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CASE REPORT

Granular Cell Tumor of Labial Mucosa: An Unusual Presentation

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ABSTRACT

Introduction: Granular cell tumors (GCTs), also called as Abrikossoff tumors, are uncommon benign neoplasms occurring at any body site and 40% of cases occur on the tongue. They have been postulated to originate from myoblasts, histiocytes, nerve sheath cells, neural endocrine, undifferentiated mesenchymal cells, and fibroblasts. They are probably neural in origin. They are more frequently found in females aged between 10 and 50 years.

Case report: A 15-year-old girl was presented with an asymptomatic swelling on the left vermillion border of the lip. The swelling was noticed a year ago, which showed as a nontender, yellowish, firm, submucosal nodule. Provisional diagnosis of traumatic fibroma was made.

Investigation: Routine histopathological examination and confirmatory periodic acid-Schiff stain (PAS) were done.

Management and prognosis: The lesion was surgically excised and the patient was under follow-up. No recurrence of the lesion was found during the course.

Conclusion: Granular cell tumor is a neoplasm with a wide variety of features and architectural patterns. Both benign and malignant lesions have been reported; hence, careful postsurgical histopathological examination and follow-up are required.

Keywords: Abrikossoff tumor, Granular cell tumor, Labial mucosa.

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INTRODUCTION

Granular cell tumors were primarily reported by Abrikossoff in 1926 as an uncommon, benign, soft-tissue tumor and named it as myoblastomas. They were thought to be derived from smooth muscle and hence, also termed as Abrikossoff tumors.[1]

In 1935, Feyrter suggested that these are probably derived from nerve tissue and hence, labeled them as granular cell neuromas.[2,6] Fust and Custer confirmed this theory in 1948, and they named it as granular cell neurofibroma.[2] In 1962, Fisher and Wechsler performed electron microscopic and immunohistochemistry study and found that they originated from Schwann's cells, and thus called them as granular cell schwannomas.[2,7] However, the World Health Organization adopted the nomenclature as GCT.[2]

Although initially classified as a myoblastoma, recent literature found that it is probably neural in origin. The GCTs are commonly seen in the head and neck region. They are also seen in dermal and subcutaneous regions. Intraorally, the tongue is the common site in 40% of cases. Internal organs like stomach, bronchus, larynx, pancreas, anus, biliary ducts, rectum, and oral tissues are affected in 40% of cases.

Granular cell tumors are seldom found in the extremities and large intestine. There is only 10% occurrence of multiple GCTs. Benign lesions are frequent and the malignant cases occur rarely, in only 1 to 2% of cases. In children, only twenty cases have been accounted till date.[2]

CASE REPORT

A 15-year-old girl presented with an asymptomatic swelling on the left vermillion border of the lip. The swelling was diffused measuring 2 × 2 cm in size and was noticed a year ago, which showed a nontender, yellowish, nonbleeding, firm, and smooth submucosal nodule, which was freely moving under the overlying mucosa, and the skin overlying swelling was soft (Fig. 1). History revealed that it showed as a small painless swelling, which gradually progressed and increased in size and became slightly hard. She had no relevant medical history. Provisional diagnosis of traumatic fibroma was made.

Total excision of the swelling was done under local anesthesia in relation to the left lower lip. The excised specimen was 1 × 1 cm, yellowish white, firm in consistency, and irregular, smooth surface (Fig. 2). The specimen was processed and routine histopathological examination was done. After histopathological examination, sections showed large fascicles of tumor cells demonstrated as nests or sheets. The tumor cells were large in size, polygonal- or oval-shaped, with small, uniform,

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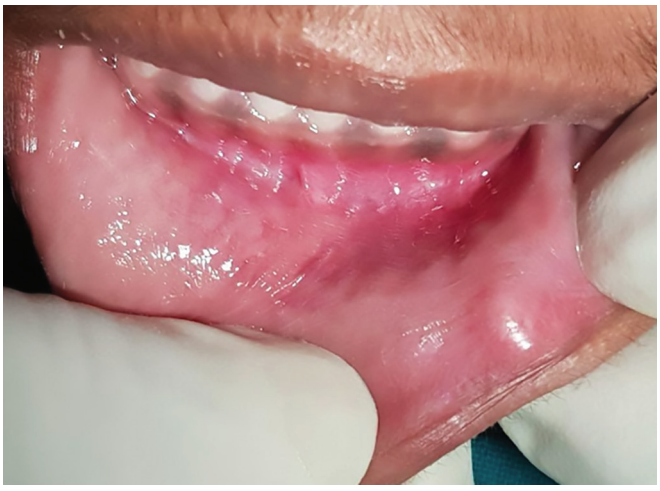

