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Severe decompression retinopathy after medical treatment of acute primary angle closure

We describe a case of medically treated acute primary angle closure (APAC) resulting in severe visual loss owing to retinal haemorrhage. We are unaware of previous reports on this phenomenon in the world literature.

Case report

A 65-year-old woman visited her optometrist for a routine check. Her pupils were dilated and no pathology was found. Her vision progressively deteriorated that evening.

She attended the hospital and was noted to have unreactive mid-dilated pupils bilaterally. Visual acuity was 20/60 OU (pinhole). Intraocular pressure (IOP) was 60 mm Hg OD and 66 mm Hg OS. Gonioscopy showed occluded angles. The patient had moderate hypermetropia but otherwise had no other ocular risk factors for angle closure.

A diagnosis of iatrogenic APAC was made and the patient was given intravenous acetazolamide, topical dexamethasone, iodine 1% and pilocarpine 4%.

Figure 1 Colour fundus photograph showing severe pre-retinal haemorrhages at the posterior poles of the right eye, which occurred 2 h after medical intraocular pressure reduction for acute primary angle closure.

Figure 2 Colour fundus photograph showing severe pre-retinal haemorrhages at the posterior poles of the left eye, which occurred 2 h after medical intraocular pressure reduction for acute primary angle closure.

Ninety min later, IOPs were found to be 10 mm Hg OS. Acuity had improved to 20/30 OD and 20/20 OS.

Discharge was planned. After 2 h, the patient was still waiting for treatment when her vision deteriorated suddenly.

Visual acuity was hand motion OU. Pupils were small and IOP remained at 10 mm Hg OU. A clear fundal view was not possible because of miosis. B-scan ultrasound was undertaken, which raised the suspicion of a "macular bleed".

She was admitted and re-examined the next morning. IOP remained controlled and corneas were clear. Visual acuity had improved marginally to counting fingers OD and 20/200 OS. She had bilateral YAG peripheral iridotomies and was subsequently dilated. Funduscopy showed extensive bilateral retinal and pre-retinal haemorrhages (figs 1, 2). IOP remained normal 1 h after dilatation.

A diagnosis of bilateral decompression retinopathy secondary to rapid reduction in IOP was made. The patient was not taking aspirin, and had no evidence of bleeding diathesis, diabetes, vasculopathy or other systemic abnormalities that may have contributed to this complication.

Due to the severity of the visual loss it was elected to treat her surgically. She underwent bilateral vitrectomy within 1 month of onset and best corrected visual acuity improved to 20/20 in both eyes with normal IOP.

Comment

The case described is, to the best of our knowledge, unique in the literature. Decompression retinopathy is a rare complication that typically appears after glaucoma surgery. It usually manifests as transient scattered intraretinal haemorrhages at the posterior pole, which resolve within a few weeks, with visual acuity unaffected or slightly reduced.

Only three previous publications have described similar conditions.

Lai and colleagues¹ describe two cases of self-limiting retinal haemorrhages after argon laser peripheral iridoplasty for APAC. Both cases resolved conservatively and vision was restored. In other case reports following Waheeb and coworkers² describe a case of unilateral retinal haemorrhage YAG peripheral iridotomies for APAC. Nah *et al*³ describe a patient with APAC treated by peripheral iridotomy. In the latter report scattered retinal haemorrhages developed 2 weeks after treatment. The haemorrhages resolved over time with no visual sequelae.

The exact aetiology of decompression retinopathy is still a matter of speculation. The effect of acute hypotony on the eye may result in mechanical changes in the scleral shell, resulting in shearing forces, or the pressure reduction may simply facilitate an increase in arterial inflow compromising retinal capillaries by hydrostatic mechanisms. Autoregulation of these capillaries may already be defective, particularly when they have been subjected to persistently raised IOP. Decompression retinopathy may be part of the spectrum of central retinal vein occlusion, where IOP reduction causes buckling of the lamina cribrosa with compression of the central retinal vein.

This patient shows a worrying complication of acute reduction in IOP. The development of retinal haemorrhages after solely medical treatment for APAC is exceedingly rare. The severity of the retinopathy despite the absence of major hypotony at any stage is clinically relevant. The prognosis seems to be excellent even when the condition is severe.

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Cytological vitreous findings in a patient with infantile neurological cutaneous and articular (CINCA) syndrome

The chronic infantile neurological cutaneous and articular (CINCA) syndrome is a rare inflammatory paediatric disease identified by Prieur *et al*¹ in 1987. Ocular manifestations have been observed in 26% of patients² and include anterior uveitis, abnormal optic disc appearance and posterior inflammation with vitritis and vasculitis.³ This article describes cytological vitreous findings, obtained by pars plana vitrectomy, in patients with CINCA syndrome.

