Case Report

Neuroepithelial Structures Similar to Juxtaoral Organ of Chievitz Present in the Mandibular Torus of a 41-year-old Man

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The Juxtaoral organ of Chievitz (JOOC) is a normal neuroepithelial structure that is located in the pterygomandibular space. Neuroepithelial structures resembling those seen in the Juxtaoral organ of Chievitz in other locations are diagnostically challenging and can be potentially misdiagnosed as squamous cell carcinoma. Here, we report the first case of such structures presenting on the right lingual surface of the mandible of a 41-year-old man with torus mandibularis. Awareness of the possibility that neuroepithelial structures similar to the JOOC may be found in mandibular tori will prevent unwarranted costly diagnostic procedures and unnecessary surgical interventions.
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INTRODUCTION

The formation of head and neck structures results from precise in-utero events where molecular signaling and interactions between components of different germ layers result in coordinated histomorphological changes. While most embryonic structures undergo continuous and progressive changes that lead to fully developed tissue types, others persist for defined periods of time and then involute.[1] However, some embryologic structures persist well into adulthood and are commonly seen in and around the oral cavity. Examples of such structures of epithelial origin include dental lamina rests, rests of Malassez, nasopalatine duct remnants and the JOOC.[2]

Chievitz first described JOOC in 1885 while studying human embryos.[3] JOOC is not unique to humans and has been reported in other species as well. Typically, JOOC is located deep to the medial pterygoid muscle (unilaterally or bilaterally) in the pterygomandibular space. JOOC starts as an epithelial thickening of the stomodeum, which invaginates into the subjacent mesenchyme. This epithelial bud then detaches from the oral epithelium, becomes innervated by a buccal nerve branch and receives vascular supply from the buccal artery.[4]

We report the presence of neuroepithelial structures similar to the JOOC in a mandibular torus, a heretofore not previously described finding in the English literature.

CASE REPORT

A 41-year-old male presented with multilobular exostoses on the right lingual surface of the mandible. The patient was scheduled to receive a complete removable mandibular denture and thus, excision of the exostoses was done under local anesthesia. The surgical specimens were fixed in 10% buffered formalin. Multiple bony tissue fragments were received in our laboratory from two different but adjacent sites. The bony tissue fragments measured 1.5 x 1.2 x 0.5 cm and 2.0 x 1.8 x 1.5 cm, respectively. These fragments were decalcified overnight, formalin-fixed, paraffin embedded, sectioned at 5 μm and then stained with hematoxylin and eosin.

Microscopic examination revealed the presence of multiple fragments of cortical and trabecular bone, some of which exhibited a dome shaped configuration. Both, cortical and trabecular bone showed osteocytes, basophilic reversal lines and fatty bone marrow. In addition, fragments of surface oral mucosa and slivers of dense fibrous connective tissue with interspersed fibroblasts and capillaries were also noted. The most significant finding was the presence of an aggregate of epithelial islands, intimately associated with nerve bundles that were partially surrounded by perineurium. The epithelial islands consisted of oval to columnar basal cells with scarce

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eosinophilic cytoplasm and prominent ovoid to elongated nuclei with occasional nucleoli. Subtle nuclear palisading of the basal cells was observed in focal areas. No evidence of cellular pleomorphism or abnormal mitotic activity was noted. The surrounding connective tissue stroma was loose and slightly myxomatous, containing interspersed spindle mesenchymal cells and occasional scattered mast cells (**Figure 1** and **Figure 2**). Further characterization of the perineural epithelial structures was attempted with a number of immunohistochemical stains. Unfortunately, the epithelial structures we were attempting to characterize with the immunohistochemical markers were no longer present at deeper levels (pictures not shown). S-100 decorated the nerve fibers present in the specimen. However, synaptophysin was negative. AE1/3 stained fragments of surface oral mucosa that were present in the specimen. Ki-67 showed a low proliferative index for the epithelial fragments of the oral mucosa. Based on morphological features, a diagnosis of mandibular tori with neuroepithelial structures similar to the Juxtaoral organ of Chievitz was issued.

