



DEPARTMENT OF PATHOLOGY

Case of the Week

ENT Pathology: Perioral Organ of Chievitz

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History

A 60-year-old female presented to the Otolaryngology service for further management of a tumor on the left cheek, of 2 months duration. Physical examination revealed a 9.0 x 9.0 cm exophytic, fungating mass with erythema and yellow purulent drainage. The mass appeared to extend to the left oral commissure, left buccal mucosa, left gingival mucosa, and left upper lip's labial mucosa. CT imaging showed a 4.8 x 4.4 x 4.4 cm heterogeneous mass in the left buccal space that extended to the skin surface, eroded through the buccinator muscle, and extended to the left maxillary sinus and hard palate. She underwent a biopsy which showed invasive squamous cell carcinoma, well-differentiated. A wide local excision of the left cheek mass was performed and sent to Pathology for intraoperative evaluation of the surgical margins. Frozen sections of the pterygoid margin shows small nests of epithelium in the submucosa, an unusual finding (Fig 1, 2, 3). Permanent sections of the same block shows similar small nests of epithelium in the submucosa (Fig 4, 5). An additional pterygoid margin was submitted for permanent section shows small nests of epithelium intermixed with nerve branches (Fig 6, 7, 8).

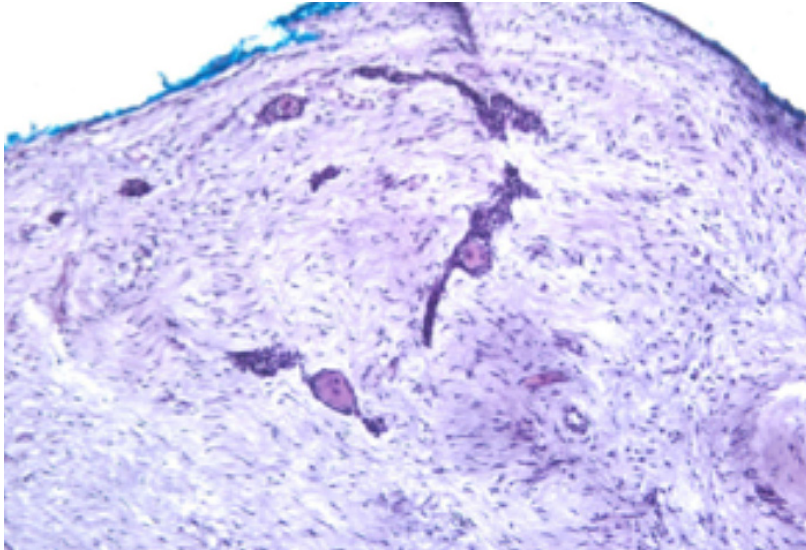


Figure 1: Small nests of squamous epithelium in the submucosa.
(Frozen section, H&E, 100x)

