ANSWER TO PHOTO QUIZ (PAGE 452)

PROGRESSIVE VISUAL DECLINE IN A ROTTERDAM HARBOR CRANE OPERATOR

DIAGNOSIS

Fundoscopy and fluorescence angiography demonstrated papilledema and posterior uveitis, which may include optic neuritis from neurosyphilis in its differential diagnosis, but may also be due to central retinal artery or vein occlusion or elevated intracranial pressure because of a tumor, hypertension, a subarachnoid hemorrhage or a subdural hematoma.[1] The patient denied casual male sexual contact and had no history of sexually transmitted diseases. He could not remember having had a chancre or other clinical features of syphilis, and HIV testing was negative. Rapid pathogen reagin (RPR) testing and a treponema pallidum (TP) antibody test was positive in blood samples with an RPR titer of 1:64. Cerebrospinal fluid (CSF) tested negative for RPR but positive for TP Western blot. CSF also showed decreased glucose (2.1 mmol/l), increased protein (0.61 g/l), increased white blood cell count (12 x 10^6/l) and an increased immunoglobulin G (IgG) index (0.72). Hence, a diagnosis of neurosyphilis-associated posterior uveitis with papilledema was made and treatment was started with intravenous (i.v.) benzylpenicillin of three million international units, six times daily. However, visual abilities did not improve after two weeks of treatment. After deliberation, 60 mg prednisolone orally, once-daily was added with considerable improvement of visual ability.

Syphilis is a sexually transmitted disease, caused by the spirochete bacterium *Treponema pallidum*. Its presentation and clinical course are unpredictable, which is why it is called "the great masquerade". The clinical course of syphilis is divided into three different stages.[2,3] The first stage occurs after an incubation period of three to six weeks and is characterized by the appearance of a painless chancre at the site of transmission. The second stage occurs one to two months later, usually after resolution of the first stage.[2-4] This second stage is characterized by a macular rash on the palms and soles of the feet, fever, body aches, arthralgias and malaise.[2,3] Of patients with untreated secondary syphilis, two thirds will remain asymptomatic or latent after this stage, while the remaining one third will develop tertiary syphilis, months to decades after initial infection.[2] Asymptomatic syphilis has two stages: early latent syphilis (lues latens recens), where infection occurs within one year of infection; and late latent syphilis (lues latens tarda), where infection occurs more than one year after infection.[5] Tertiary syphilis results in slow, progressive damage to the nerves and blood. Neurosyphilis can occur at any stage.[3]

In neurosyphilis, ocular manifestations can be the presenting feature, but these typically occur in the secondary or tertiary stage.[2,3] The most common ocular finding in syphilis is uveitis, which occurs in 2-5% of patients in the tertiary stage.[2] Uveitis may be anterior, affecting the iris and ciliary body or posterior, involving the choroid, retina and retinal pigment epithelium.[3] Optic nerve involvement includes perineuritis, anterior or retrobulbar optic neuritis or papilledema.[3,4]

Primary treatment of syphilis involves high-dose i.v. penicillin. In addition, the use of corticosteroids (CSs) in syphilis may avoid/diminish the Jarisch-Herxheimer reaction, a hypersensitivity reaction caused by penicillin-induced spirochete death. Anecdotal case reports suggest that topical, periocular and systemic CSs may have an adjunctive role in the management of ocular syphilis, perhaps by suppressing intraocular inflammation and reducing uveitic macular edema. Adjunctive topical CSs have been found to be effective in the management of interstitial keratitis and anterior uveitis. Additional oral and i.v. CSs are used to treat posterior uveitis, scleritis and optic neuritis and led to notable improvement of visual ability in the present case.[3,4,6]

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