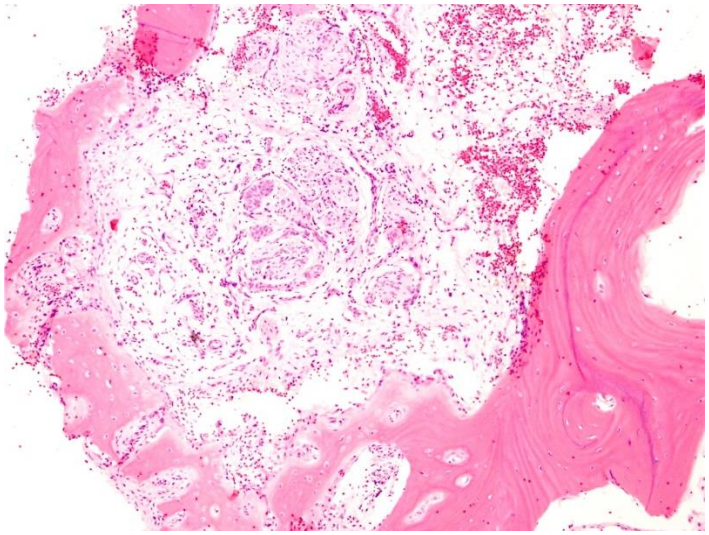

Figure 1. Low power view showing trabecular bone with interspersed fibrous connective tissue stroma. Epithelial islands are seen adjacent to neurovascular structures (hematoxylin and eosin, original magnification x400).

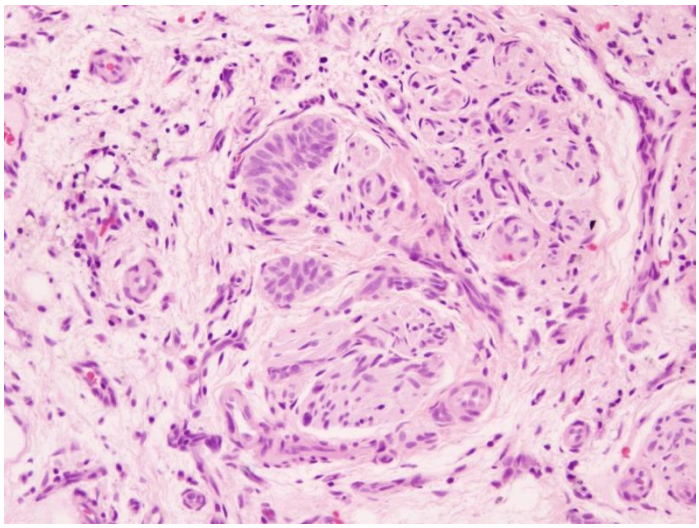

Figure 2. High power view demonstrating the presence of nests of epithelial cells with ample cytoplasm and slightly elongated columnar type of nuclei. Nerve fibers are seen in close proximity to the epithelial structures (hematoxylin and eosin; original magnification x200).

DISCUSSION

Chievitz thought that the neuroepithelial structures that bear his name were a transient fetal structure destined to involute prenatally.[3] However, Zenker reported that JOOC persists well into adulthood.[5] More recently, Mérida-Velasco in an extensive study of human embryos demonstrated the presence of a rich neurovascular complex of the JOOC capsule, a finding that militates against the notion suggesting that JOOC represents an involuted structure.[4]

JOOC is usually located in the pterygomandibular space, deep to the medial pterygoid muscles.[4] A typical JOOC should not be clinically palpable due to its small dimension and deep location. However, the threshold at which the JOOC becomes clinically evident is when it acquires a dimension of 10 mm or more in diameter.[3] Clinically evident lesions at intra-oral and extra-oral locations, resulting from hyperplastic JOOC are seen both in children and adults.[7-10] A superficial, submucosal locus in the ascending ramus of the mandible of a child that protruded into the oral cavity has been reported.[3] An unexpected location for JOOC has the potential to create a diagnostic challenge.

JOOC is characterized at the microscopic level by the presence of distinct epithelial and connective tissue structures.[10,12] The epithelial component consists of circumscribed nests of non-keratinizing squamous, columnar (glandular-like) and occasionally, basaloid epithelial cells. Three concentric domains of connective tissue encase the epithelial islands. The inner layer, stratum fibrosum internum, consists of dense collagen fibers that are separated from the epithelial islands by a distinct basal lamina.[12.13] The middle layer, stratum nervosum, is characterized by loose connective tissue stroma, populated with myelinated and nonmyelinated fibers.[12-14] The outer layer, the stratum fibrosum externum, connects to the muscle fascia of the buccotemporalis.[13]

Fontana–Masson stain has shown the presence of melanin pigments in some JOOC.[15] Alkaline phosphatase activity of the epithelial component of the JOOC and a possible mechanoceptor function due to close approximation of JOOC to structures resembling Pacinian corpuscles have also been documented.[12,16] In addition, a uniform intracytoplasmic

signal with pancytokeratin and selective staining of the center of epithelial islands with cytokeratin 19 have been reported.[6,7,15]