Fig. 1: Clinical appearance of tumor

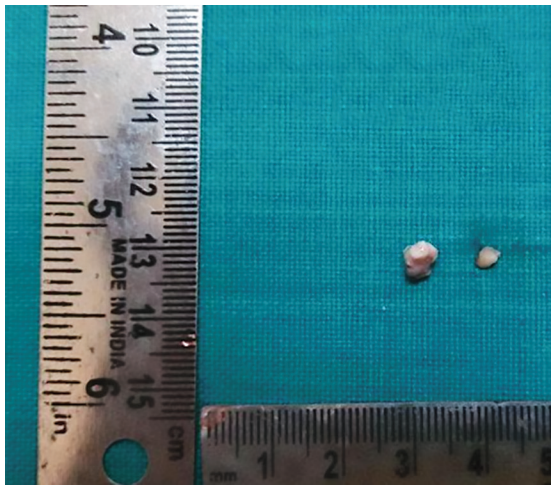

Fig. 2: Gross specimen

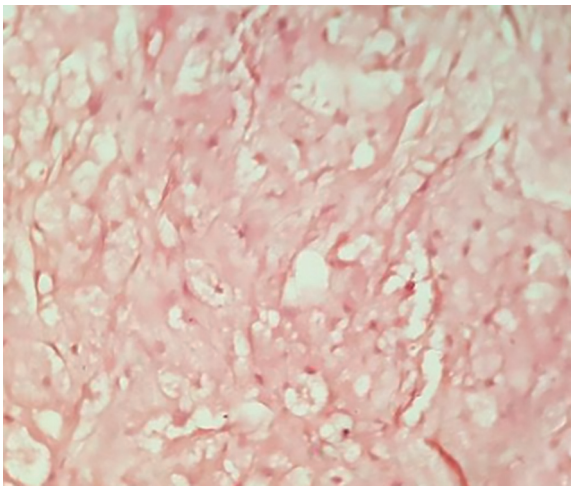

Fig. 3: Granular cells, hematoxylin and eosin, 40×

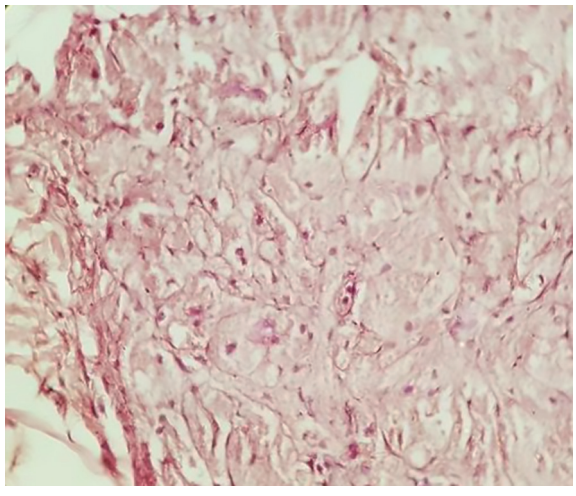

Fig. 4: Special stain (PAS)

eosinophilic cytoplasmic granules filling the cytoplasm, and small, round-to-oval nuclei. The lesion was rather well circumscribed, but unencapsulated (Fig. 3). So it was reported as GCT and we confirmed it with special stain PAS (Fig. 4). The patient was under follow-up. No recurrence was found during the course.

DISCUSSION

Abrikossoff was the first to describe GCT by the name granular cell myoblastoma. Granular cell tumors commonly develop in soft tissues. They have a frequency of 0.5% among soft-tissue tumors, and they chiefly materialize in or under the skin, or in the submucosa of the chest wall, bronchus, throat, and tongue.[3,8]

Tumors occurring in the systemic organs are diagnosed later than the skin lesions, as they are asymptomatic. The tumor occurs commonly in the age between 10 and 50 years with women predilection with a proportion of about 2:1. However, the frequency in adults is particularly high. Over 60 years of age, it is rare. The tumor is usually presented as a slow, growing, solitary mass, which is asymptomatic. Malignant variant of GCT is reported in about 2% of all cases.

