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Case report

Glial choristoma of the ventral part of the tongue: first report in an elderly patient

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Key words

Choristoma • Glial tissue • Tongue

Summary

Introduction. The aims of this report are to illustrate the first case of glial and adipose tissue choristoma at the ventral part of the tongue in an elderly patient, to discuss the possible differential diagnoses and to speculate about its pathogenesis.
Case report. A 65-year-old female was admitted to our hospital with a swelling at the base of the tongue. MRI revealed an oval lesion with indistinct borders without contrast enhancement. The patient underwent surgical complete excision and grossly, the specimen consisted of a gray-white mass measuring 25 mm in its great diameter. Microscopically the lesion contained fibrocollagenous stroma, mature adipose tissue and mature astrocytes. In the absence of cellular atypia, mitoses and necrosis a diagnosis of adipose and glial choristoma was performed. The patient is healthy 18 months postoperatively.
Discussion. Choristomas are cohesive tumor-like masses histologically composed by normal tissue occurring in an unusual anatomical location and mainly affecting children during the first years of life. Glial choristomas are considered malformations of the central nervous system and their localization in the tongue is exceptional. However they carry a favourable prognosis so it is of paramount importance to histologically diagnose them correctly.

Introduction

Choristomas of the head and neck region are cohesive tumor-like masses histologically composed by normal tissue occurring in an abnormal anatomical location. Several different tissues can occur, like salivary gland, cartilage, bone, glial or glioneuronal tissue, gastric or enteric mucosa and thyroid parenchyma and choristomas are classified accordingly, based on the tissues they are composed by [1].
Glial choristomas are usually considered malformations of the central nervous system, affecting mainly newborns, infants and children, with a typical localization in nasal cavity, the so called nasal glioma [2]. However, lingual localization has been described, though extremely rare, with the lesion being generally located on the dorsum of the tongue [3]. In this setting, a few studies described it also in association with cleft palate [4].
Glial choristoma has a favorable course and can be treated by surgical excision alone. The most important point is to diagnose it correctly, differentiating it from many entities like hamartomas and from true neoplasms [1].
The aims of this report are to illustrate the first case of glial and adipose tissue choristoma at the ventral part of the tongue in an elderly patient, to discuss the possible differential diagnoses and to speculate about its pathogenesis.

Case report

A 65-year-old woman was admitted to our hospital complaining a swelling at the base of the tongue. Her past medical history was unremarkable. Clinical examination revealed a firm and well circumscribed mass, located at the midline ventral zone of the tongue (Fig. 1A). Magnetic Resonance Imaging (MRI) revealed an oval lesion without contrast enhancement. Signs of intrusion of adjacent muscles were observed. The patient underwent a surgical complete excision (transoral enucleoresection) of the lesion and the sample was sent to the Pathology Lab for histological examination. Grossly, the specimen consisted of an unencapsulated tender mass measuring 25 mm in its major axis (Fig. 1B): the cut surface was yellowish and smooth. Microscopically the mass showed a globular ap-

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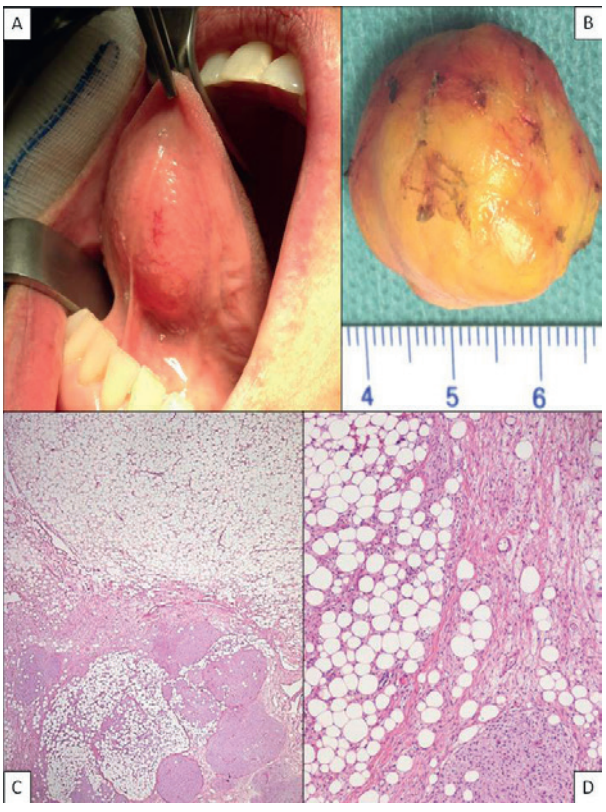

