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Epibulbar complex/osseous choristoma: clinicopathological study in a tertiary eye hospital

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Epibulbar choristoma is a congenital lesion that arises from ectopic pluripotent cells capable of differentiating into several elements: skin, adipose tissue, bone, lacrimal gland, cartilage, and rarely myxomatous tissue. The incidence is 1 in 10,000 and it affects the cornea, limbal conjunctiva and subconjunctival space. In our study, we aimed at presenting our experience with these lesions in a Tertiary Eye Hospital focusing on the complex and/or osseous type of choristomas and their ophthalmic and/or systemic associations. We collected all cases with the tissue diagnosis of epibulbar choristoma during the period: January 2000 to end of December 2016 for review. Out of a total 120 patients with epibulbar choristoma, complex choristoma constituted 13/15 patients (10.8%) while 2 patients only had osseous choristoma (1.7%). All cases were from Saudi Arabia. 11/15 (73.3%) had other ophthalmic manifestations with the commonest being upper lid coloboma in 1/3 followed by optic nerve anomaly, while half had associated syndromes. Goldenhar's syndrome was the most common in 5/13. Other associations included linear nevus sebaceous syndrome (LNSS) and encephalo-cranio-cutaneous lipomatosis (ECCL). One patient with osseous choristoma had an associated Coat's disease in the same eye. All cases were managed surgically with a mean duration of 44.6 months between the presentations to surgical intervention. The most common indication for surgery was cosmetic. Histopathologically, the choristoma in our series were not different than what has been reported. Interestingly the presence of smooth muscle was significantly associated with a larger size choristoma. In conclusion, in our series 73.3% of complex choristoma had associated ophthalmic abnormality (mostly lid coloboma). We had the first reported case of combined Goldenhar's and ECCL. Therefore, we recommend further studies on the pathogenesis of these lesions with consideration of molecular genetic etiology.

Recent Publications:

1. Tasse C, Böhringer S, Fischer S, et al. (2005) Oculo-auriculo-vertebral spectrum (OAVS): clinical evaluation and severity scoring of 53 patients and proposal for a new classification. *European Journal of Medical Genetics* 48:397–411.
2. Vachette M, Moulin A, Zografos L and Schalenbourg A (2012) Epibulbar osseous choristoma: a clinicopathological case series and review of the literature. *Klin Monbl Augenheilkd* 229(4):420–423.
3. Kim B J and Kazim M (2006) Bilateral symmetrical epibulbar osseous choristoma. *Ophthalmology* 113(3):456–458
4. Shields C L, Demirci H, Karatza E and Shields J A (2004) Clinical survey of 1643 melanocytic and nonmelanocytic conjunctival tumors. *Ophthalmology* 111(9):1747–1754.
5. Sangwan V S, Sridhar M S and Vemuganti G K (2003) Treatment of complex choristomas by excision and amniotic membrane transplantation. *Arch Ophthalmology* 121:278–280.

Biography

Hind Alkatan has completed her Ophthalmology from King Saud University, Riyadh, Saudi Arabia and her Postdoctoral studies from Departments of Ophthalmology/Pathology, University of Manitoba and University of British Columbia, Canada. She is an Assistant Professor (College of Medicine), Consultant (Departments of Ophthalmology and Pathology), Chief of Ophthalmic Pathology Division, and Director of the KSU Post-Graduate Residency & Fellowship Training Programs in Ophthalmology, King Saud University Medical City (KSUMC), Riyadh, SA. She is a member in many international organizations in her field: Eastern Ophthalmic Pathology, Canadian Ophthalmology Society, International Society of Ocular Oncology, and Saudi Ophthalmology Society. She has been contributing as an invited speaker in many international symposia such as the World Congress of Ophthalmology and the European Society of Pathology Annual meetings. She has published more than 120 papers in reputed journals and has been serving as an Editorial Board Member for several journals.

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