Abstract

Congenital cyst of the optic nerve is a rare congenital anomaly, scarcely reported. We report a large case series from a single institute over a period of 21 years, consisting of 3 cases with rare presentations. Retrospective study of 1100 orbital cases revealed three cases with congenital cyst of optic nerve reported here. In these patients, a cyst was replacing almost all of the intraorbital portion of the optic nerve, with their unique features. Management of the cases is also discussed.

Key Words: Congenital cyst, optic nerve, orbit.

enlargement of the bony orbit with thinning and curving of the lateral wall, but no erosion (figure: 1b). Both eye balls were of the same size. No abnormality was found in her right eye. Anterolateral trans-conjunctival orbitotomy revealed large cystic mass that replaced almost all of the intra-orbital part of the optic nerve (figure: 1c). It was first drained, which did not cause softening of the globe, and then excised in toto. Histopathology revealed glial tissue. No malignant cells were found on cytology.

Case 3
Our third patient was a 5-month-old female with right inferonasal dystopia since birth. On visual assessment of her right eye, the child was not following light. Ipsilateral cornea was smaller by 2mm (figure: 2a). There was limitation of extraocular movements in all gazes. Pupil was non-reactive and fundus examination revealed a pale optic disc. Computed tomography revealed a large intra-conal cyst replacing the optic nerve (figure: 2b, c, d). Both the eye ball measurements were equal. It was excised through an anterior, trans-conjunctival lateral approach after initial drainage. It contained clear fluid. Acceptable cosmetic results were found with improved dystopia. Histopathology revealed glial tissue. No malignant cells were found on cytology.

DISCUSSION
Congenital abnormalities of the optic disc may include, size abnormalities, like optic disc aplasia, hypoplasia, megalopapilla and optic disc cupping in prematurity. MEDLINE search revealed only few isolated cases of optic nerve cysts, reported so far. Kim et al, reported a case of an optic nerve cyst involving distal end of the intra-orbital part of the optic nerve while in the current series, the cyst involved almost all of the intra-orbital part of the optic nerve. There was no associated corneal pathology in their case and the patient was male. Whereas all our cases were females and ipsilateral corneas were smaller by 2mm. Holland et al, also reported an optic nerve cyst but unlike the current series, it was associated with a cystic eye ball. Shankar et al, reported bilateral retrobulbar cyst with associated optic disc colobomas. Optic disc coloboma with an associated cyst in the optic nerve sheath was also reported by Wiggins et al.

Optic nerve in previously reported cases was intact while the intraorbital part of the optic nerve is entirely...
replaced by a cyst in our series. Optic canal part of the optic nerve was intact. The only explanation for cyst involving only the intraorbital part seems to be the loose space available for the cyst to expand within the orbit. The intracanalicular portion of the optic nerve was not biopsied by us or by other observers in the past. We do not know whether the axon migration is normal or there is a developmental abnormality beyond orbit.

At 6 weeks of embryological development, closure of fissure starts from the center extending anteriorly towards the optic cup and posteriorly towards the optic stalk. During week 7, axons begin to grow in to the optics talk and development of the optic nerve is completed by the eighth week. It is believed that during 6th to 8th week of development, instead of axon growth and cell migration in to the optics talk, it is replaced by fluid giving rise to an optic nerve cyst. The cause may be any developmental anomaly, genetic event or environmental factor, which is still a matter of debate.

CONCLUSION
Complete replacement of the intraorbital part of the optic nerve by congenital cyst has not been reported before to the best of our knowledge. It is important to rule out secondary causes of optic nerve cysts before labeling a cyst as benign. Our study shows female preponderance and association of ipsilateral smaller corneal in such cases.

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REFERENCES


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