Eastern Ophthalmic Pathology Society Meeting
Washington, DC
September 13-15, 2018

Nasreen A. Syed, MD
F.C. Blodi Eye Pathology Laboratory
Univ. of Iowa
233 Medical Research Center
Iowa City, IA 52242-1182, USA
319-335-7192 FAX: 319-335-7193
nasreen-syed@uiowa.edu

Case materials:
Glass slide (H&E) – 0
Images in powerpoint – 1
Written protocol – 1

Conjunctival Complex Choristoma

EYE18-19

HISTORY: The patient is a Caucasian female who presented initially at 3 months of age for a pedunculated mass at the left lateral canthus that had been present for 2 months. Clinically, the mass was thought to be a pyogenic granuloma and measured 2 mm x 1.5 mm. Excision was postponed until age 6 months due to anesthesia risk. She was seen at 6 months of age. The mass had increased to 3 mm x 2 mm. Observation was recommended. Her vision and extraocular motility were not affected. The eye was white and quiet. At the request of the parent, the mass was excised at 10 months of age.

POH: Unremarkable
PMH/PSH: Normal, full-term delivery, no trauma, meeting all milestones
FH: No history of eye disease, 4 other siblings all healthy

CLINICAL EXAM: See clinical photo.

MANAGEMENT: An excisional biopsy was performed.

PATHOLOGY: Grossly, the tissue consisted of a tan-white, rubbery, irregular piece of tissue measuring 4 mm x 3.5 mm x 3 mm. Microscopically, the mass was devoid of surface epithelium and scattered neutrophils and granulation tissue were present. The deeper tissue demonstrated fibrosis with an infiltrative proliferation of cells with bland, oval to spindle-shaped nuclei. Occasional nuclear pseudo-inclusions were present and there was a moderate amount of uniformly staining cytoplasm. Some cells were arranged in a whorl-like pattern. Some of the cells were separated by thick bundles of collagen. Immunohistochemistry demonstrated that most cells were positive with S100 and EMA. Occasional cells were positive with progesterone receptor. Somatostatin receptor, CD31, and CD68 were negative.

Deep to these cells is a fragment of bone with a fibrous pseudocapsule. Occasional osteoclasts surround the bone.

FURTHER EVALUATION AND MANAGEMENT: The patient underwent an MRI of the brain and orbits with and without contrast to screen for any other abnormalities of the head and neck and none were identified. She was seen in follow up at age 15 months. There was no evidence of recurrence of the lesion.

DISCUSSION:

Conjunctival choristomas are uncommon lesions that typically occur in children or young adults. Occasionally, they may be congenital. Lesions most commonly arise at the limbus or in the conjunctival fornix. They may be unilateral or, less often, bilateral and may be associated with other developmental anomalies in the ocular adnexa, eye or elsewhere. “Simple” choristomas, such as the dermoid or lipodermoid, are the most common of these developmental proliferations. They consist of epidermal/dermal components with a stroma consisting of dense, obliquely arranged bundles of collagen. Complex conjunctival choristomas are characterized by variable combinations of ectopic tissues such as cartilage[1,2], bone[3-8], lacrimal gland[9-13], adipose tissue, smooth muscle, and, more rarely, tooth[14] and brain[15]. The superficial component may resemble a dermoid in some cases with the other components present in the deeper tissue.

Meningothelial proliferations have only been rarely reported in ocular adnexal choristomatous lesions[16]. However, developmental “meningiomas” are well known to occur in the skin, albeit, uncommonly[17-23]. Cutaneous meningiomas are classified into 3 categories: Type I – primary cutaneous meningioma, Type II – ectopic meningioma of the deeper soft tissues extending into the skin, and Type III – CNS meningioma with extension into the skin[24]. Type I lesions tend to occur on the scalp or the paraspinal skin and are typically noted in childhood or early adulthood. These lesions tend to be of the meningothelial type. Almost all cutaneous meningiomas demonstrate the presence of psammoma bodies[25]. Type I cutaneous meningiomas typically express vimentin, S100 and EMA similar to intracranial lesions[21,26]. They may occasionally be

associated with genetic syndromes such as neurofibromatosis type 1[27]. These lesions may occasionally be mistaken for squamous neoplasms and careful evaluation of the microscopic features and clinical presentation are necessary to avoid a misdiagnosis as the immunophenotype can overlap with SCCA[28]. Cutaneous meningiomas and rudimentary meningoceles have been postulated to be two ends of a spectrum of the same developmental anomaly.[29,30] Another origin of cutaneous meningioma is previous trauma involving a skull or vertebral fracture[31,32].

