Optic disc drusen precipitating central retinal vein occlusion in young

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DESCRIPTION

A 17-year-old boy presented with sudden painless diminution of vision in right eye for 3 days. The best corrected visual acuity was 20/400 and 20/20 in right and left eye, respectively. The intraocular pressures were 12 mm Hg in both eyes. There was associated relative afferent pupillary defect in right eye (OD). Anterior

Figure 1 Colour fundus photograph of right eye (A) shows hyperemic and swollen disc with extensive intraretinal and disc haemorrhages suggestive of CRVO. Similar optic disc features sans the retinal haemorrhages in left eye (B) is suggestive of pseudopapilloedema. CRVO, central retinal vein occlusion.

Figure 2 Horizontal optical coherence tomography scan through macula in right eye (A) shows cystoid oedema of outer nuclear and plexiform layer, neurosensory detachment and thickening of retinal nerve fibre layer. A marked reduction in oedema is noted after treatment with intravitreal bevacizumab (B).

Figure 3 Ultrasound B-scan image shows high reflective ovoid lesion at the optic disc representative of optic disc drusen in both eyes. OD, right eye; OU, both eyes.

segment examination was otherwise normal in both eyes (OU). Dilated fundus examination OD revealed optic disc oedema with hyperaemia, extensive intraretinal haemorrhages, engorged tortuous veins, macular oedema and cotton wool spots (figure 1A). Left eye had a swollen disc with blurred margins and the physiological cup was obscured (figure 1B). Optical coherence tomography (OCT) through macula in right eye showed thickened retinal nerve fibre layer, intra-retinal cystoid spaces and neurosensory detachment (figure 2A). B-scan ultrasound revealed pronounced disc oedema OD and an ovoid echogenic lesion deep within the optic disc with corresponding high amplitude spike on A-scan OU (figure 3A,B). He did not report any medical history and was not on any systemic medications. Based on above clinical and imaging findings, a diagnosis of optic disc drusen OU and central retinal vein occlusion (CRVO) with macular oedema OD was made. Investigations for thromboembolic risk factors were unremarkable. His blood pressure (120/80 mm Hg), fasting blood glucose (86 mg/dL) and serum lipid profile were normal. The macular oedema ebbed with two intravitreal bevacizumab (1.25 mg) injections at 1 month intervals and the corrected vision improved to 20/80 at 3 months before he was lost to follow-up (figure 2B).

Optic disc drusen are calcified hyaline-like deposits in the optic nerve head concentrated anterior to the lamina cribrosa.¹ Its incidence is 0.4%–20.4% in general population with female preponderance.² They are often bilateral

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Table 1 Patient demographics of the reported cases of CRVO association in eyes with optic disc drusen

Author	Year	Age/sex	CRVO severity	Systemic association	Treatment
Brodrick ¹²	1973	48 years/ female	Optic nerve head drusen associated with extensive retinal haemorrhages	Hypertension	None
Chern <i>et al</i> ¹³	1991	43 years/ male	Non-ischaemic CRVO	No systemic associations	None
Gallagher and Clearkin ¹⁴	2000	33 years/ female	Non-ischaemic CRVO	On oral contraceptives	None
Honkanen <i>et al</i> ⁹	2011	13 years/ male	Ischaemic CRVO	No systemic associations	Pan-retinal photocoagulation
Law <i>et al</i> ²	2014	52 years/ male	Non-ischaemic CRVO No macular involvement	No systemic associations	None

CRVO, central retinal vein occlusion.

(67%–91%) and are commonly found at the nasal aspect of the optic disc.² Largely they are benign and vision sparing but rarely may complicate with visual field defects, haemorrhages, choroidal neovascularisation, serous maculopathy and non-arteritic ischaemic optic neuropathy.^{3–5} Enlargement of drusen with advancing age is believed to cause mechanical compression at the lamina cribrosa leading to the complications.¹ Elevation of central retinal venous pressure may result in CRVO.^{1 2 6} The enlargement is observed mostly in teenage^{7 8} but to the best of our knowledge only a single case has been reported with concomitant CRVO in teenage.⁹ Unlike the hitherto reported case which resulted in neovascular glaucoma and retinal detachment, we achieved favourable outcome with intravitreal bevacizumab injection in the early treatment period. Follow-up at a later date could throw light on the long-term course of this vascular occlusion. This underscores active and early treatment of symptomatic CRVO in young when associated with optic disc drusen. Universal eye screening in children could go a long way detecting optic disc drusen early and imaging modalities such as ultrasound B scan and enhanced

depth optical coherence tomography can increase the sensitivity of the detection in buried drusen.^{10 11} Table 1 lists the reported association of optic disc drusen with CRVO in literature.

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Patient's perspective

I am happy that my disease could be diagnosed accurately at an early stage and the treatment in the form of eye injection restored my vision. I was informed about the rarity of the haemorrhagic complication and the risk of the other eye being involved.

Learning points

- Subjects with optic disc drusen should be made aware of the ominous prospects of a venous occlusion and followed closely.
- Ultrasound B scan of optic nerve head is a simple imaging tool to reveal embedded drusen.
- Optic disc drusen should be one of the differentials in young onset central retinal vein occlusions.

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