CASE REPORT

Ectomesenchymal chondromyxoid tumor of the anterior tongue: A rare case

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Abstract Ectomesenchymal chondromyxoid tumor is a rare tumor seen in the anterior dorsal aspect of the tongue. Diagnosis of this lesion without a prediagnosis clinically is made only with histopathological findings including the immunohistochemical examinations. However, diverse results are being reported in the literature with keratin staining. While glial fibrillary acid protein positiveness maintain being a fixed repeating feature, we emphasized for our case of a 28-year-old Caucasian woman having the findings of qualities supporting the ectomesenchymal origin of the mentioned tumor.

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Introduction

Ectomesenchymal chondromyxoid tumor (ECT) of the anterior tongue was firstly reported by Smith et al. [1] in 1995. It was a unique chondromyxoid lesion. Besides this, myxoid and chondromyxoid ones of the oral soft tissues usually form a heterogeneous group [2]. The mesenchymal tumors having myxochondroid features have been grouped under the termination of ECT and most of them have been detected in the anterior tongue [1]. Clinically, ECT exhibits a slowly growing, firm, well-circumscribed painless nodule on the anterior part of the tongue. Histologically, it is characterized by well-limited lobular non-capsulated proliferation of fusiform, oval, spindle, and polygonal cells on chondromyxoid ground frequently having multilobulated nuclei and foci of atypia [3,4]. Immunohistochemically, tumor cells are reactive for glial fibrillary acidic protein (GFAP) and cytokeratin, and less frequently for smooth muscle actin, S-100 protein, and CD-57 (Leu-7). However, tumor cells are not reactive for desmin and epithelial membrane antigen (EMA) [4]. While GFAP positiveness maintain being a fixed repeating feature, we emphasized

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for our case of a 28-year-old Caucasian woman that incorporation of muscle fibrils to the lesion is an important characteristic in the differential diagnosis as well as being lobulated with well-defined borders. Findings of the present case have qualities supporting the ectomesenchymal origin of ECT.

**Case presentation**

A 28-year-old Caucasian female presented with a smooth and painless mass lesion on the anterior dorsal side of the tongue that grew progressively for the last 1 year. A painless and immobile mass lesion with regular borders, diameter 0.5 cm and elastic consistency was found in the oral examination on the anterior dorsal side of the tongue. No other notable finding was present in the examination of mouth, head, and neck. Excisional biopsy was performed under local anesthesia, and soft tissue was observed in the gross examination with size 0.5 × 0.5 × 0.5 cm covered with intact mucosa. The entire lesion was removed according to gross examination.

In the Hematoxylin & Eosin stained slides, a lesion was observed that grew by pushing the surrounding tissues but with well-defined borders. Borders were sharply separated from the surrounding tissue and have a lobular appearance with the thin collagen fibrous septa (Fig. 1). It was separated from the intact mucosal epithelium with a thin layer consisting of compacted collagen bundles and fibroblasts. Tumor was created from the cup-shaped, ovoid, and fusiform cells on the chondroid ground surrounding in the center, and from the stellate cells in the myxoid ground fully surrounding this chondroid area (Fig. 2). Pseudo-infiltration of the muscle fibrils was recognized in one area of the tumor. No any nuclear atypic characteristics, mitosis, necrosis, hemorrhagia, calcifications, osteoid areas, and intranuclear pseudooinclusion were seen as reported in the literature. We have not detected any inflammatory cell infiltration, and there were no minor salivary glands in the surrounding tissue. While cells of the specimens displayed the diffuse staining with GFAP and S-100 immunohistochemically, they were negatively stained with epithelial markers pankeratin and EMA. Besides, they were focally stained with vimentin and actin. Finally, ECT diagnosis was made based on histopathological, clinical, and immunohistochemical characteristics.

**Discussion**

In the presence of the cartilaginous lesion in the oral soft tissue, the possibility of chondroid pleomorphic adenoma must be considered particularly if the lesion is well defined and directly in contact with salivary glands. The immunohistochemical stains of cytokeratin and EMA appear useful to determine any epithelial component in the lesion in such cases [5].