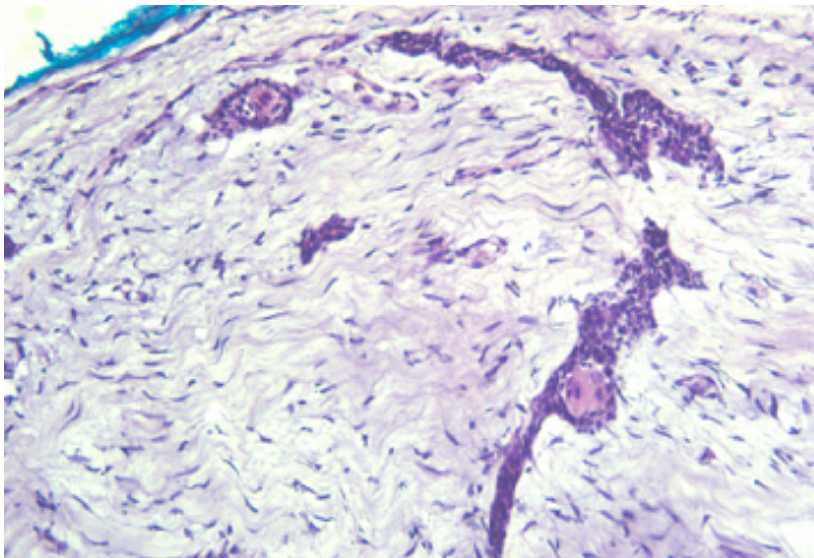
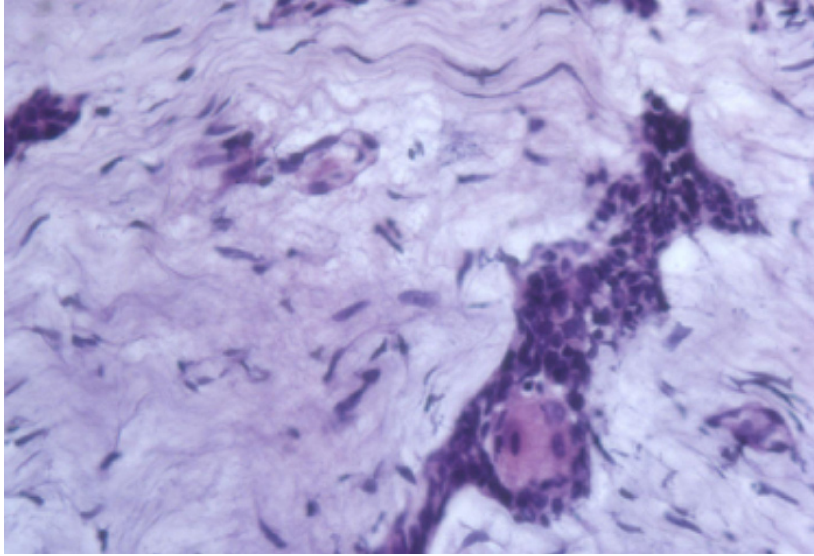
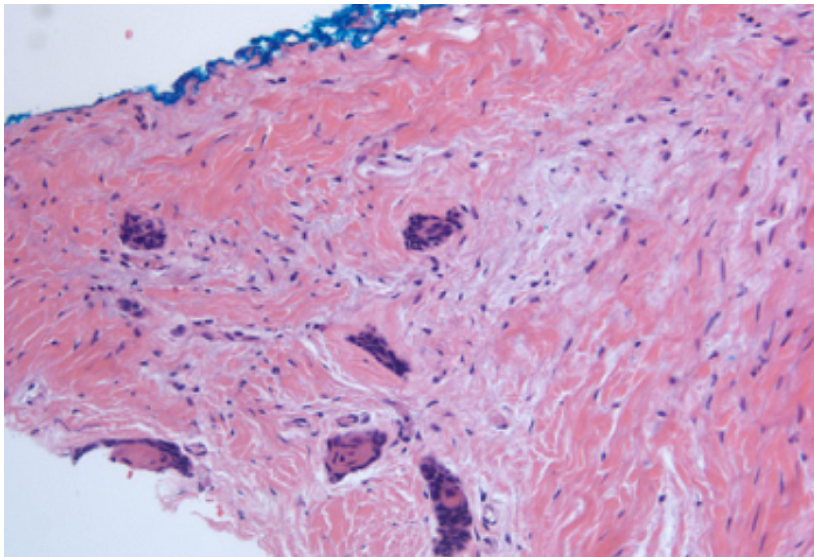


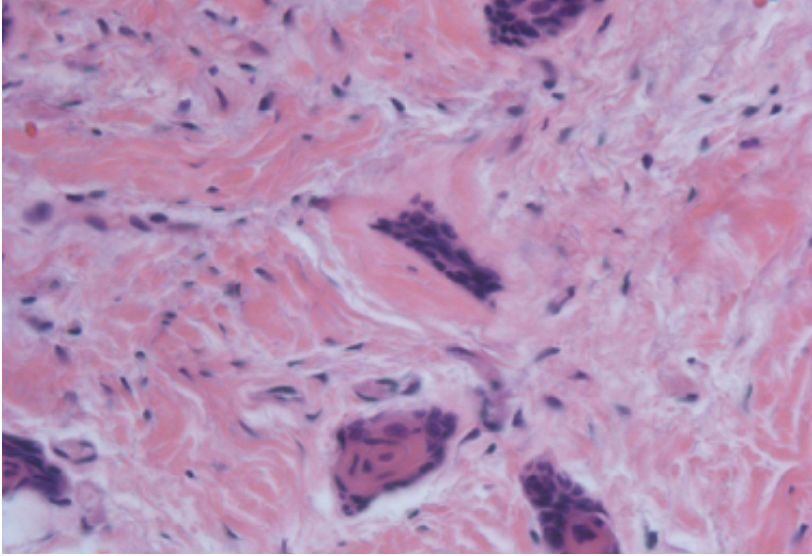
Figure 2: Small nests of squamous epithelium in the submucosa.
(Frozen section, H&E, 200x)



