LETTER TO JCP

Juxtaoral organ of Chievitz presenting clinically as a tumour

F Ide, K Mishima, I Saito

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An extremely rare hamartomatous lesion of the juxtaoral organ of Chievitz (JOOC) in a 63 year old man is reported. The tumour appeared as a large mass in the infratemporal fossa with associated mandibular bone resorption; histologically, it was well encapsulated and composed of numerous tangled masses of benign squamous epithelial nests and mature fibrofatty tissue. There were no histological features suggestive of neoplastic transformation. A literature survey confirmed that this is the first adult case of JOOC presenting clinically as an extraoral tumour.

A 63 year old man was referred for evaluation of a painless, slowly growing buccotemporal tumour that had been present for two years. Computed tomography scan and magnetic resonance imaging revealed a well demarcated, 6 cm mass lesion in the right infratemporal fossa near to the mandible (fig 1). There was an apparent bony erosion of the inner surface of the mandible. On opening of the mouth, a small mass was protruded intraorally at the level of the ascending ramus. The resected specimen was completely encapsulated and had a yellowish white colour and friable consistency. The surgeons' impression was that of a lipoma. He was free from recurrence four years after surgery.

HISTOLOGY

The tumour showed a conglomerate of numerous epithelial nests and mature fibrofatty tissue, in approximately equal amounts. Individual epithelial islands were composed of benign squamous cells with a peripheral rim of cuboidal basal

cells (fig 2A). Although many nests exhibited nodular hyperplasia (fig 2B), even in serial sections, both epithelial and mesenchymal components showed no evidence of neoplastic change. Among the epithelial islands, fine nerve fibres were conspicuous. Because of its specific location and characteristic histopathological features, a diagnosis of hamartoma of the juxtaoral organ of Chievitz (JOOC) was given.

Immunohistochemically, three wide spectrum cytokeratins (CKs; AE1/AE3, 34 β E12, and MNF116) were diffusely and strongly positive in all epithelial cells (fig 2C). CK14 was exclusively expressed in the central squamous cells. Both CK10 and CK19 were detected and central squamous cells often stained more intensely than the peripheral basal cells. Epithelial cells were uniformly negative for CK13, CK18, filaggrin, involucrin, and epithelial membrane antigen.

DISCUSSION

The JOOC is a normal anatomical structure located in the buccotemporal fascia on the medial surface of the ascending ramus.¹ Regardless of its function, the only practical importance lies in the potential for it to be misdiagnosed as perineural invasion in a patient with oral squamous cell carcinoma.¹ Pathological conditions associated with JOOC are rare. Leibl *et al* incidentally found, in a 68 year old woman, a single example of nodular hyperplasia in 100 postmortem specimens.² We are aware of only two reports describing an intraoral tumour-like mass in a child.^{3,4} Soucy *et al* reported a 5 year old girl with a small buccal mass containing JOOC, but provided neither clinical nor histological illustrations.³ Recently, a possible tumorous proliferation of JOOC was

Abbreviations: CK, cytokeratin; JOOC, juxtaoral organ of Chievitz

Figure 1 (A) Computed tomography scan and (B) magnetic resonance imaging of a large mass lesion in the infratemporal fossa.

Figure 2 (A) Benign squamous epithelial islands. Haematoxylin and eosin stained; original magnification, $\times 40$. (B) A nodular hyperplastic nest. Haematoxylin and eosin stained; original magnification, $\times 200$. (C) AE1/AE3 in squamous cells. Immunoperoxidase stained; original magnification, $\times 200$.

Take home messages

- This is the first adult case of juxtaoral organ of Chievitz (JOOC) presenting clinically as an extraoral tumour
- The two previously reported JOOC lesions had an intraoral location and onset was during childhood
- This is an extremely rare hamartomatous lesion of the JOOC, which appears to be benign

described in a 12 year old girl by Vadmal *et al.*⁴ The lesion that they described was composed of oval to spindle shaped cells, arranged in a whorled pattern and immunoreactive for vimentin and epithelial membrane antigen, suggesting a meningothelial nature. The clinical findings of our case differed considerably from these examples. The lesion that we describe was far larger and caused tumour related bone resorption.

“The description of further cases should shed more light on the pathogenesis of this peculiar lesion, which has an apparently benign clinical course”

Various wide spectrum CKs have been found to react with the epithelial nests of JOOC.^{4–6} In agreement with our findings, Mandl *et al* reported CK19 immunoreactivity in the central squamous cells.⁶ We also found positive staining for CK10 and CK14 in the epithelial cells. The available CK profiles to date suggest that the epithelial nests of JOOC share the immunohistochemical phenotype of non-keratinised stratified squamous cells.

Onset in childhood and an intraoral location are the common features of two previously reported lesions.^{3,4} Inflammation, trauma, and persistent growth have not been recorded; therefore, we favour the hamartomatous nature of this condition. The description of further cases should shed more light on the pathogenesis of this peculiar lesion, which has an apparently benign clinical course.

Authors' affiliations

F Ide, K Mishima, I Saito, Department of Pathology, Tsurumi University School of Dental Medicine, 2-1-3 Tsurumi, Tsurumi-ku, Yokohama 230-8501, Japan

Correspondence to: Dr F Ide, Department of Pathology, Tsurumi University School of Dental Medicine, 2-1-3 Tsurumi, Tsurumi-ku, Yokohama 230-8501, Japan; ide-f@tsurumi-u.ac.jp

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