Choristomatous tumors of the eye

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Clinical History: A 6 weeks old Caucasian male was presented with a slowly growing mass of the medial right lower eyelid that had been present since birth. The clinical differential diagnosis was made inclusion cyst, dermoid cyst or choristoma. No comorbid conditions were noted. The mass was excised and sent for pathologic evaluation. There was an uneventful convalescence.

Pathology: the specimen was pale, firm and rounded, measuring 1x0.6 cm.

Discussion: Phacomatous choristoma of the eyelid was first described by Zimmerman in 1971 and subsequently was reported to a total number of 24 cases. Phacomatous choristoma is consistently reported to be located in the medial lower eyelid, generally with involvement of the inferior anterior orbit. The tumor is often noticed immediately after birth and often slowly grows in size to a maximum diameter of 20 mm. The histological appearance is typical and exists of desmoplastic-like stoma with island of cuboid to polygonal cells surrounded by a thick layer of eosinophilic amorphic PAS-positive material. Other specifics are the presence of Wedl-bladder like cells and psammomatous calcifications. The epitheloid cells stain positive for vimentin and S100 and are negative for keratins.

The differential diagnosis of a tumor of the eyelid is large but can be restricted, because of the age of the patient, to infection, chalazion, nasolacrimal obstruction, dermoid cyst or complex choristoma. A short overview of choristomas of the eye is discussed, according to location.


 Conjunctiva, cornea and sclera: limbal dermoid, infratemporal epibulbar. Dermolipoma, superotemporal, epibulbar, no relation to limbus. Ectopic choristoma, often containing ectopic lacrimal gland, no predilection quadrant.


Multiple hypotheses have been proposed as to the origin of the choristomas of the eye. The three major lines of thought, atavism, sequestration and regulation are discussed.

14. Seregard S: Phakomatous choristoma may be located in the eyelid or orbit or both, Acta Ophthalmol Scand 1999, 77:343-346