Unusual lesion of the temporal region, benign tumor of the Chievitz’s organ: a case report

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Abstract
Normally present in the temporal region, the juxtaoral organ of Chievitz has considerable importance for both the surgeon and the pathologist. Although very rare, the nodular hyperplasia of the Chievitz’s organ has been misinterpreted as invasive carcinoma. The pathological aspect mimics the mucoepidermoid carcinoma, squamous carcinoma, basal cell carcinoma with sebaceous differentiation or tumor of the skin appendages. We present the case of a 71-year-old patient admitted with temporal tumor. The initial pathological diagnosis was mixed squamous basal cell carcinoma. The subsequent evaluation of sections and immunohistochemistry allowed us to demonstrate a benign tumor of the Chievitz’s organ. In this case, therapy and prognosis are strongly related to the correct diagnosis and avoid an over treatment of a benign tumor.

Keywords: juxtaoral organ of Chievitz, temporal region, benign tumor.

Introduction
The juxtaoral organ is less known in histology and pathology, in spite of the fact it was described many years ago by Johan Hendrik Chievitz [1]. It is a small structure located within the soft tissue of the buccotemporal space. Today, it is believed to represent a primordium of the parotid gland but without true connection with the parotid. It is situated close to the fascia of the buccotemporal muscle. Microscopically, it is made up of an epithelial parenchyma, embedded in a highly organized connective tissue rich in nerves and sensory receptors [2].

The tumor that arises from the juxtaoral organ is very rare. Until now, only benign nodular hyperplasia was reported. Its sprouts, cell nests, gland-like structures or even follicles, lymphatic vessels admixed with small branches of the buccal nerve have been misinterpreted as squamous carcinoma with perineural invasion, mucoepidermoid carcinoma or tumor of epidermal appendages.

In our opinion, the knowledge and understanding of this lesion is extremely helpful for the pathologist.

Patient Methods and Results
A 71-year-old man was admitted with a right, temporal, cutaneous tumor. The symptoms included a painless, ulcerative tumor of the temporal region that involved all skin layers. Over one year before admission, the tumor had been 2 cm in its larger diameter and gradually grew, extending towards the right temporal region, that induced facial asymmetry.

The macroscopic examination revealed a round invasive tumor with firm borders having 5/3.5 cm in diameter. The tumor was firmly adherent to the skin but remained mobile towards the deeper layers. No regional lymph nodes enlargement was noticed and salivary secretion was not altered. All basic biochemical investigations had normal values.

After obtaining the informed consent of the patient, the tumor excision was decided. The surgery was performed under general anesthesia and consisted in excision of the tumor in clinical oncology safety limits. Plastic surgery was performed in the second stage. The tegumentary defect was covered with a free skin graft (Figure 1a). The patient was monitored for two years, the control being performed at one month, six months and then annually. Clinical and paraclinical examinations presented no local tumor recurrence or presence of metastases.

The specimen was fixed in buffered formalin and embedded in paraffin. The tissue sample prepared in paraffin was sliced by using a microtome, thus obtaining 4 μm histological sections, which were applied on specially prepared glass slices covered with a layer of positive charged amino acid waste (poly-L-Lysine) (Sigma) in order to increase the adherence of the samples to the slices.

For current pathological diagnosis, cross sections were stained with Hematoxylin–Eosin (Figure 1b) and Gömöri’s trichrome methods. Additional sections were immunostained for pancytokeratin (clone MNF116) cytokeratin 8 (clone 38BH11) (Figure 3a), chromogranin A (clone DAK-A3) (Figure 3b), neuron specific enolase (clone BBS/NC/VI-H14), S100 protein (Figure 2a), α-smooth muscle cell actin (clone 1A4) (Figure 2b), leukocyte common antigen (clone 2B118PD7/26), carcinoembryonic antigen (clone 11-7), and vimentin (clone V9). Both LSAB2 and EnVision system (DAKO, Denmark) were used. The most suggestive images are presented in (Figures 2 and 3).

Postoperative evolution was favorable, with no signs of local recurrence or distant metastasis.
Histological examination revealed the presence of cell nests with scattered gland-like and tubular lumens. We noticed a lobular arrangement of epithelial cells, sometimes forming keratotic pearls, isolated spindle cells and many capillaries were found between cords of epithelial cells. Around and within the nests of epithelial cells many small nerve fibers and a rich network of lymphatic capillaries were found. At first glance, these findings suggested an invasive tumor and the first diagnosis was squamous-basal cell carcinoma. On the other hand, mitosis were rarely found, without cellular atypia. The stroma connective tissue was rich in fibroblasts, plasma cells, macrophages and blood vessels.