JOOC carries no risk for malignant transformation and no recurrence after its removal.[7,9,13] Eversole and Leider published in 1978 a case where they found neuroepithelial structures identical to the JOOC in the anterior maxilla of an individual.[11] Our case is similar to Eversole's case and documents a previously unreported location for these neuroepithelial structures. It is conceivable that these neuroepithelial structures originate from invaginations of the anterior mandibular embryonic oral mucosa in a process similar to the one described for the JOOC in the pterygomandibular space area. These neuroepithelial structures show an uncanny resemblance to the JOOC and have the potential to create a diagnostic challenge. However, the lack of epithelial pleomorphism and the presence of a well-defined basement membrane, help to separate JOOC from a squamous cell carcinoma showing perineural invasion.[13] At times, the glandular-like epithelial cells of JOOC can mimic a mucoepidermoid carcinoma (MEC).[14,17,18] However, the typical mucous cells, intermediate and squamous cells with characteristic histopathologic findings as seen in MEC are not present. Mucicarmine stain was negative in this biopsy material. Another consideration is high-grade mucoepidermoid carcinoma where typically the tumor shows a scarcity of mucus cells, with prominent cellular pleomorphism and malignant features of the squamous component, easily separating this diagnosis from JOOC.

The bland cellular microscopic characteristics of neuroepithelial structures similar to the JOOC coupled to awareness of a possible presentation in association with a mandibular exostosis, should prevent diagnostic confusion and unnecessary surgery.

To the best of our knowledge, this report represents the first documented case of neuroepithelial structures similar to the JOOC in a mandibular torus.

CONFLICT OF INTEREST STATEMENT

The authors have no conflict of interest to disclose.

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REFERENCES

1. Schoenwolf GC, Bleyl SB, Brauer PR, Francis-West PH. Larsen's Human Embryology, 4th ed.: Churchill Livingstone, Publishers; 2009.
2. Wysocki GP, Wright BA. Intraneural and perineural epithelial structures. Head Neck Surg. 1981;4(1):69-71.
3. Chievitz JH. Beiträge zur Entwicklungsgeschichte der Speicheldrüsen. Arch Anat Physiol. 1885;9:401-436.
4. Mérida-Velasco JR, Rodríguez-Vázquez JF, de la Cuadra-Blanco C, Salmerón JI, Sánchez-Montesinos I, Mérida-Velasco JA. Morphogenesis of the juxtaoral organ in humans. J Anat. 2005;206(2):155-163.
5. Zenker W. Organon bucco-temporale (Chievitzsches Organ), ein nervös epitheliales Organ beim Menschen. Anat Anz. 1953;100:257-265.
6. Kusafuka K, Kameya T. Juxtaoral organ of Chievitz, radiologically suspicious for invasion of lingual squamous cell carcinoma. Pathol Int. 2007;57(11):754-756.
7. Ide F, Mishima K, Saito I. Juxtaoral organ of Chievitz presenting clinically as a tumor. J Clin Pathol. 2003;56(10):789-790.
8. Leibl W, Pflüger H, Kerjaschki D. A case of nodular hyperplasia of the juxtaoral organ in man. Virchows Archiv A Pathol Anat Histol. 1976;371(4):389-391.
9. Soucy P, Cimone G, Carpenter B. An unusual intraoral mass in a child: the organ of Chievitz. J Pediatr Surg. 1990;25(11):1200.
10. Bénateau H, Rigau V, Comoz F, Benchemam Y, Galateau F, Compère JF. Tumor of the juxtaoral organ. Int J Oral Maxillofac Surg. 2003;32(1):101-103.
11. Eversole LR, Leider AS. Maxillary intraosseous neuroepithelial structures resembling those seen in the organ of Chievitz. Oral Surg Oral Med Oral Pathol. 1978;46(4):555-558.
12. Müller E, Zenker W. Enzyme-histochemistry of the juxtaoral organ in man(" organ of chievitz"). Hisochemistry. 1981;71(2):279-290.
13. Pantanowitz L, Balogh K. Significance of the Juxtaoral Organ (of Chievitz). Head Neck. 2002;25(5):400-405.
14. Tschen JA, Fechner RE. The juxtaoral organ of Chievitz. Am J Surg Pathol. 1979;3(2):147-150.
15. Ide F, Mishima K, et al. Melanin pigmentation in the juxtaoral organ of Chievitz. Pathol Int. 2003;53(4):262-263.
16. Ide F, Mishima K, Saito I. Pacinian corpuscle in the juxtaoral organ of Chievitz. J Oral Pathol Med. 2004;33(7):443-444.
17. Mikó T, Molnár P. The juxtaoral organ-a pitfall for pathologists. J Pathol. 1981;133(1):17-23.
18. Sciubba JJ, Sachs SA. Schwannoma of the inferior alveolar nerve in association with the organ of Chievitz. J Oral Pathol. 1980;9(1):16-28.