A 65 years old female, visited our institute in January 2007 with the chief complaint of redness of both eyes. She had noticed redness of eye for last 9-10 months. She was known case of hypertension for last 12 years.

On ocular examination, Her best-corrected visual acuity was 6/6, N6 in both eyes. Intraocular pressure (IOP) by applanation tonometry was 38 mmHg in right eye (RE) and 24 mmHg in left eye (LE). Anterior segment examination showed dilated episcleral vessels in both eyes (figure 1a & 1b). The RE episcleral veins were more dilated compared to LE. Gonioscopy showed open angles with blood in Schlemm's canal in both eyes. Dilated fundus examination showed CDR (cup to disc ratio) to be 0.6 with inferior rim thinning in RE. In LE, CDR was 0.5 with healthy neuroretinal rim. Peripheral retina showed normal caliber retinal vessels with no sign of choroidal hemangioma. Central corneal thickness was 588 microns in RE and 586 microns in LE. Automated perimerty (HFA (24-2) showed normal field. As her IOP was high, she was diagnosed as ocular hypertension and was started on Timolol 0.5% BE and Latanoprost eye drop at night in RE.

She was examined again after 3 days by orbit specialist. Ocular examination was negative for any signs suggestive of thyroid eye disease. The ultrasound (USG) of orbit showed normal caliber of superior ophthalmic vein in both eyes. Her thyroid profile (T3, T4, TSH, TSH receptor antibody) was repeated twice and were normal. Her magnetic resonance imaging (MRI) & magnetic resonance angiography (MRA) orbit was not suggestive of carotid cavernous fistula and low flow Dural AV fistula. Provisional diagnosis of idiopathic elevated episcleral venous pressure was made. She was also given course of systemic steroid in tapering dose, which did not reduce dilated episcleral vein.

During follow up visit, her IOP always remain in high thirties in RE and mid twenties in LE. She was added topical Alpa-2 agonist (Brimonidine Tartarate three times a day) and topical carbonic anhydrase inhibitor (CAI) (dorzolamide, twice a day) in RE. On maximum medical therapy, her IOP fluctuated between 32 – 42 mm Hg in RE and 24-32 mm Hg in LE between Jan 07 to May 07. In May 07, her repeat visual field, optic disc and OCT findings were similar to her first visit findings. In view of her persistent elevated IOP and dilated episcleral veins, She underwent digital subtraction angiography (DSA), to rule out low-grade dural arterio venous fistula. The DSA showed normal brain vasculature. She was asked to have chest physician opinion to rule out

primary pulmonary hypertension & / or tumor at apex of lung. The Chest X-ray and computerized tomography (CT scan) of chest were normal.

In December 2007, as her IOP was always in high thirties in RE despite maximum medications and was advised trabeculectomy. It was thought that due to high episcleral venous pressure, she would have high chances of uveal effusion after trabeculectomy, so partial thickness sclerectomy with sclerotomy in infero-temporal region was planned. Right eye trabeculectomy with partial thickness sclerectomy with sclerotomy in infero-temporal region was performed on 7th January 2008. Initially, partial thickness sclrectomy with sclerotomy was performed, however the incision was not deepened into the suprachorioidal space. Subsequently routine fornix based trabeculetomy with mitomycin (large surface area technique) was performed superiorly. After completion of trabeculectomy, there was difficulty in forming anterior chamber, it was though that patient has developed chroidal effusion and the sclerotomy was deepened into suprachoroidal space and straw color fluid was drained. After the choroidal fluid drainage the anterior chamber was formed.

Postoperatively on day 1, her IOP was 12 mm Hg, good bleb and shallow anterior chamber with collerate iridocorneal touch. On fundus examination, she had bullous choridal detachment, which was documented on ultrasonography. As the choroidals were not kissing and central anterior chamber was formed, she was treated conservatively. She was given systemic steroids for 15 days. After 3 weeks of surgery, her choroidals subsided with conservative management. Her IOP was on 23rd day of surgery was 20 and 25 mm of Hg in RE and LE respectively. On anterior segment examination, she had good bleb, well formed anterior chamber. Last she was seen 7 months after the surgery, her IOP was 22 mm Hg in RE without any medicines and 25 mm Hg in LE on anti-glaucoma medications.

Discussion:

Dilated episcleral vein with secondary glaucoma is always difficult to diagnose and mange. IDEV is considered congenital in origin and other all pathology should able to diagnose by way of non-invasive or invasive investigations. In patients with IDEV a congenital abnormality in vasculature and familial predisposition have been suggested as the cause; however some reports suggest that the ocular injection is acquired. Most of reports in literature report this entity in 2nd or 3rd decade. It is actually diagnosis of exclusion; however in our case the presentation was late and despite normal

investigation we don't think it could be IDEV. Radius-Maumenee syndrome is another possibility but it usually unilateral and the episcleral veins usually regresses in caliber and congestion after trabeculectomy, which did not happen in our case. Low areterio venous grade fistula would come very close to as first differential. The gold standard to rule out the presence of a low-grade dural AV fistula is an angiography as in a small percentage of cases, it can be missed on MRA. The MRA and DSA both were normal and we could not confirm the diagnosis. However it is possible that low-grade fistula can spontaneously get closed and does not show any abnormality on angiography. We did not measure the episcleral blood flow or episcleral venous pressure in our patient but feel that would not have lead to any specific diagnosis.

The management is always difficult in these cases. Some argue for early filtration surgery. It is argued that drug that works on uveoscleral pathway may able to reduce better than aqueous suppressant. However in our patient, prostaglandin analogue did not reduce IOP. It is thought that due to high episcleral venous pressure, the surgery would have higher complication rate. We assumed that higher episcleral venous pressure and sudden IOP lowering after trabeculectomy would lead to uveal effusion like syndrome. From this perspective we decided partial thickness sclerectomy and sclerotomy during trabeculectomy to take care of cilliochoroidal detachment and choroidal effusion. However we thought that entering into suprachoroidal space before trabeculectomy will lead to hypotony and will make surgery more difficult and did not enter in to suprachoroidal space. This lead to sudden hypotony and development of choroidal effusion intra-operatively which resolved very slowly.

To conclude, this case highlights that in some cases with dialted episcleral vein, we may not able to diagnose the exact etiology despite through investigation. It also highlights the importance of sclerotomy in such cases of high episcleral venous pressure. It may be better to do sclerotomy and enter suprachoroidal space before trabeculectomy as sudden hypotony after trabeculectomy may lead to transudation of fluid in to suprachoroidal space and massive choroidal effusion during surgery.

Legend:

Figure 1a & 1b: anterior and lateral view of right eye showing dilated episcleral veins