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 monocytic/macrophagic 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

left eye. A more recent photograph of his left fundus shows optic atrophy and retinal fibrosis. The draining veins of the affected temporal retina are occluded completely (fig 1). This is a unique fundus appearance not noted in vascular occlusions with more common calcific, fibrinoplatelet or cholesterol emboli.

Comment

The aetiology of retinal arteriolar occlusion is diverse. The most common causes were cholesterol and fibrinoplatelet (from a carotid source) followed by calcific emboli (from a cardiac source) in one case series of 70 patients.¹ Risk factors can be multifactorial and a comprehensive physical examination and systemic work-up is essential.² The treatment is directed to the cause. However, acute treatment of the condition to aid visual recovery is still disappointing.

The case described is an unusual, iatrogenic event. To our knowledge only one case of central and posterior ciliary artery occlusion after PVA embolisation has been reported³ (PubMed search for articles in English). Although reporting is not a reflection of true incidence, such an occurrence is still rare as it is only the external carotid system that is involved in embolisation, whereas the ophthalmic circulation is derived from the internal carotid system. This perceived notion of safety has been further supported by the advocacy of embolisation treatment as the primary treatment of intractable epistaxis, citing low complication rates in a large case series.⁴⁻⁵

The mechanism of PVA particles affecting the ocular circulation distant from their site of injection is manifold. Post-nasal bleeding is usually arterial and the vasculature is derived from both external (sphenopalatine and greater palatine branches of the internal maxillary artery) and internal carotid systems (anterior and posterior branches of the ophthalmic artery). The presence of these normal or any abnormal anastomoses (such as the artery of foramen rotundum, to the carotid siphon) provides anatomical access of embolic agents to the ophthalmic circulation.⁶

PVA particles are available in sizes ranging from 45 to 2000 µm. The smaller the particle size, the greater the ability to travel downstream and cause tissue destruction. Smaller particles occlude arterioles including retinal arterioles, as in our patient. Over-forceful injection can result in a reflux into the internal carotid system.⁴

The clinical appearance of retinal arterial occlusion owing to fibrotic agents like PVA is unique and distinct. This is due to the fact that polyvinyl alcohol particles are capable of inducing an intense fibrotic response (with a minimal amount of inflammatory cells) in tissues.⁷ This

Figure 1 Photograph of the left fundus showing retinal fibrosis.

can be of significance in pinpointing the aetiology where other risk factors may coexist. ENT and ophthalmic surgeons need to be aware of this rare but devastating complication occurring after PVA embolisation for epistaxis.

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MAILBOX

Patching treatment and bullying

We read with great interest the "Bullying and eye patching" commentary by Williams *et al*,¹ in which they report on the psychosocial implications of early versus late treatment of amblyopia. In their birth cohort study, they found that early detection (screening at 3 years and 1 month) and subsequent early treatment of amblyopia resulted in fewer reports of bullying victimisation by 8 years. On this basis, they appraised pre-school vision screening and suggested that treatment should be carried out as early as possible, to lessen the likelihood of bullying. While literature suggests that sustained bullying in childhood may have adverse psychosocial consequences in adulthood,² and although we agree that prevention of bullying is important, no evidence indicates that the children who undergo patching treatment for amblyopia have adverse consequences in terms of their well-being in the long term.

A growing body of evidence suggests that patching is associated with peer victimisation.^{1,3,4} In a recent study, we found that children reported not only experiences of bullying but also feelings of stigmatisation regardless of whether they were subjected to victimisation.⁵ This resulted in negative psychosocial outcomes for some children. However, whether bullying or feelings of stigmatisation have longer term psychosocial sequelae remains unanswered. Ideally, prospective longitudinal studies are needed to investigate the long-term psychosocial

consequences of patching for amblyopia. Although Williams *et al* have conducted a prospective study, they have only assessed bullying at one point in time (at the age of 8.5 years) and do not appear to have asked children what they perceived to be the cause of their victimisation. Without at least the child's perspective on this, it is impossible to ascertain whether these experiences relate to patching. Furthermore, the treatment of amblyopia usually always ceases well before the end of the first decade of life. Hence, patching, the source of a child's differentness within a group of peers, also ceases early in life. It is possible that short-term patching does not necessarily have an adverse effect on a person's psychosocial well-being in the long term. Evidence indicates that the removal of a stigmatising feature can have positive implications for a person. For instance, it has been reported that psychosocial functioning improves with corrective surgery for strabismus.⁶⁻⁹ More research is needed to investigate whether this similarly relates to patching.

We agree with the authors that bullying and other psychosocial factors should form part of treatment outcome measures; however, there is need for future research to also be attentive to the longer-term effects of patching treatment for amblyopia. Given the nature of this birth cohort study, the Avon Longitudinal Study of Parents and Children team may be able to discuss this in follow-up studies.

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Patient safety and ophthalmic surgery marking: which side are you on?

We read with interest the paper by Fraser and Adams,¹ which suggested many reasons for wrong site surgery and also proposed ways of