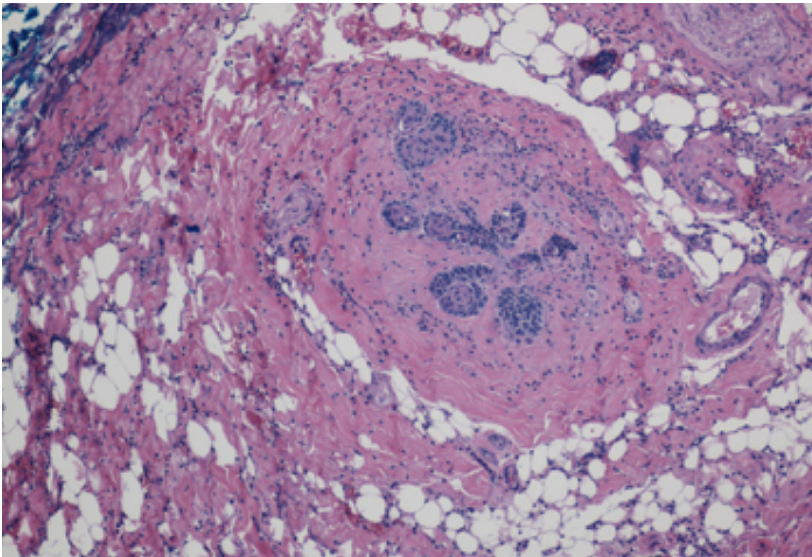
*Figure 3: Small nests of squamous epithelium in the submucosa.
(Frozen section, H&E, 400x)*



*Figure 4: Small nests of epithelium in the submucosa.
(Permanent section, H&E, 200x)*



*Figure 5: Small nests of epithelium in the submucosa.
(Permanent section, H&E, 400x)*



*Figure 6: Connective tissue plug containing small nests of non-keratinizing squamous epithelium intermixed with nerve branches.
(Permanent section, H&E, 100x)*

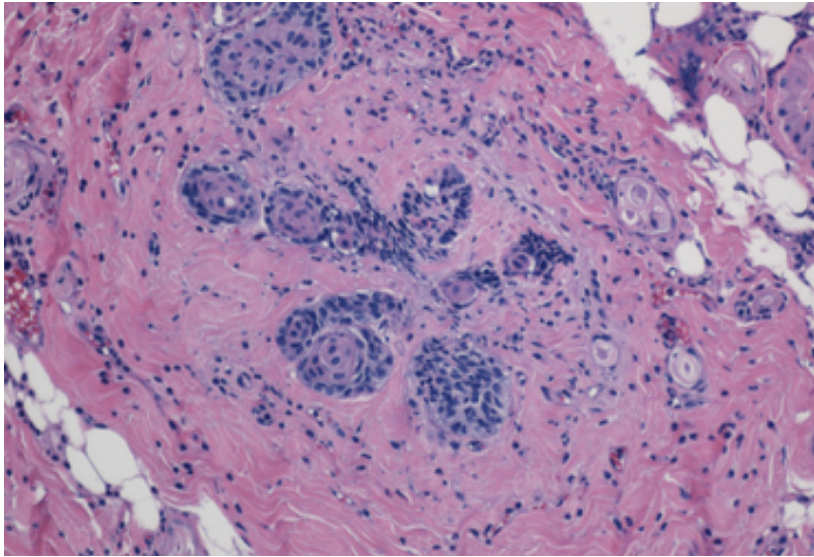


Figure 7: Connective tissue plug containing small nests of non-keratinizing squamous epithelium intermixed with nerve branches. (Permanent section, H&E, 200x)

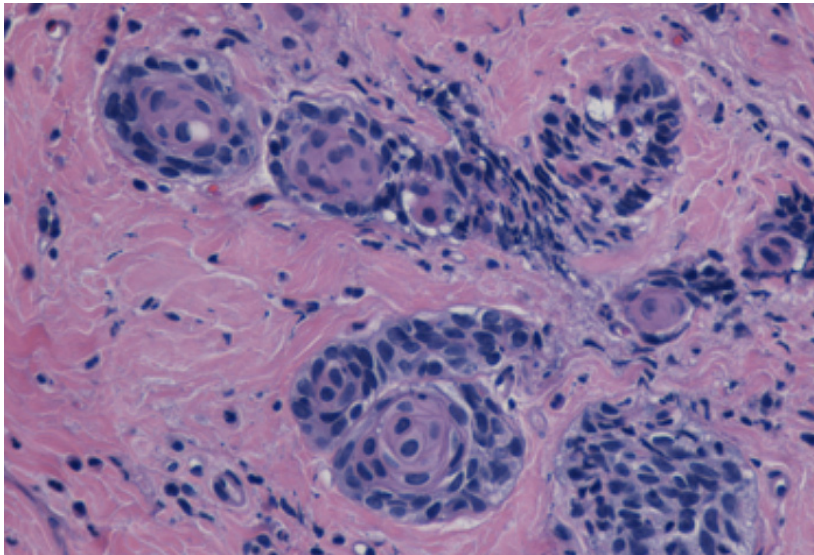


Figure 8: Connective tissue plug containing small nests of non-keratinizing squamous epithelium intermixed with nerve branches. (Permanent section, H&E, 400x)

