A case of bilateral uveitis and optic disc swelling with Chiari I malformation

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Abstract

We report a case of bilateral uveitis and optic disc swelling with Chiari I malformation. A 16-year-old girl was admitted to our clinic due to conjunctival hyperaemia and blurred vision in her right eye. Ophthalmologic and systemic examinations were performed. Visual acuity was 0.7 (OD) and 1.0 (OS). Bilateral optic disc swelling was observed. Fluorescein angiography demonstrated bilateral retinal vasculitis as well as optic disc hyperfluorescence due to leakage. Laboratory examinations were within normal limits. Cranial magnetic resonance venography imaging revealed neither cranial mass nor cerebral venous thrombosis but a Chiari I malformation. The patient was started oral cetazolamid, topical and oral corticosteroids. After six months follow-up, bilateral optic disc swelling was resolved completely and visual acuity was 1.0 in both eyes. Optic disc swelling may be associated with intraocular inflammation; however, patients with bilateral optic disc swelling should be suspected of having an accompanying intracranial pathology.

Keywords: Uveitis, Optic disc swelling, Chiari I malformation, Papilledema

Introduction

Chiari I malformation is a congenital malformation, characterized by herniation of cerebellar tonsils below the foramen magnum and into the cervical canal. Associated findings include a small posterior fossa, mild displacement of the medulla and/or the fourth ventricle, hydrocephalus, syringomyelia and osseous anomalies. Presentation frequently does not occur until the third decade of life. Although the most common presenting symptoms in patients with Chiari I malformation are headache, neck pain, extremity pain, weakness and paresthesias; patients may also be asymptomatic. In patients with Chiari I malformation associated hydrocephalus occurs in 7–10% of cases. Neuroophthalmological signs associated with Chiari I malformation consist of papilledema and ocular motility disturbances, including downbeat nystagmus and sixth cranial nerve palsy. Acute presentations of Chiari I malformations with isolated papilledema but without hydrocephalus are uncommon and are very rare in children. Co-occurrence of ocular inflammation and Chiari I malformation in a patient has not been reported in the published literature yet.

We report a patient of bilateral uveitis and optic disc swelling with Chiari I malformation to emphasize that optic disc swelling can occur either with intraocular inflammation or secondary to raised intracranial pressure.

Case report

A 16-year-old girl presented with a 4-day history of conjunctival hyperaemia and blurred vision in her right eye. Her past medical history and family history were unremarkable.
Her ocular examination revealed best-corrected visual acuities of 0.7 OD, and 1.0 OS. Intraocular pressures were 8 mm Hg OD, and 10 mm Hg OS. Biomicroscopic anterior segment examination revealed mild anterior chamber inflammation with ciliary injection, mutton-fat keratic precipitates, aqueous cells and flare in the right eye and normal findings in the left eye. Her fundus examination revealed bilateral optic disc swelling and venous engorgement without retinal exudates and oedema in both eyes (Fig. 1). A fundus fluorescein angiogram showed bilateral retinal vasculitis and optic disc hyperfluorescence due to leakage in both eyes (Fig. 1).

The laboratory testings included complete blood count, erythrocyte sedimentation rate, serum protein electrophoresis, immunoelectrophoresis, anti-nuclear antibody, anti-neutrophilic cytoplasmic antibody, anti-double stranded DNA antibody, anti-cardiolipin antibody, anti-phosphatidylserine antibody, rheumatoid factor, calcium and angiotensin converting enzyme levels, complements levels, Lyme western blot, Brucella agglutination, HLA tissue typing, rapid plasma reagin, urinalysis were all unremarkable. Chest X-ray and skin tests including tuberculin skin test and pathergy were performed and within normal limits.

Lumbar puncture revealed elevated cerebrospinal fluid opening pressure level which was 31-cm water and normal cerebrospinal fluid analysis levels. Cranial magnetic resonance imaging demonstrated a type I Chiari malformation without hydrocephalus and cranial magnetic resonance venography imaging demonstrated neither cranial mass nor cerebral venous thrombosis.

Treatment was initiated and consisted of oral acetazolamide 250 mg three times per day for intracranial hypertension, hourly instillation of topical corticosteroid, three times per day instillation of mydriatic drops (cyclopentolate 1%) and oral prednisolone 1 mg/kg/day, which was tapered and stopped in several weeks. The response of medical treatment was closely followed up. At the sixth month examination, visual acuity was 1.0 in both eyes. Ocular inflammation was inactive and disc swelling was disappeared in both eyes. A fundus fluorescein angiography was performed, neither retinal vasculitis nor optic disc hyperfluorescence were observed (Fig. 2).

Discussion

To our knowledge, this is the first report presenting coexistence of intraocular inflammation and Chiari I malformation with optic disc swelling without hydrocephalus. Obstruction of the cerebrospinal fluid flow at the level of foramen magnum in Chiari I malformation may cause intermittent intracranial pressure elevations. Acute presentations of Chiari I malformations without hydrocephalus but with isolated papilledema are uncommon. However, Milhorat and colleagues reported that nine (2%) of 364 patients symptomatic from Chiari I malformation had papilledema. Seven of the nine patients (80%) had normal-sized ventricles on cranial magnetic resonance imaging. In this age group, idiopathic intermediate uveitis may cause bilateral intraocular inflammation and primarily involves the anterior vitreous, pars plana and peripheral retina with an increased incidence of bilateral papilledema. The absence of vitreous snowballs, snowbanks and the presence of an elevated cerebrospinal fluid opening pressure with normal laboratory examinations made us to exclude intermediate uveitis as the reason of this clinical appearance in our patient.

Examinations revealed that our patient had uveitis and optic disc swelling. Although optic disc swelling may be caused by intraocular inflammation, intracranial hypertension should be ruled out in cases with bilateral papilledema. Our patient had an elevated cerebrospinal fluid opening pressure as well. Cerebral magnetic resonance imaging demonstrated that the patient also had a Chiari I malformation without hydrocephal-
lus. After the treatment with topical/oral corticosteroids and oral acetazolamid, disc swelling and vasculitis disappeared in both eyes and visual acuity improved in the right eye.

In conclusion, uveitis may exist with intracranial pathologies such as Chiari I malformation coincidentally. As already known, optic disc swelling may be associated with intraocular inflammation; however, patients with bilateral optic disc swelling should be suspected of having an accompanying intracranial pathology.

References