Cases of type I cutaneous meningioma of the eyelid are rarely reported in humans. This lesion has been reported in dogs[33]. I could not find any cases of ocular surface meningioma reported in the literature. There are cases of type II cutaneous meningioma of the eyelid and periocular tissue reported in the literature[34-37]. These typically arise from the orbital soft tissue. Our case is from the conjunctiva but does have some parallels to the cutaneous form of meningioma including age of the patient and immunophenotype. However, our case lacked any psammoma bodies and included a deep rest of mature bone, consistent with a complex choristoma. Because there is no data regarding the presence of meningothelial cells in an ocular surface choristoma, we recommended imaging of the head to exclude a deeper lesion or other lesions that could suggest a related syndrome. The MRI was negative.

SUMMARY: Our case represents an example of a complex choristoma of the conjunctiva with the presence of meningothelial tissue. To our knowledge, this is the first report of this occurrence in the conjunctiva.

REFERENCES:

1. Zhang Z, Yang Z, Pan Q, Qin X, Deng Y, Cao Y. Epibulbar complex cartilaginous choristoma: A distinctive clinicopathological case series and literature review. *Medicine (Baltimore).* 2018;97(7):e9902. doi: 9910.1097/MD.0000000000009902.
2. Ojha PR, Deshpande AH, Gargade CB, Nigam JS. Epipalpebral conjunctival chondroid choristoma: Interesting developmental anomaly presenting in an adult. *Indian J Ophthalmol.* 2017;65(7):613-614. doi: 610.4103/ijo.IJO_4139_4117.
3. Harkins KA, Perry D, Suh DW. Epibulbar osseous choristoma: A case report. *Am J Ophthalmol Case Rep.* 2017;5:4-6.(doi):10.1016/j.ajoc.2016.1010.1002. eCollection 2017 Apr.
4. Vachette M, Moulin A, Zografos L, Schalenbourg A. Epibulbar osseous choristoma: a clinicopathological case series and review of the literature. *Klin Monbl Augenheilkd.* 2012;229(4):420-423. doi: 410.1055/s-0031-1299256. Epub 1292012 Apr 1299211.
5. Gayre GS, Proia AD, Dutton JJ. Epibulbar osseous choristoma: case report and review of the literature. *Ophthalmic Surg Lasers.* 2002;33(5):410-415.
6. Ortiz JM, Yanoff M. Epipalpebral conjunctival osseous choristoma. *Br J Ophthalmol.* 1979;63(3):173-176.
7. Ferry AP, Hein HF. Epibulbar osseous choristoma within an epibulbar dermoid. *Am J Ophthalmol.* 1970;70(5):764-766.
8. Boniuk M, Zimmerman LE. Epibulbar osteoma (episcleral osseous choristoma). *Am J Ophthalmol.* 1962;53:290-296.
9. Tuncer S, Araz B, Peksayar G, Buyukbabani N. Solitary lacrimal gland choristoma of the limbal conjunctiva. *Ophthalmic Surg Lasers Imaging.* 2010;41(Online):e1-2. doi: 10.3928/15428877-20101025-15428809.
10. Alyahya GA, Bangsgaard R, Prause JU, Heegaard S. Occurrence of lacrimal gland tissue outside the lacrimal fossa: comparison of clinical and histopathological findings. *Acta Ophthalmol Scand.* 2005;83(1):100-103. doi: 110.1111/j.1600-0420.2005.00365.x.
11. Pokorny KS, Hyman BM, Jakobiec FA, Perry HD, Caputo AR, Iwamoto T. Epibulbar choristomas containing lacrimal tissue. Clinical distinction from dermoids and histologic evidence of an origin from the palpebral lobe. *Ophthalmology.* 1987;94(10):1249-1257.
12. Firat T, Emuler U. Two cases of aberrant lacrimal gland. *Ann Ophthalmol.* 1971;3(1):50-51.
13. Pfaffenbach DD, Green WR. Ectopic lacrimal gland. *Int Ophthalmol Clin.* 1971;11(3):149-159.
14. Jakobiec FA, Nguyen J, Mandell K, Fay A. Complex palpebral odontogenic choristoma: A reappraisal of the origin of teeth-bearing periocular lesions. *Am J Ophthalmol.* 2009;147(3):531-543.e531. doi: 510.1016/j.ajo.2008.1009.1033.
15. Emamy H, Ahmadian H. Limbal dermoid with ectopic brain tissue. Report of a case and review of the literature. *Arch Ophthalmol.* 1977;95(12):2201-2202.
16. Wolter JR, Benz SC. Ectopic meningioma of the superior orbital rim. *Arch Ophthalmol.* 1976;94(11):1920-1922.
17. Konopinski J, Prieto VG, Ivan D, et al. Isolated Ectopic Cutaneous Atypical Meningioma of the Scalp: Another Mimicker of Primary Adnexal Tumor. *Am J Dermatopathol.* 2017;39(7):545-547. doi: 510.1097/DAD.0000000000000821.
18. Miedema JR, Zedek D. Cutaneous meningioma. *Arch Pathol Lab Med.* 2012;136(2):208-211. doi: 210.5858/arpa.2010-0505-RS.
19. Hussein MR, Abdelwahed AR. Primary cutaneous meningioma of the scalp: a case report and review of literature. *J Cutan Pathol.* 2007;34(Suppl 1):26-28. doi: 10.1111/j.1600-0560.2007.00720.x.
20. Penas PF, Jones-Caballero M, Garcia-Diez A. Cutaneous heterotopic meningeal nodules. *Arch Dermatol.* 1995;131(6):731.
21. Gelli MC, Pasquinelli G, Martinelli G, Gardini G. Cutaneous meningioma: histochemical, immunohistochemical and ultrastructural investigation. *Histopathology.* 1993;23(6):576-578.

