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Case report: Rare case of phakomatosis pigmentovascularis with bilateral advanced glaucoma

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Introduction: Phakomatosis Pigmento-Vascularis (PPV) is a congenital syndrome characterized mainly by the concurrent presence of capillary malformation and pigmentary nevi. This rare condition was first described by Ota et al. in 1947.

Methods: A 7 yr old male who underwent trabeculotomy with trabeculectomy at the 3 months of age presented to us with intra-ocular pressure of 40 mmHg in both eyes on maximal medical therapy.

His BCVA was 6/60P in both eyes. He had diffuse flat bluish grey pigmented patches on the sclera with flat bleb superiorly in both eyes. He was evaluated by the ophthalmology, neurology, cardiology, dermatology department and was diagnosed to have Phakomatosis pigmentovascularis type II B with advanced bilateral glaucoma with ADHD.

Conclusion: Phakomatosis pigmentovascularis is a neural crest disorder. When oculodermal melanocytosis and nevus flammeus (phakomatosis pigmentovascularis) occur together, with each extensively involving the globe, there is a strong predisposition for congenital glaucoma, when one or both are present with only partial regular intervals for the development of glaucoma.

Biography

Sneha Kankaria is a Dynamic Young Glaucoma Specialist practicing at Agarwal's Eye Hospital India. She has a vast exposure in Medical management/Lasers in Glaucoma. She has a keen interest in Glaucoma implant and Congenital glaucoma. She has to her credit various publications and presentations at National and International Conferences where she participates to keep abreast of latest development. Her latest challenge is now Incorporating MIGS in her practice being in a developing country.

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