Case report

A 1-month-old boy was initially examined because of a bilateral anterior uveitis for 10 days. He was born after a normal pregnancy. On the second day, however, a generalised urticaria was observed and his serum C reactive protein level was raised. He presented with recurrent fever, leucocytosis, neutrophilia, increased erythrocyte sedimentation rate and

Figure 1 Ophthalmoscopic view during pars plana vitrectomy. The dense vitreous cells, and whitish deposits over the retina, pars plana and optic nerve are evident.

raised serum level of immunoglobulin (Ig)G. No evidence of infection was found, but hepatosplenomegaly was observed. At the same time arthritis developed, involving the knees, elbows and ankles. Neurological findings included chronic meningitis, and the patient's development was markedly delayed.

Anterior-segment examination showed two positive cells in the anterior chamber without posterior iris synechias. Funduscopy showed three positive dense vitreous cells that prevented the fundus of the eye being seen in both eyes. Topical steroids and mydriatics were initiated with high-dose intravenous methylprednisolone for 3 days followed by oral prednisone (0.5 mg/kg body weight/day). As no response to treatment was observed, a three-port pars plana vitrectomy was performed in the patient's right eye. During surgery, dense vitreous cells were observed with white infiltrates over the retina, optic nerve and pars plana (fig 1). Cytological examination of the vitrectomy specimen showed abundant cellularity, with a small number of polymorphonuclear leucocytes and lymphocytes and a dominant population of larger cells with wide, vacuolated cytoplasm and reniform nuclei with bland chromatin (fig 2A). Immunocytochemistry using a CD68 antibody was diffusely positive, confirming their macrophagic nature (fig 2B). Other diagnostic possibilities, such as granulomatous infections, xanthogranulomatous inflammation, metabolic storage disease, Whipple's disease and Langerhans cell histiocytosis were ruled out. The CINCA syndrome was established as a result of the clinical and pathological findings. To decrease steroidal side effects, ciclosporin A

(5 mg/kg/day) was added with no clinical benefits. The patient finally died at the age of 2 years.

Comment

The CINCA syndrome is an inflammatory disease characterised by persistent rash and chronic aseptic meningitis, with extensive infiltration of polymorphonuclear and macrophage cells at the sites of inflammation.⁴ The CINCA syndrome belongs to the group of systemic autoinflammatory diseases characterised by episodic or fluctuating degrees of inflammation, without evidence of high-titre autoantibodies or antigen-specific T cells. The disease is caused by mutations in the *CIAS1* gene that encodes a protein cryopyrin, NALP3 or PYPAF1.⁵ Mutations in cryopyrin have a profound pro-inflammatory effect. Cryopyrin is a caspase 1 activator, which in turn causes the activation of interleukin (IL)1 β . The activating mutations of cryopyrin induce an excessive activation of IL1 β , which causes an influx of macrophages and polymorphonuclear cells to the site of inflammation, in our patient, in his eye.⁶

We could not find any reference to previous pathological descriptions of the cell composition of the vitreous fluid in the CINCA syndrome. Only scattered reports on this entity contain pathological descriptions of skin and liver biopsy specimens, with unspecific inflammatory infiltrates composed of lymphocytes, neutrophils and eosinophils. However, current knowledge about the nature of this syndrome, which seems to be a phagocytic cell-mediated autoinflammatory disease, raises the possibility that the macrophagic infiltrate could be the hallmark of the disease, at least in the vitreous fluid.

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Figure 2 (A) Cytological smear of vitreous fluid: numerous histiocytes with bland nuclei and wide cytoplasm (May-Grunwald-Giemsa, 400 $\times$). (B) Immunocytochemistry in vitreous fluid: strong and diffuse cytoplasmic positivity with the monocyte/macrophage marker CD68.

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Intraocular central retinal artery occlusion during treatment for epistaxis

Maxillary artery embolisation with polyvinyl alcohol (PVA) is a procedure used for the treatment of intractable epistaxis. We report a rare but devastating complication of this otherwise safe and successful procedure.

Case report

A 46-year-old man presented to the accident and emergency department with a 1-day history of left epistaxis. He had had mild infrequent episodes in the past. He had cauterisation of the Little's area, but as the bleeding persisted, nasal packing was performed and blood tests were done. The platelet count was $310 \times 10^9/l$; other tests were normal. The epistaxis persisted the next day and a new bismuth iodoform paraffin paste nasal pack was inserted. He underwent a nasal endoscopy the next day and an arterial bleed in the area of the left sphenopalatine artery was observed. Electrocautery to this area was performed and a post-nasal pack was applied.

Despite these measures, the epistaxis continued for a further couple of days. The patient underwent carotid angiography via the left femoral artery under local anaesthesia. The catheter was advanced through the left common carotid artery, left external carotid and left internal maxillary artery. Selective endovascular embolisation of the left internal maxillary artery with 45–150 μ m polyvinyl alcohol (PVA) particles was carried out. By the end of the procedure, the patient experienced left-sided loss of vision. His blood pressure was 167/100 mm Hg, pulse 52 beats/min and SpO_2 99%.

The patient was examined by an ophthalmologist and was found to have no perception of light, afferent pupillary defect and classic signs of a central retinal artery occlusion in the