Aniridia associated with congenital Aphakia and Secondary glaucoma.

A 35-year-old gentleman presented with a decrease of vision in both eyes (OU) since 15 years. His past ocular and medical history was not significant. He and his parents did not give history of trauma or ocular surgery. On ocular examination, he had jerky nystgamus, best-corrected visual acuity (BCVA) of 20/400 in left eye (OS) with aphakic correction and no PL in right eye (OD), corneal stromal oedema with an intercalary staphyloma in OD and peripheral corneal scar in OS, aniridia and aphakia in OU (figure 1, left image). Intact and formed anterior hyaloid face was seen in OU. Intraocular pressures (IOP) were 28 mm Hg in OD and 36 mm Hg in OS. Gonioscopy showed rudimentary angle structures with cilary body band inferiorly in OS. Fundus examination showed total glaucomatous optic atrophy in OD and 0.9:1 cup disc ratio with bipolar notch and foveal hypoplasia in OS. After a month of medical treatment with topical 0.5% Timolol maleate and 0.15% Brimonidine tartarate, the IOP remained uncontrolled; we performed a planned trabeculectomy with Mitomycin C (0.04% of MMC for 3 minutes) and anterior vitrectomy under local anesthesia in his OS. His intra-operative and post-operative course was uneventful.

Post-operatively, at the end of five weeks the patient maintained BCVA of 20/400 in the left eye. A diffuse bleb was present (figure 1, right image). IOP in the left eye was 7 mm Hg and he did not require any topical anti-glaucoma medications. At last visit (6 months post-operatively), he maintained BCVA of 20/400 in the left eye and IOP of 6 mm Hg.

Visual function may be affected in Aniridia due to other associated anomalies like secondary glaucoma, cataract, foveal hypoplasia. To our knowledge there is no reported case in literature where aniridia has been associated with congenital aphakia. In aniridia, secondary glaucoma are attributed to number of mechanisms like increased confluence of irregular attachments from iris stroma to angle wall, absence of schlemm's canal, or secondary angle closure following miotic therapy. In the present case, vision in right eye was lost due to glaucomatous optic atrophy and in left eye there was advanced cupping. Timely, planned surgery with Trabeculectomy with MMC and anterior vitrectomy could control his IOP without medication and gave stable vision. Number of surgical procedures have been described with unsatisfactory and varying results for the management of secondary glaucoma in aniridia like ALT, goniotomy, Trabeculectomy , cyclocryotherapy, Molteno implantation.[3]

This case highlights that aniridia can also be associated with primary aphakia and secondary glaucoma and timely surgical intervention in such cases achieves adequate control of IOP with preservation of good visual function.

FIGURE LEGENDS:

FIGURE 1: Left eye of the patient with aphakia and aniridia.