A confusional tumour of the tongue

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ABSTRACT

Granular cell tumour (GCT) is a fairly rare benign lesion typically seen in intra oral sites though there are several cases reported in extra oral sites. A review of the literature also reveal malignant versions of Granular cell tumours. This tumour has been coined with different terminologies due its controversial histological origin. We report a case of a granular cell tumour of the tongue in a 37 year old male patient.

INTRODUCTION

The history of granular cell tumours dates back to 1926 when Russian pathologist Alexi Ivanovich Abrikossoff supposed the tumour to be of muscular origin and termed it granular cell myoblastoma. Though its histogenic origin remains unclear, the finding of nerve sheath differentiation in these tumours has confirmed their neural origin. GCT typically presents as a solitary tumour but cases of multicentric presentations have also been reported.

CASE REPORT

A 37 year old male patient presented to us with the complaint of swelling over posterior aspect of tongue for the past 1 year which was progressively increasing in size. He did not have complaints of associated pain, bleeding, or dysphagia. An intra oral examination revealed the presence of a hard hemispherical non tender submucosal swelling over posterior aspect of tongue in midline. The patient underwent excision biopsy of the tumour. Sections showed tissue lined by stratified squamous epithelium with marked pseudoepitheliomatous hyperplasia with formation of occasional keratin pearls. Sub epithelium showed nests of granular cells which was seen extending between the muscle bundles. IHC studies showed S100 positive in the tumor cells and Ki-67 positivity index was low. Thus a diagnosis of granular cell tumour was made.

DISCUSSION

Granular cell tumours are neoplasms that more commonly affect females in the 4th-5th decade of life. These lesions may mimic several other benign lesions which include lipomas, dermoid cysts, benign mesenchymal neoplasms or neuromas. 70% of these lesions affecting the head and neck region are intraoral. Other sites affected are varied including the skin, nervous system, gastrointestinal tract, urinary bladder, female reproductive tract, bronchus. Majority of these tumours are asymptomatic and measures not more than 3 cm. 1-3% of the GCTs present in a malignant way.

An AFIP study stated any 3 of the following 6 criteria for the tumour to be categorized as histologically malignant

1) > 2 mitoses/10 fields at 200X magnification
2) necrosis
3) high nuclear-cytoplasmic ratio
4) spindling
5) vesicular nuclei with large nucleoli
6) pleomorphism.

It is classified as atypical GCT if 2 of the above mentioned criteria are met with.

Cases of coexistence of malignant and benign lesions have also been cited in literature. The key to differentiate between a benign and malignant lesion includes particular note of the tumour size, rapid progression of swelling, invasion of adjacent structures and distant metastasis. Unlike other malignant conditions there is no universally accepted staging system for these tumours.

Surgical excision is the treatment of choice but extent and location of tumour as well as lack of capsule may not always allow an excision with a safe margin. Other modalities of treatment like radiotherapy and chemotherapy have not proven to be effective in curing these lesions.

The characteristic marked pseudoepitheliomatous hyperplasia is deceiving as they closely resemble carcinoma. Definitive diagnosis is made by immunohistochemistry as the tumour cells react strongly with S 100 protein and thus considered precursors of Schwann cells. Other immunohistochemical analyzers include myelin base proteins, CD-68, P75, neuron specific enolase. A low Ki 67 index is a good prognostic factor but if Ki 67 is > 10% it is indicative of malignancy.

Usually recurrences are uncommon if surgical resection is adequate. But literature states that recurrence rates can vary from 2-50% and sometimes even noted several years later.
based on the infiltrative pattern\textsuperscript{15}. Thus regular follow up of these patients are needed to rule out malignant transformation but the slow growth and rare aggressiveness of the tumor lend it a good prognosis\textsuperscript{17}.

Figure 1: Hematoxylin and eosin stain showing marked pseudoepitheliomatous hyperplasia

Figure 2: S 100 positivity

Figure 3: Cytokeratin uptake which is negative in granular cells but positive in the epithelium
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