22. Barr RJ, Yi ES, Jensen JL, Wuerker RB, Liao SY. Meningioma-like tumor of the skin. An ultrastructural and immunohistochemical study. *Am J Surg Pathol.* 1993;17(8):779-787.
23. Theaker JM, Fletcher CD, Tudway AJ. Cutaneous heterotopic meningeal nodules. *Histopathology.* 1990;16(5):475-479.
24. Ragoowansi R, Thomas V, Powell BW. Cutaneous meningioma of the scalp: a case report and review of literature. *Br J Plast Surg.* 1998;51(5):402-404.
25. Lopez DA, Silvers DN, Helwig EB. Cutaneous meningiomas--a clinicopathologic study. *Cancer.* 1974;34(3):728-744.
26. Possanzini P, Pipolo C, Romagnoli S, et al. Primary extra-cranial meningioma of head and neck: clinical, histopathological and immunohistochemical study of three cases. *Acta Otorhinolaryngol Ital.* 2012;32(5):336-338.
27. Argenyi ZB, Thieberg MD, Hayes CM, Whitaker DC. Primary cutaneous meningioma associated with von Recklinghausen's disease. *J Cutan Pathol.* 1994;21(6):549-556.
28. Fox MD, Billings SD, Gleason BC, Thomas AB, Cibull TL. Cutaneous meningioma: a potential diagnostic pitfall in p63 positive cutaneous neoplasms. *J Cutan Pathol.* 2013;40(10):891-895. doi: 810.1111/cup.12198. Epub 12013 Aug 12197.
29. Brantsch KD, Metzler G, Maennlin S, Breuninger H. A meningioma of the scalp that might have developed from a rudimentary meningocele. *Clin Exp Dermatol.* 2009;34(8):e792-794. doi: 710.1111/j.1365-2230.2009.03519.x. Epub 02009 Oct 03510.
30. Sibley DA, Cooper PH. Rudimentary meningocele: a variant of "primary cutaneous meningioma". *J Cutan Pathol.* 1989;16(2):72-80.
31. Pacheco Compana FJ, Midon Miguez J, Avellaneda Oviedo EM, Busto Lodeiro E. Post-Traumatic Cutaneous Meningioma. *Arch Plast Surg.* 2016;43(4):381-384. doi: 310.5999/aps.2016.5943.5994.5381. Epub 2016 Jul 5920.
32. Borggreven PA, de Graaf FH, van der Valk P, Leemans CR. Post-traumatic cutaneous meningioma. *J Laryngol Otol.* 2004;118(3):228-230. doi: 210.1258/002221504322928035.
33. Teixeira LB, Pinkerton ME, Dubielzig RR. Periocular extracranial cutaneous meningiomas in two dogs. *J Vet Diagn Invest.* 2014;26(4):575-579. doi: 510.1177/1040638714533116.
34. Decock CE, Kataria S, Breusegem CM, Van Den Broecke CM, Claerhout IJ. Ectopic meningioma anterior to the lacrimal gland fossa. *Ophthalmic Plast Reconstr Surg.* 2009;25(1):57-59. doi: 10.1097/IOP.1090b1013e3181936811.
35. Kakizoe S, Kojiro M, Hikita N. Primary cutaneous meningioma. Report of a case. *Acta Pathol Jpn.* 1987;37(3):511-514.
36. Lee DH, Sim HS, Hwang JH, Kim KS, Lee SY. Extracranial Meningioma Presenting as an Eyebrow Mass. *J Craniofac Surg.* 2017;28(4):e305-e307. doi: 310.1097/SCS.0000000000003555.
37. Tan LT, Stewart CM, Sheerin F, MacDonald B, Silva P, Norris JH. Ectopic orbital meningioma: Fact or fiction? *Orbit.* 2017;36(3):144-146. doi: 110.1080/01676830.01672017.01279663. Epub 01672017 Mar 01676833.