EUROPEAN OPHTHALMIC PATHOLOGY SOCIETY

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Case number:	TVU21-12766
Material distributed:	1 histology slide

Title Of Case Presentation:
Unusual finding(s) in a dermoid tumour of the orbit; etiological considerations

Clinical History:
The patient was a 21 year old male, who had noticed a swelling in the superotemporal quadrant of the orbit and a mild ptosis. He had no other complaints, such as pain or diplopia. An MRI was made, showing a swelling in the anterior orbit, superotemporal, pressing against the globe and displacing the lacrimal gland. The process was strongly hyperintense, comparable to fatty tissue. Differentially, a dermoid cyst, a vascular anomaly or a (hemorrhagic) pleomorphic adenoma of the lacrimal gland were considered. The lesion was excised

Ocular Pathology:
Macroscopy:
The specimen measured 1,6 x 1,0 x 1,0 cm and gave the impression of an already opened cyst. Upon slicing the cyst contained a beige, semisolid material. It was completely enclosed in 3 lamellae.

Light microscopy:
Microscopy revealed a cystic structure that showed a somewhat variable aspect of the epithelium. In some areas there was a keratinizing squamous epithelium, with some adnexal structures attache4d, i.e. hairs and sebaceous glands. In other places the epithelium was non-keratinising and showed a few goblet cells within the epithelium, reminiscent of conjunctival epithelium. In yet another place the epithelium was multilayered with ciliated cells on the surface, and in yet another place there was a bilayered epithelium, reminiscent of adnexal or apocrine epithelium. The cyst content was somewhat granular, generally amorphic, with no hairs or squamous cells. In the surrounding tissue some fatty tissue, nerves, blood vessels and connective tissue was seen, but in proportional amounts to what was to be expected in tissue from this location.

Immunohistochemistry:
A variety of keratin stains was performed. In general the squamous epithelia were positive for keratins 5/6 and 13 and p63 and p40; in a few places keratin 7 and CAM 5.2 positive cells were seen on the surface. In areas were the epithelial layers were thinner the entire epithelium stained for keratin 5/6 and there was strong staining in all superficial cells and weaker staining of the basal cells for keratin 7 and strong staining of the basal cells for p63 , with the superficial cells being negative. The PAS stain revealed a couple of goblet cells

EM and Molecular pathology: Not performed

Discussion:
Dermoid cysts are among the commonest orbital lesions. In the majority of cases they are found in the superotemporal quadrant of the orbit (from 61 – 86 %) (Cavazza et al, 2011; Lieb,2010; Pushker et al, 2019; Reissis et al, 2014; Shields et al, 1997), with the superomedial location being the next most common. Rarely, other localisations are reported, such as intraconal (Samal et al, 2021)These lesions are considered to be congenital, caused by epithelial rests caught between boney structures along fetal lines of closure, for instance the frontozygomatic fissure (Cavazza, et al, 2011, Lieb, 2010).The term epidermoid is used to describe cases showing only surface (squamous) epithelium, while the term dermoid denotes the presence of adnexal structures such as hairs and/or sebaceous glands, with sweat glands being much less frequently present. There is a wide age range of patients, but most patients are younger, on average 17 years old (range: 5 months to 85 years) (Pushker et al, 2019, Shields et al, 1997). There is some debate about the occurrence of inflammation in these excision specimens, but there is a reasonable agreement on the frequency of significant inflammation, ranging from 25 – 40%; this is considered to be caused by rupture of the cysts and contact of the cysts content with the surrounding tissue (Abou-Rayyah et al, 2002, Reissis et al, 2014) The keratinous contents are highly irritative and dense inflammation after rupture is to be expected.

Though most reports just mention the cyst wall is lined by squamous epithelium, in our case the histology and immunohistochemistry were suggestive of more than just 1 type of epithelium in the cystic process. This was not always clear-cut, because reactive changes can occur in epithelia. But there was clearly conjunctival epithelium (squamous epithelium with strong p63 expression and keratin 5/6 in every layer except the most superficial cells, and there was squamous epithelium intimately associated to adnexal structures, i.e. sebaceous glands. In addition there was ciliated respiratory-type epithelium and clearly bilayered epithelium. In the literature in general dermoid cysts are said to be lined by squamous epithelium and little or no mention is made of other epithelial types. In a paper by the Shields', describing a fairly large number of desmoid cysts some 5% showed conjunctival epithelium (Shields et al, 1997). In isolated cases combinations of different epithelial types have been
describes. Hence, the term cystic epithelial choristoma was proposed to describe these lesions (Verdijk et al, 2015). The question remains what the explanation is for the occurrence of other types of epithelium in these lesions. Probably, theproximity of the conjunctiva explains a lining of conjunctival epithelium by retention of nests of that epithelium. But other types of epithelium can probably only be explained by metaplastic changes, possible by nearby Inflammation. It illustrates a pathologist's adagium: Everything is possible….

Literature:

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