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Eye (2006) **20,** 845–847. doi:10.1038/sj.eye.6702016;
published online 21 October 2005

Sir,

Disc drusen and peripapillary subretinal neovascular membrane in a child with the VACTERL association

We report a case of peripapillary subretinal neovascular membrane in association with optic disc drusen in a patient with the VACTERL (Vertebral defects, anal atresia, cardiac malformations, tracheoesophageal fistula with atresia, renal anomalies, and limb anomalies) association.

Case report

A 13-year-old girl with the VACTERL association was referred to the ophthalmology department complaining of reduced visual acuity in the left eye for the past 4 weeks and a possible diagnosis of papilloedema. A CT scan of the brain performed by the paediatricians was reported as normal. A lumbar puncture was considered, but a decision was made to refer her to the ophthalmic department for further investigations prior to the lumbar puncture.

On examination, her best-corrected Snellen acuity was 6/5 in the right and 6/18 in the left eye. The anterior segments were clear. Fundoscopy revealed a mildly elevated disc in the right eye. The left eye showed an elevated disc and a surrounding elevated lesion with subretinal exudation and haemorrhage. Extensive spotty and linear peripheral RPE lesions were also noted in the left fundus (Figure 1a). Ultrasound examination showed evidence of buried disc drusen in both eyes. Fundus fluorescein angiography revealed a large peripapillary subretinal neovascular membrane with

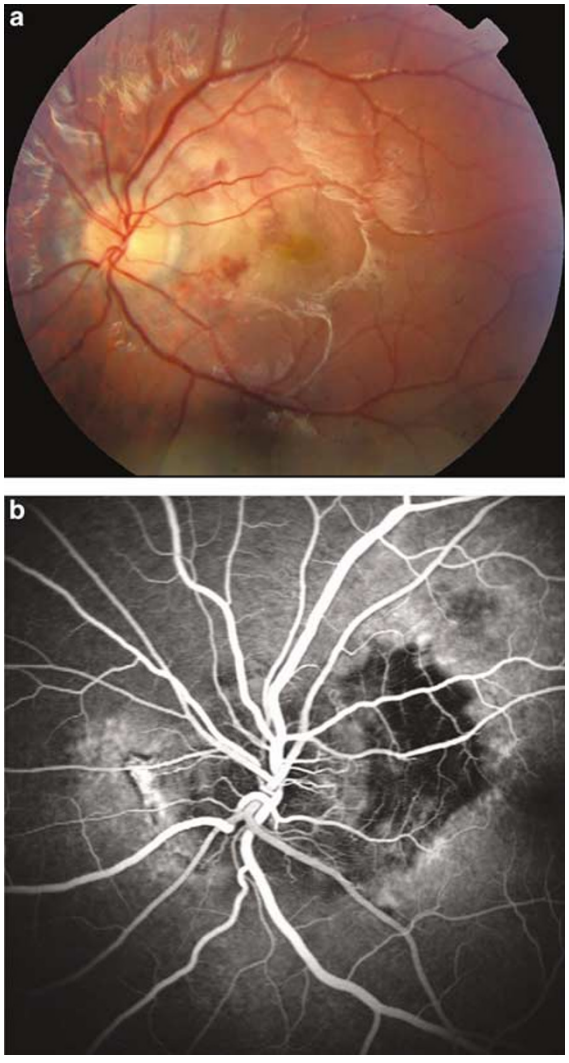

Figure 1 Coloured fundus photograph of the left eye showing a large peripapillary disciform with accompanying exudation and haemorrhage. (a) Pre-treatment fundus fluorescein angiography demonstrating the extent of peripapillary disciform. (b) Post-treatment fluorescein angiogram demonstrating a well-treated peripapillary disciform.

retinal thickening extending up to the fovea.
A diagnosis of a large peripapillary disciform in association with optic disc drusen was made. As the peripapillary disciform was threatening the fovea, it was decided to treat the lesion with argon laser photocoagulation. Follow-up at 2 weeks following treatment revealed an improvement in visual acuity, which was now 6/9–1 with marked resolution of retinal oedema. Fundus fluorescein angiography showed a well-treated peripapillary disciform (Figure 1b), although there was some residual leakage at the temporal disc margin.

Comment

The VACTERL association is a nonrandom association of malformations, which include vertebral anomalies, anal atresia, cardiac malformations, tracheooesophageal fistula, renal anomalies, and limb anomalies. Most cases of the VACTERL association are sporadic, with no recognised teratogen or chromosomal abnormality. However, features of VACTERL association have been reported with distal 13q deletion[1] and mitochondrial cytopathy.[2] It has been hypothesised that notochord anomalies allow ectopic expression of molecular signals in the developing embryo, and thus lead to VACTERL malformations.[3]

Ocular associations have very rarely been reported with the VACTERL association. Say *et al*[4] described ophthalmic abnormalities in four patients, which included ptosis, strabismus, cloudy cornea, severe myopia, anisocoria, and heterochromia iridis. Bilateral lacrimal anlage ducts, microphthalmos, anophthalmos, microcornea, optic nerve hypoplasia, nystagmus, and hemifacial microsomia[5,6] are the other ocular abnormalities that have been reported with the VACTERL association.

The present case adds optic disc drusen and a peripapillary subretinal neovascular membrane to the list of ocular associations seen in VACTERL association.

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Eye (2006) **20,** 847–848. doi:10.1038/sj.eye.6702018;
published online 24 February 2006

Sir,

The Ahmed glaucoma valve in refractory glaucoma: experiences in Indian eyes

We read with interest the article by Das *et al*[1] on their experience on the use of Ahmed valve in the treatment of refractory glaucoma among Indian eyes. The encapsulation rate shown in this paper are indeed very different from our paper published using a similar glaucoma implants in Asian eyes.[2]

It was mentioned in the article that no 'hypertensive phase' was observed and the authors attributed this to the continuous egression of aqueous through the dissected scleral flap. Such scleral flap is expected to be quite thin and certainly will not be as deep as what one would expect in nonpenetrating trabeculectomy as the authors made no attempt to create such depth at the time of dissection. If that was the case, egression of fluid through the scleral flap is not likely. If the egression of fluid is from the anterior chamber entry wound, the presence of the scleral flap would make no difference.

Furthermore, if there is still drainage through the scleral flap 4 weeks after the operation, it would be hard to determine whether the control of the IOP is due to the slceral flap draining or the Ahmed valve. If there is no encapsulated bleb, what would be the causes of failure?