A benign tumor will usually measure below 3 cm. However, if the tumor progression is rapid and develops an ulcer, malignancy could be doubted. The GCT diagnosis is confirmed through pathology tests. The tumor shows existence of a large volume of dense cytoplasmic lysosomes in different fragmentation phases, which gives a granular form under microscopy. The disease displays itself in the form of subdermal or submucosal nodules and the cells will be organized in diffuse masses and strands.

The GCTs are not encapsulated and have imprecise borders. They might invade and infiltrate nearby tissues. The tumor cells are large fusiform or polygonal which shows a distinct cytoplasmic membrane and a pale cytoplasm, filled abundantly with acidophilic granules.[3,9] Cell nuclei are small, oval to round vesiculated, seen centrally, and some cells may show more than one nucleus.

The typical granules seen inside the cells are PAS-positive, diastase-resistant, and phosphotungstic acid hematoxylin negative. In immunohistochemistry, GCTs are positive for the protein neuron-specific enolase, CD-68 and S-100. The most typical feature of granular cells is the membrane-bound cytoplasmic granulation with microvesicles of various densities resembling lysosomal structures along with microtubules, and myelinic developments.

The presence of sphingomyelin and lipoproteins seen in granular cells shows that the granules in the polygonal cells contain myelin or the result of its degradation, and positivity for enolase, myelinic proteins PO and P2, and protein S-100 by immunoperoxidase techniques.

From a histopathologic point of view, Smith et al suggested the following six standards to determine the malignancy of a tumor which include: (1) The appearance of spindle cells, (2) the presence of necrosis, (3) enlarged vacuolar nucleus (4) increase in mitosis (two or more mitoses/10×), (5) increase in the nuclear cytoplasmic ratio, and (6) polymorphism.[4,7]

Note: If none of these norms are seen, the tumor can be reported to be benign; if positive for one or two diagnostic norms, the tumor is considered to show atypia; if three or more criteria are positive, the tumor is confirmed to be malignant.[4,9]

Granular cell tumor shows a rare occurrence and mainly occurs as a solitary nodule on the skin, oral cavity, and the tongue. The disease arises in the skin in 30 to 40% of cases, then the head and neck region, where they are mostly seen in the soft palate, the hard palate, and the tongue. In the whole body, it often arises as subdermal tumors in and under the skin.

In the oral cavity, it has been described to appear submucosally in the buccal mucosa, floor of the mouth, palate, lower lip, tongue, and other tissues. Other locations include the thyroid gland, the respiratory tract, the gastrointestinal tract, the urinary bladder, female genitalia, the central nervous system, and the breast. Granular cell tumor in the breast is often misdiagnosed as invasive ductal carcinoma.[4]

It is diagnosed based on the microscopic findings because the presence of a submucosal nodule is the only subjective symptom exhibited with no apparent clinical symptoms induced by this tumor. Theories like histiocytic, fibroblastic, and myogenic have been discussed to explain the histogenesis of this tumor.[10] Based on the mechanism of elucidation by which it originates, GCT is considered as a true neoplasm by various researchers.

Intraorally, this disease often arises in regions which receive chronic mechanical trauma like buccal mucosa, tongue, and the gingiva. So, it may not be a true neoplasm, but can be a lesion based on metabolic abnormality, degeneration, and reactive growth. Since the tumor occurred in the labial mucosa in the present case, the possibility that the tumor was a reactive lesion cannot be ruled out. Our case also showed connective tissue stroma with sheet-like arrangement; it showed large polygonal cells with ample eosinophilic granular cytoplasm and a prominent nucleus intermixed with collagen fibers and extravasated red blood cells. Overlying epithelium was not seen.

Tumors are usually wide resected, involving adjacent tissues are advocated, as they are unencapsulated and are found to be quiet effective. The recurrence is extremely rare in benign GCT. However, on the contrary, malignant GCTs are associated with increased rate of recurrence and metastasis and mortality rate of 40%.[5]

CONCLUSION

Granular cell tumor is a tumor with an extensive variety of features and architectural patterns. Both benign and malignant lesions have been reported; hence, careful postsurgical histopathological examination and follow-up are required.

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