Ectomesenchymal chondromyxoid neoplasm. An unusual presentation. A case report

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**A R T I C L E   I N F O**

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**A B S T R A C T**

Ectomesenchymal chondromyxoid tumor (ECMT) is a rare benign intraoral tumor. Clinically, it presents as a slow growing, painless, firm, submucosal swelling exclusively occurring on the anterior dorsum of the tongue. Till date not more than 45 cases have been reported in literature. Histopathologically the tumor is characterized by a well circumscribed, lobular proliferation of round, polygonal, ovoid or fusiform cells in a net-like pattern in a myxoid to chondromyxoid background. Here, we present a rare case of ECMT occurring in a 17 years old male who presented to us with chief complaint of slow growing painless mass arising from anterior Dorsum of the tongue measuring about 1.5 cms × 1.5 cms. Mass was completely resected as excisional biopsy, it was found unencapsulated with muscle entrapment.

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1. Introduction

ECMT is a rare benign tumor occurring as a submucosal swelling on the anterior two third of the dorsum of the tongue. It was first described by Smith et al. in 1995 and only 45 cases have been reported till 2010 [1]. The tumor is thought to arise from undifferentiated ectomesenchymal cells, hence the name. They typically present as unencapsulated though well circumscribed tumor masses Muscle entrapment within the tumor can be seen and should not be confused as malignant behaviour of the tumour [2–5].

These cases are typically positive for S100 protein and GFAP Immunohistochemical stains. The tumor usually exhibits positivity for Cytokeratin,GFAP and other myoepithelial markers such as SMA. This lesion usually affects the anterior 2/3rd of the dorsum of the tongue [12,14,15]. Age of affected person ranges from 9 to 78 years. Both Males and Females are affected equally.

Treatment of choice of ECMT is conservative surgical excision but rarely it may recur. The chances of recurrence and the histopathological evidence of Foci of pleomorphic hyperchromatic cells, sporadic mitotic figures and muscle infiltration suggest the importance of regular follow up of patients after treatment [6–11,13,16].

This work has been reported in line with the SCARE criteria [17].

2. Case report

A 17 years old male presents to our ENT Opd with painless swelling on the anterior dorsum of the tongue of approximately 7 months duration. On Examination, a solitary well defined submucosal mass measuring 1.5 cms × 1.5 cms in diameter located on the right side of the dorsum of the tongue approximately 1.5 cms from the tip and 1 cm from the lateral border was found. The mass was firm in consistency, mobile and non-tender. The Rest of oral, Head and Neck examination was within normal limits.

FNAC was taken and was reported as unremarkable. Surgical Excision was performed and the specimen was about 1.5 × 1.5 × 1 cms in dimensions.

Specimen was send for Histopathological Examination and was reported as Ectomesenchymal Chondromyxoid Neoplasm.

The hematoxylin and eosin stained sections of the formalin fixed and paraffin embedded tongue mass show an encapsulated (Fig. 1), nodular lesion composed of rounded to ovoid and polygonal cells with eosinophilic and “frothy” cytoplasm. Some cells showed intracytoplasmic vacuoles (Fig. 2). Focally, the lesion demonstrated net-like growth pattern with slit-like spaces (Fig. 3). Multinucleated tumor giant cells were seen (Fig. 4) although the tumor lacked in calcifications. Focal myxoid stroma was present although no chondroid matrix was seen in our case. Mild nuclear atypia and pleomorphism were also present in this case.

Our tumor showed focal S100 protein positivity, focal neuron specific enolase (NSE) positivity (Fig. 5), and negative staining with AE1/AE3 immunohistochemical stains (IHC).

Immuo-Histochemistry was performed and the tumor cells were found to be positive for S100, SMA and NSE and negative for
Fig. 1. Lower magnification of the encapsulated tongue mass (20X; H&E).

Fig. 2. The tumor is composed of rounded of ovoid cells with eosinophilic, somewhat frothy, cytoplasm with scattered cells having intracytoplasmic vacuoles (200X; H&E).

Fig. 3. Although focal, the lesion demonstrated net-like growth pattern with slit-like spaces (200X; H&E).

Fig. 4. Multinucleated giant tumor cells were also evident although no calcifications were noted in our case (100X; H&E).

Fig. 5. Neural immunohistochemical markers like neuron-specific-enolase (NSE) were focally positive (Inset: positive s100 protein marker).

CK 5/6, HMB45, Desmin and GFAP and highlights vessels for CD31 and CD34, CD68 and low Index (<5%) for Ki-67.

As this particular case is positive for s100 protein and negative for GFAP. Interestingly, the tumor usually exhibits positivity for cytokeratin (Though negative in this particular case) GFAP and other myoepithelial markers such as SMA, a fact that suggests a possible myoepithelial phenotype/differentiation.

3. Follow up

At 3 month follow up, Pt. was improved without any complications/Recurrence.

4. Conclusion

ECMT although a rare entity should be considered in the differential diagnosis of nodular lesions of the tongue dorsum. Although our case showed negative results to GFAP and positive for S100 protein and exhibited positivity for some myoepithelial markers such as GFAP as was described above. The immunoprofile of these lesions need further definition as more cases are added to the literature.
Therefore, increased awareness of this distinct entity may lead to a better insight into the clinical behaviour and also in understanding its histogenesis.

Conflicts of interest

No conflict of interest to declare by any of the authors.

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Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available and can be provided whenever needed.

Author contribution

Dr Abdulmohsen; data collection, writing the paper.
Dr Hussein; data analysis and contribution.
Dr Intiyaz; data collection, writing paper, study concept.

Guarantor

Dr Abdulmohsen and Dr Intiyaz.

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