Salivary glands were not observed in the serial cuts which were performed on the present case. Calcification is marked in the center which is frequently seen only in extraskeletal chondromyxoid tumors (ES-CT) that are generally found in hands and feet and that are frequently related to the tendon sheath and joint capsule. Granuloma-like proliferation of the epitheloid cells and multinuclear giant cells can be seen. While these characteristics were not present in our case, the cellular appearance of ES-CT with a rather mature peripheral region and with lesser central chondroid tissue also was not observed. Contrary to the mature and well-differentiated hyaline cartilage surrounded with connective tissue in cartilaginous choristoma and capsule with osteoid and bone formation in the ossifying fibromyxoid tumor, which has never been reported with localization in tongue, chondroid tissue surrounded with myxoid areas and the pseudo-infiltration of myofibrils at the sides of the tumor are notable.

In cases with malignant characteristics, such irregular nuclear morphology, increased cellularity, lack of intercellular matrix formation, mitosis, and infiltration to the surrounding tissues, which were not present in our case, are seen in the oral soft tissues; then primary chondrosarcoma or metastasis of osseous chondrosarcoma is more frequently seen, but nevertheless, still rather rare.
Glial choristoma and myoepithelioma including the chondromyxoid variants are included in the differential diagnosis immunohistochemically and as regards GFAP positiveness. It can easily be differentiated from glial choristoma thanks to lack of astrocytes, ganglion cells, and other neural elements and also from myoepithelioma thanks to lack of spindles or plasmacytoid cells, ductal structures, and salivary glands.

The most important item in the differential diagnosis is ECT. It appears in the form of a painless nodule with lobules and well-defined borders consisting of round, cup-shaped, fusiform, ovoid or polygonal cells with uniform small nuclei, which can be atypical sometimes, and medium-slight basophilic cytoplasm arranged as cords, columns, or layers on myxoid base. Among the differentiating characteristics, inclusion of muscle fibriles within the lesion can be an important clue as well as GFAP-positiveness is fixed \[1,3\] and this requires to make the serial cuts from the lesion.

It is important to make the diagnosis of ECT by eliminating all the differential diagnoses one by one. However, we have the opinion that it would be appropriate if the differential diagnoses, which are given within a very wide spectrum in the literature, are limited only with ES-CT, cartilaginous choristoma, chondroid pleomorphic adenoma, myoepithelioma including chondromyxoid variants and extra-skeletal myxoid chondrosarcoma among other chondromyxoid lesions of the head and neck region \[6\].

There is no consistency in stains related to epithelial markers (pankeratin, EMA) in publications after the report of Smith et al. \[1,4,6\]. In our study, staining with pankeratin and EMA was also negative. Vimentin and actin expressions confirm mesenchymal origin and S-100 and GFAP expressions confirm neurogenic origin. Presence of myxoid and chondroid regions indicates the multi-potential characteristics of the cells.

In the embryological sense, these grow by the proliferation of the mesenchymal tissues in the ventromedial aspect of the first pharyngeal arch under the lateral lingual protuberances on both sides of the tuberculum impar. These formations undergo fusion so as to cover tuberculum impar also. Two-thirds (anterior part) of the tongue is thus formed. As explained, connective tissue of the tongue differentiates from the mesenchyma of the first pharyngeal arch.

On the other hand, pluripotent neural crest cells immigrating to pharyngeal arches differentiate later to the cartilage tissues of the head—neck region (like the Meckel’s cartilage in the first pharyngeal arc) \[1\]. It is possible that ECT originates from these pluripotent neural crest cells. These cells may behave like multi-potential cells and differentiate to myxoid and cartilaginous cells.

Consequently, ECT is a unique chondromyxoid tumor and the diagnosis is only confirmed with histopathological evaluation considering immunohistochemistry. Immunohistochemical reactivity and findings of histopathological examination confirm neural and chondroid differentiation as reported in the literature. The preferred treatment modality remains the surgical excision with the minimal margins of the lesion. In our opinion, a clinician must be awake for the possibility of the ECT in the differential diagnosis of the cases having the nodular lesions on the tongue. Increased awareness may create the better insight for these types of lesions.

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References


