive granules (PAS 100x) D. cells immunoreactive for S100. (100x)

DISCUSSION

1st description of Granular cell tumor (GCT), an uncommon benign neoplasm, is credited to Abrikoss off in 1926 (Rejas *et al.,* 2011). The histogenesis of GCT has been a source of controversy since its recognition as an entity and has been associated with skeletal muscle, histiocytes, fibroblasts, myoepithelium and nerve cell connective tissue origin. (Budino‑Carbonero, 2003). Presence of autophagic vacuoles and positive IHC staining for α-1 antichymotrypsin and α-1 anti-trypsin in GCT suggest histiocytic origin Alessi and Zimmerman, 1988). Neural origin for GCT was postulated by various authors and its origin with Schwann's cells was based on ultra structural similarities between Schwann's and granular cells: similarity between the granules in granular cells and altered myelin, concentric arrangement of granular cells around nerve ends and presence of lipoproteins and sphingomyelin in granular cells. All these indicated that the granules in these cells were made of myelin or the product of its degradation; Moreover the positivity for protein S-100, enolase and myelinic proteins PO and P2 in these granules by immunoperoxidase techniques supports its neural or neuroectodermal origin. Since the main morphologic feature is the granularity of the cytoplasm, caused by a massive accumulation of lysosomes the World Health Organization (WHO) adopted the term as granular cell tumor. (GCT) (2). GCT found more frequently in the oral mucosa which accounts for 70% of all sites affected by the tumor especially the tongue and hard palate (Rejas *et al.,* 2011). Among oral mucosa 23-28% of cases are seen to occur in tongue (Sposto *et al.,* 2006). GCT manifests as solitary or

multiple asymptomatic firm pink or yellow well delimited lesion that rarely exceeds 3cms in diameter at the time of diagnosis and is covered by intact mucosa (Vered *et al.,* 2009). There are reports of tender swelling although our patient reported asymptomatic growth. The GCT manifests itself in the form of sub dermal or sub mucosal tumors with cells arranged in diffuse masses and strings. GCTs are un encapsulated and with imprecise borders. They may also invade and infiltrate adjacent tissues. In some cases, the epithelium that covers the tumor exhibits pseudoepitheliomatous the liomatous hyperplasia (Collin *et al.,* 1995). The lesion is characterized by proliferation of large fusiform or polygonal neoplastic cells with abundant pale eosinophilic cytoplasm containing cytoplasmic granules, a small round to oval dark hyperchromatic and eccentrically located nucleus and undefined cytoplasmic limits with marked cell membrane. Some cells may have more than one nuclei (Collin *et al.,* 1995). The most characteristic of granular cells is the membrane-contained cytoplasmic granules with micro vesicles, increased density areas, microtubules and myelinic formations. The pale, characteristic granules inside the cells are PAS (periodic acid-Schiff) positive and diastase-resistant (Noronha and Dias, 1997) and contain glycoprotein, lipoid material and polysaccharides (Thawley, 1974). In addition to PAS other sspecial stains which shows positivity for GCT are Sudan black B and Masson's trichrome.

In immunohisto chemistry granular cell tumors are specifically positive for protein S-100, neuron-specific enolase (NSE), Laminin, and Myelin proteins (P0 and P2). The coarse granularity in GCT is thought to be because of Phagolysosomes (Manara, 1981) which stain positive for NK1/C3 (CD 63) (nonspecific lysosomal membrane glycoprotein) and KP1 (pan macrophage CD 68), another lysosomal marker. (Basile and Woo, 2003) Granular cells can occur as a degenerative, reactive, or hamartomatous phenomenon [Basile and Woo, 2003]. 72% of granular cell tumors show loss-of-function mutationsin **ATP6AP1** or **ATP6AP2**. Silencing of these genes in vitro results in impaired vesicle acidification, redistribution of endosomal compartments and accumulation of in tracytoplasmic granules (Pareja *et al.,* 2018). Immunohistochemical analysis has shown a strong and consistent positivity for protein S-100, a finding supporting the hypothesis that GCT is of peripheral nerve sheath origin. In an immuno profile study on GCTs conducted by Rejas *et al.,* 2011 showed nerve sheath differentiation that supports neural origin of these tumors and contributing to the establishment of a differential diagnosis between this lesion and other oral granular cell tumors, whether benign or malignant. Vered *et al.,* recently tested an extensive panel of antibodies to determine the true origin of this tumor. In most cases, granular cells were strongly and diffusely positive for p75, vimentin, calretinin, NKI/C3, inhibin-a, protein gene product 9.5 (PGP9.5) and protein S-100. GCT should be clinically differentiated from other common soft tissue tumor occurring in the oral cavity such as fibroma, neurofibroma and schwannoma etc. In the oral cavity, granular cells can also be found in lesions other than GCT, including congenital epulis of the newborn, rhabdomyoma, fibroxanthoma, ameloblastoma, ameloblastic fibroma, odontogenic fibroma and odontogenic cysts. (Mirchandani R 1989) Presence of granular cells, numerous capillaries, scattered chronic inflammatory cells, overlying atrophic epithelium and not reactive to S-100, smooth muscle actin, CD-68 and desmin differentiate congenital epulis from GCT (Senoo *et al.,* 2007). GCT should be differentiated from Rhabdomyoma by presence of coarse granular cytoplasm, absence of cross striations and glycogen and positive reaction for muscle specific actin, HHF 35, desmin, and myoglobin (Amelia Souza *et al.,* 2013). Fibro xanthoma may sometime be confused with GCT. Lipid droplets, nuclear atypia and cellular pleomorphism may differentiate this tumor from GCT. Some of the odontogenic lesions also show granular cell changes but identified with their characteristic histopathological features and its anatomical location. Malignant GCT is very rare and has a poor prognosis, as patients die within 2–5 years after diagnosis. It occurs in only 1–2% of cases (Kamal 1998). Surgical excision with a safety margin is the treatment of choice for GCT, although this is not always possible because the tumor lacks a capsule, a condition histologically demonstrated by an undefined cell margin. An excisional biopsy was the treatment of choice in the present cases and was followed-up for three years with no signs of recurrence.

Conclusion

Though GCT is considered as a benign variant, it may rarely cause recurrence and behave in a malignant fashion. Early diagnosis and timely surgical treatment is of prime importance in GCT like any other granular cell lesion. Surgical excision with a safety margin is the treatment of choice for GCT, although this is not always possible because the tumor lacks a capsule, a condition histologically demonstrated by an undefined cell margin.

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