An unusual association of optic hydrops and Reiter’s syndrome

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ABSTRACT

The association of optic hydrops and Reiter’s syndrome in the literature has not been mentioned previously. This syndrome characteristically consists of 3 manifestations: arthritis, urethritis, and conjunctivitis. A 42-year-old woman with a history of Reiter’s syndrome presented with progressive vision disturbance in her left eye and with headache. Orbital MRI demonstrated enlarged periocular nerve subarachnoidal space on the left side.

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Case Reports

An unusual association of optic hydrops and Reiter’s syndrome

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The 3 most common manifestations of Reiter’s syndrome are arthritis, conjunctivitis, and urethritis.1 The involvement of the eye in Reiter’s syndrome is most commonly manifested as conjunctivitis; an autoimmune inflammation of the mucous membrane that covers the eyeball.2 Optic hydrops consists of isolated optic nerve sheath dilatation.3 It occurs without raised intracranial pressure (ICP) and may be caused by local lesions that lead to obstruction of the cerebral spinal fluid (CSF) flow in the optic nerve sheath.4

Case Report. A 42-year-old woman with symptoms of arthralgia, dysuria, fever and diarrhea was diagnosed with Reiter’s syndrome. Four weeks later she presented with progressive headache and fragmentary vision and photosensitivity in her left eye. She was not exposed to any recent febrile infection, and lumbar puncture revealed a normal opening pressure, with protein 1.3g/l, 94 white cells/(μl (100% lymphocytes) and glucose 2.7/5.6 mmol/l (CSF/plasma). Laboratory data were as follows: white blood cells 10.3 (normal 4-10 K/μl); hemoglobin 13.8 (normal 12.1-17.2g/dl); platelets 209 (normal 150-400 10e3/μl), H sensitive sedimentation rate 32 mm/h (normal, 0-20 mm h); and positive HLA-B27. Neuroophthalmologic examination revealed no afferent pupillary defect and papilloedema in each eye. Confrontational visual fields and the Humphrey automated visual fields showed no defect in both eyes. Axial T1-weighted orbital MRI, demonstrated edema-like change, and enlarged left optic nerve sheet (Figure 1). On coronal heavily T2-weighted orbital MRI in the right eye, optic nerve thickness was measured normally as 0.44 cm at the level of trunk, and 0.64 cm at the retrobulbar area (Figure 2). In the left eye, optic nerve thickness was measured 0.50 cm and 0.67 cm at the level of trunk and retrobulbar area respectively (Figure 2). Axial T1-weighted cerebral MRI was normal. Pharmacological treatment with a maximum dosage (3 x 250 mg) of acetazolamide for 2 weeks was effective. The aim of this paper is to present a very unusual association of optic hydrops and Reiter’s syndrome.

Discussion. Reiter’s syndrome is an autoimmune multisystem disease commonly triggered by a genitourinary infection or bacterial enteric infection.1 Reactive arthritis, is also called Reiter’s syndrome, the most common type of inflammatory polyarthritis in young men.2 The HLA-B27 genotype is a predisposing factor in over two-thirds of patients with reactive arthritis, as in our patient.5 The involvement of the eye in Reiter’s syndrome is most commonly manifested as conjunctivitis, which is present in up to 50% of affected individuals, and can develop at any time during the disease.2 Anterior uveitis, an inflammation of the inner eye, is the second most common ocular symptom of Reiter’s syndrome, occurring in up to 12% of affected persons.6 Other ocular conditions also associated with Reiter’s syndrome are scleritis, cataract, glaucoma, keratitis, papillitis, retinal and disc edema, and retinal vasculitis.6 Optic hydrops consists of isolated optic nerve sheath dilatation.4 It occurs without elevated ICP and may be caused by local lesions, which lead to obstruction of the CSF flow in the optic nerve sheath. However, an enlarged periocular subarachnoidal space occurs in some healthy persons and represents a variant of normal CSF spaces distribution without pathological value.4 However, our patient exhibited
fragmentary vision and photosensitiviness in her left eye after 4 weeks of the relapse. Pressure or arterial pulsation on the lower, open area of the arachnoidal sheath within the optic canal may cause a blockade of the perioptic CSF-filled spaces or with a valve-like effect, which can lead to optic hydrops. The subarachnoid space of the optic nerve cannot be regarded as a homogeneous space filled with CSF, but rather as a multi chambered and subdivided tubular system with a blind end (cul de sac) behind the ocular globe. It is agreed that there is a bulk circulation of fluid from the site of origin to the site of absorption that is, from the ventricles to the arachnoid villi in the cranial subarachnoid spaces. Jinkins, reported on 11 patients with optic hydrops secondary to various conditions, such as trauma, neoplasm, cysts and arachnoid adhesions. All these etiological conditions were eliminated in our patient. Magnetic resonance imaging demonstrates perioptic subarachnoidal space dilatation, but no obstruction along the optic subarachnoidal space. These are also observed in pseudotumor cerebri. In pseudotumor cerebri, there is raised ICP with normal CSF constituents, and MRI is usually normal. The abnormal immunologic function that was observed in the ocular inflammatory disease studied, may be the result of 2 immunopathologic phenomena. They are finalized by the non-self transformation of the antigens linked to various eye structures. These are represented by autoimmunity (of humoral cellular, or mixed type), and the immediate type III hypersensitivity reaction. On the contrary, this association may be coincidental in this patient, however, concurrent autoinflammatory changes of the arachnoid trabecula and septae may have had a role in the pathophysiology of this condition. The MRI scan of the brain and orbits, however, demonstrated localized and isolated stasis of fluid in the left optic nerve subarachnoid space only. The reason for this fluid congestion causing an optic nerve sheath compartment syndrome could not be identified by neuroimaging.9

Treatment of optic hydrops consists of the elimination of the causative lesion, an optic nerve decompression, or optic nerve sheath fenestration is caused by local lesions, which lead to obstruction of the CSF flow in the optic nerve sheath. Owing to normal ICP value, we administered acetazolamide orally aiming to decrease the CSF production. Generally systemic therapy (including immunosuppressive treatment) and the use of antibiotics to treat the underlying infections and anti-inflammatory medications are required to control obstinate ocular inflammation.10 In our patient, she recovered dramatically after administration of acetazolamide, which was evidence of a decrease in CSF production and provided the optic nerve decompression. If the administered pharmacological agents did not show any success, an underlying pseudotumor cerebri may be suspected, and ventriculo-peritoneal shunting and systemic therapy (including immunosuppressive treatment) might be required to control the ocular inflammation and to prevent progressive visual loss in such cases.

References


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**STATISTICS**


Describe statistical methods with enough detail to enable a knowledgeable reader with access to the original data to verify the reported results. When possible, quantify findings and present them with appropriate indicators of measurement error or uncertainty (such as confidence intervals). Avoid relying solely on statistical hypothesis testing, such as the use of *P* values, which fails to convey important information about effect size. References for the design of the study and statistical methods should be to standard works when possible (with pages stated). Define statistical terms, abbreviations, and most symbols. Specify the computer software used.