Fig. 1. A: Clinical examination revealing a firm and well circumscribed mass at the midline ventral zone of the tongue; B: Macroscopic appearance of the mass; C: Histological appearance at low magnification, mature adipose tissue intermingled with nodules of glial elements (H&E, 2x); D: Higher magnification showing astrocytes with amphophil cytoplasm and bland, oval nuclei intermixed with mature adipose tissue (H&E, 10x).

pearance and was composed by fibrocollagenous stroma containing several islands of elongated elements with amphophil cytoplasm and bland, oval, vescicular nuclei (Fig. 1 C-D). Immunophenotypic profile was determined by a panel of antisera including pS100, Ki67, CD34 and GFAP (Ventana) and MDM2 (Life Technologies). The above mentioned cells showed strong expression of glial fibrillary acidic protein (GFAP) and were therefore interpreted as mature astrocytes; no neural element was observed. The glial nests were intermingled with mature adipose tissue showing immunohistochemical positivity for pS100 protein and negativity for MDM2. The lesion intruded the surrounding skeletal muscle and was superficially lined by stratified squamous epithelium. Neither cellular atypia, nor mitoses nor necrosis were detected and therefore a diagnosis of adipose and glial choristoma of the tongue was performed. The postoperative course was uneventful and the patient is alive and healthy 18 months postoperatively.

Discussion

Choristomas are composed by normal tissue occurring in an abnormal anatomical location and are characterized by an excellent prognosis without recurrence after complete surgical removal.

Glial choristoma in the head and neck region has been named throughout the literature as “heterotopic brain tissue”, “extracranial glioma” or “teratoma”, but these terms are unsatisfactory. Therefore the definition of “glial choristoma” proposed by Chou is actually favored encompassing both the presence of normal ectopic glial tissue and its tumor-like growth pattern [5].

This entity was first highlighted by Reid in 1852 and in 1922 Peterer described its occurrence in the tongue [6], a very unusual site with only 18 reported cases (including the present) usually affecting newborns, infants and children. In fact glial choristomas are usually present at birth or reveal themselves during the first years of life since they grow in an expansively pattern with a rate similar to the normal surrounding tissue [7]. The peculiarities of our case are the patient’s age and the anatomical site of occurrence: the present case is the first description of a glial choristoma manifesting itself in an elderly patient at the midline ventral tongue, instead of posterior part of the lingual dorsum [3].

From a histological point of view, we had a wide range of entities to rule out in order to achieve the correct diagnosis. We had to keep in mind the advanced patient’s age during which the probability of true neoplasms is higher than that of malformations. In the first setting, we ruled out adipose tissue tumor as spindle cell lipoma and atypical lipomatous tumour since we did not observe ropey collagen bundles, or lipoblast or CD34 or MDM2 immunohistochemical positivity. Concerning teratoma, the lack of an endodermal component contributed in dismissing this hypothesis. Squamous cell carcinoma, granular cell tumour, neurofibroma, hemangioma and other soft tissue neoplasms were also excluded basing on morphology and immunohistochemistry. The possibility of developmental malformations, like hamartomas (i.e. tumor-like malformations composed of an overgrowth of mature normal cells normally located) and thyroid duct cysts, was also rejected since the oral cavity is an ectopic location for glial tissue and no cystic changes or thyroid follicles were observed.

Due to the rarity of glial choristomas, generally considered a congenital malformation, the extact pathogenesis of this entity is still not clear and various theories have been proposed.

The first one is that of herniation of the fetal cerebral tissue, like encephalocele but with separation from the cranial cavity. The *locus minori resistentiae* of this theory is that the tongue is far from known embryonic canals [1].

The second proposed hypothesis is that glial choristoma may develop from the neural crest cells in the head and neck which have the ability to undergo neuroglial development. Another theory that may explain the occurrence of glial choristomas in the lingual location is that of the separation of cells from the anterior part of the brain in early embryonal development and particularly from the occipital somites from which tongue muscles originate. It seems that the nests of pluripotential cells become separated prior

to complete fusion of the neural tube and are brought to the periphery in association with normally migrating cells [1]. Herein we report the first case of glial and adipose tissue choristoma occurring in an elderly female patient. Moreover we argue that, although very rare, it is important to make the correct histopathological diagnosis especially in such a clinical setting where the advanced patient's age favoured the hypothesis of a true neoplasm. In our case, the wrong diagnosis might have led the patient to an incorrect surgical overtreatment, with possible functional impairment, as for example decreased ability of speech. Adipose and glial choristomas of the tongue have an excellent prognosis and the surgical conservative excision is appropriate and curative, in fact, our patient is alive and healthy 18 months after surgical treatment without recurrences.

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