Ocular features, clinical profile and results of surgical intervention in a series of patients with port wine stain and neurofibromatosis

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Abstract
Purpose: To describe the ocular features, clinical profile and results of surgical intervention in a series of patients with facial port-wine stain (PWS) and neurofibromatosis (NF) in the pediatric age group.

Methods: All consecutive patients having PWS and NF presenting between January 2011 and April 2012 were included. Their systemic and ocular features were recorded. Imaging study of brain and orbits was carried out where required. Appropriate surgical interventions were carried out as indicated. These included trabeculectomy, cycloablation, excision of episcleral haemangioma and vitreo-retinal surgery. Patients were followed up for complications and control of intra ocular pressure (IOP).

Results: A total of 9 eyes of 8 patients were included. At presentation IOP in the involved eye ranged from 6 to 42 mmHg (mean 25.00 ±12.23). Glaucoma was present in 6 eyes. Episcleral haemangioma was noticed in one patient, retinal detachment (RD) was seen in 2 patients. Diffuse choroidal thickening on B scan was noted in 5 eyes of patients with PWS. After applying the appropriate management option, IOP on last follow up ranged from 6 to 24mmHg (mean 13.44±5.43) in the involved eye the last follow up ranged from 6 to 24mmHg (mean 13.44±5.43) in the involved eye. The difference between IOP of the involved eyes at initial presentation and that recorded at the last follow-up was significant (p=0.005).

Conclusion: Patients with SWS and NF present with varied clinico-ocular profiles and therapeutic interventions are individualized according to the ocular involvement. Post operative complications after treatment of glaucoma in such patients can largely be prevented with special measures taken per-operatively. Al-Shifa Journal of Ophthalmology 2013; 9(1): 16-21 © Al-Shifa Trust Eye Hospital, Rawalpindi, Pakistan.