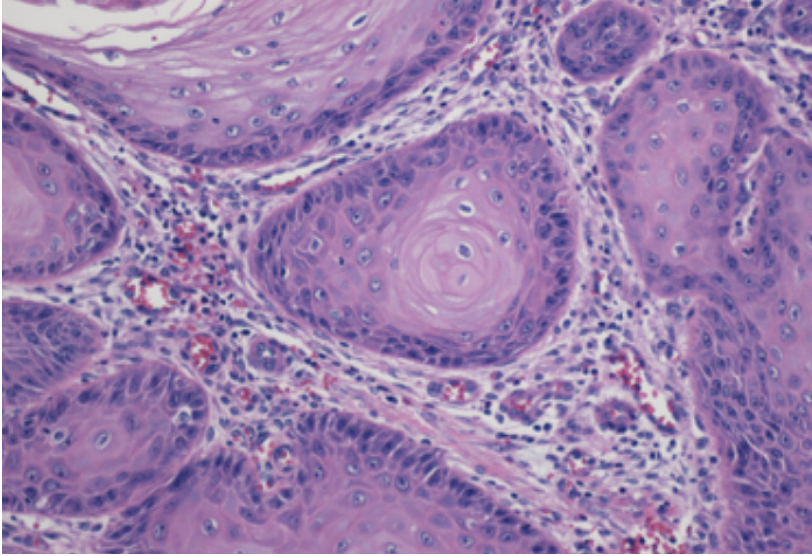


Figure 9: Section of tumor showing epithelial cell nests with keratin pearl formation, invasive into the lamina propria. (Permanent section, H&E, 100x)

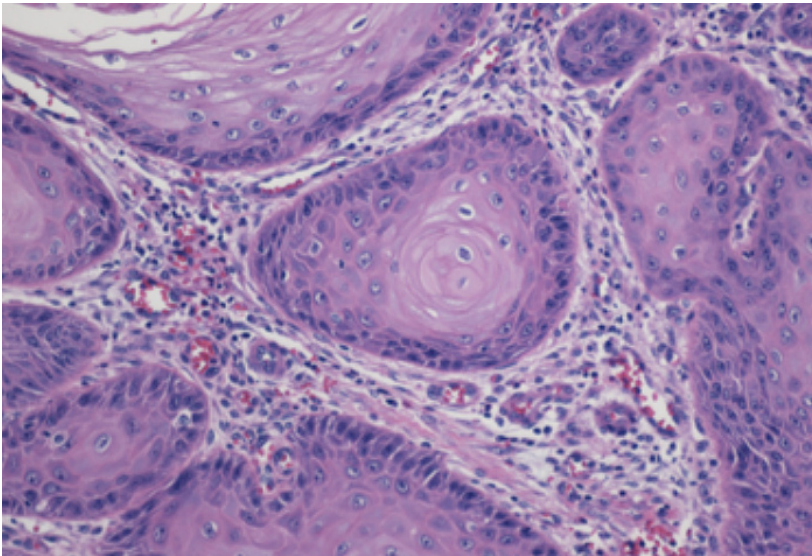


Figure 10: Section of tumor showing epithelial cell nests with keratin pearl formation, invasive into the lamina propria. (Permanent section, H&E, 400x)