These controversial aspects found on routine stained slide, lead to the idea of the presence of a benign tumor of the juxtaoral organ. The immunohistochemical profile revealed no reactivity with pancytokeratin and only moderate reaction with cytokeratin 8 (Figure 3a). An intense reaction was noticed with both chromogranin A and neuron specific enolase (Figure 3b). S100 protein and alpha-smooth muscle cell actin were expressed only in isolated cells of the stroma and in the spindle or “dendritic” cells of the epithelial nests (Figure 2a). The other immune reactions were negative in the tumor cells.

**Discussion**

The juxtaoral organ of Chievitz is normally present in temporal region. It is a rather small organ that measures 0.7–1.7 cm in length and 0.1–0.2 cm in diameter [3, 4].

The organ of Chievitz is multilobated and has a connective fibrous capsule and the stroma is fibrous, containing many nerve fibers. The parenchyma consists of nests of squamous epithelial cells that sometimes form pseudolumens. Although the epithelial component is located in the deeper part of the region, step sections show the branched and continuous arrangement [3, 4]. The epithelial cells of the large nests have clear cytoplasm and nuclei are oval or round in shape. Intracellular bridges may be noticed between epithelial cells and small nerve fibers of the facial nerve creates the false impression of perineural invasion of a squamous cell carcinoma.

The recognition of epithelial nests with nodular hyperplasia that belong to the juxtaoral organ may define the therapeutic strategy and prognosis. There are some reports of nodular hyperplasia of the juxtaoral organ in the literature but carcinoma of the juxtaoral organ was not yet described [6–9]. In spite of the benign character, Eversole and Leider [10] described intraosseous maxillary extension of the lesion.

The function of the organ of Chievitz is not understood [11]. The Danish anatomist Johan Hendrik Chievitz described the organ in 1885 in a 10-week-old human embryo. In spite of the fact that it is known for more than one hundred years, there is extremely few data about the function of this organ.

According to certain authors [12], the above-mentioned structure is being formed during the embryogenensis process through the proliferation of an epithelial bud of the oral mucosa, that gets barred in the subjacent tissue like a diverticulum, then grows by creating itself a conjunctive stroma and a capsule at its periphery, afterwards getting loose from the oral cavity and developing independently. The structure keeps and grows its own somatic and vegetative innervations and receives vascularization from the oral cavity vessels [13]. Other authors consider the Chievitz’s organ to be originally neuro-
epithelial like several other oral cavity structures [14]. Initially, it was assumed that this organ would be present only during the embryogenesis process, afterwards disappearing. Many studies in the last seven decades proved the Chievitz’s organ to continue its development being frequently identified in children and adults [15]. However, because of its small dimensions, its deep localization similar consistence as its surrounding tissues, therefore the considered structure cannot be clinically identified [16]. It becomes clinically evidenced in children and adults when its dimensions exceed 10 mm following a hyperplasia or a tumor growth process [7, 17–19].

In our study, the stroma cells showed an intensely positive reaction to chromogranin A and neuron specific enolase. For that, we consider the Chievitz’s organ a neuroepithelial structure. The particularity of our case is the rapid proliferation within a single year, reaching from 2 cm to 5/3.5 cm.

In our consulted references, we could not precisely identify the function and importance of the Chievitz’s organ. It was considered by some authors an abortive salivary gland due to intense activity of certain enzymes in the epithelial parenchyma (hydrolase, alkaline phosphatase and carbonic anhydrase) in similarity to salivary glands or a rudimentary structure by others. Relatively ultrastructural and immunohistochemical data suggest its sensorial [11] or neurosecretory function. Another peculiar behavior of this organ is that it develops benign tumor forms, not malignant ones [2]. This rare form of tumor has never been reported in our clinic in a period of over 30 years of anatomical and pathological examinations.

Conclusions
Rapid wound healing, the absence of recurrence and metastasis suggest the benign character of the lesion. Therapy and prognosis are strongly related to the correct diagnosis and avoid an over-treatment of a benign tumor. In this anatomical area, the differential diagnosis with an invasive mucoepidermoid carcinoma is also necessary. The pathological aspect mimics the mucoepidermoid carcinoma, squamous carcinoma, basal cell carcinoma with sebaceous differentiation or tumor of the skin appendages. We think that it is important to know about the organ of Chievitz in order to avoid confusion with invasive carcinoma of the temporal region.

References

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