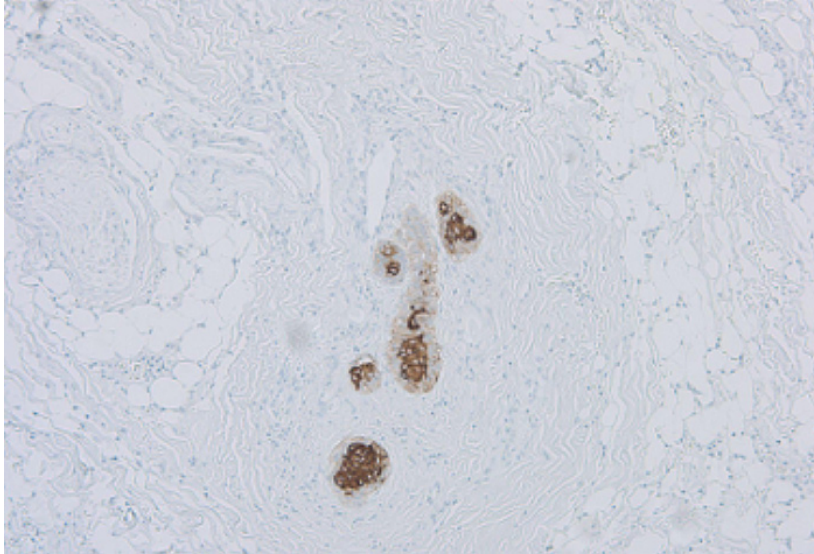


Figure 11: Immunostain for pancytokeratin highlights epithelial components. (Permanent section, AE1/AE3, 100x)

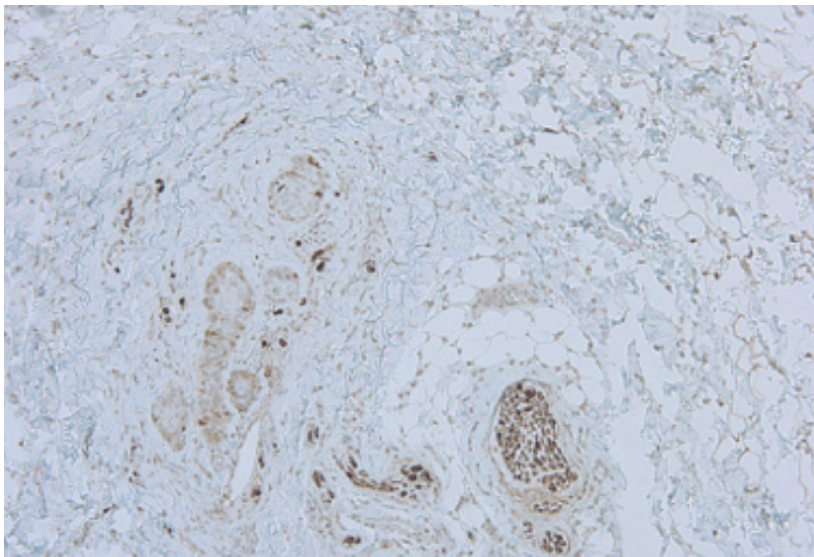


Figure 12: Immunostain for S100 highlights neural elements in vicinity of epithelial nests. (Permanent section, S100, 100x)

Diagnosis

Perioral Organ of Chievitz.

Discussion

The perioral organ, also known as Chievitz organ, is a diagnostic pitfall even for the experienced pathologist. The organ is a vestigial neuroepithelial structure located in the soft tissue overlying the angle of the mandible in the buccotemporal space. The perioral organ is found between the buccotemporal fascia and pterygoid muscle, and is innervated by branches of the buccal nerve. Its appearance as squamous nests can result in misdiagnosis as metastatic squamous cell carcinoma during head and neck frozen section evaluation. Seeing epithelium in an unusual location brings up the possibility of metastatic squamous cell carcinoma when there is a known clinical history of squamous cell carcinoma, and seeing the epithelial nests with nerve fibers brings up the possibility of neural invasion. However, it is important to note that the perioral organ has several features that may help the pathologist distinguish it from a malignant entity. The organ is composed of well-circumscribed bland squamous cell nests with no keratinization as opposed to metastatic squamous cell carcinoma composed of atypical squamous cells and keratin pearl formation (Fig 9, 10). The organ has three layers of connective tissue that surround the epithelial nests: stratum fibrosum internum, stratum nervosum, and stratum fibrosum externum. The stratum fibrosum internum is a thin capsule composed of dense collagen fibers, few elastic fibers, and is delineated from the epithelial nests by a basal lamina. The stratum nervosum consists of loose connective tissue composed of myelinated and unmyelinated nerve fibers. The stratum fibrosum externum is the outer capsule that encapsulates the entire organ, and is connected to the buccotemporalis. These different components can be highlighted using immunohistochemistry stains for epithelial and neural elements (Fig 11, 12).

References

1. Bommanavar SB, Hema KN, Baad R. Juxtaoral organ of Chievitz: An innocuous organ to be known. *J Oral Maxillofac Pathol.* 2017;21(1):162–164.
2. Pantanowitz L, Balogh K. Significance of the juxtaoral organ (of Chievitz). *Head Neck.* 2003 May;25(5):400-5; discussion 400. Review. PubMed PMID: 12692878.