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Syphilis mimicking pseudotumor cerebri in an immunocompetent patient

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Abstract

Ocular syphilis and neurosyphilis are infrequently reported in immunocompetent patients. Generally, these conditions are seen in HIV positive individuals and commonly present with uveitis. Here, we describe a 40-year-old immunocompetent female who presented with ocular symptoms, found to have papilledema and was diagnosed with neurosyphilis.

Keywords: Syphilis; neurosyphilis; ocular syphilis; papilledema; optic disc edema.

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Introduction

Syphilis, often known as 'The Great Pretender,' may present with various signs and symptoms, making it difficult to distinguish from other diseases [1]. Ocular and neurosyphilis may occur during any stage of syphilis. Visual impairment occurs in 2-10% of the patients and can sometimes be the initial manifestation [2,3]. This report describes an immunocompetent female who presented with ocular symptoms and was found to have papilledema and diagnosed with neurosyphilis.

Case presentation

A 40-year-old female with history of attention deficit hyperactivity disorder, migraines and chronic obstructive airway disease presented to the hospital for persistent headaches and blurry vision. A few days after taking naproxen for headaches, she developed a pruritic, erythematous rash on her back, chest, and arms. She was prescribed prednisone and diphenhydramine, which helped clear the rash. The headaches were not abating, and she started retaking naproxen. She endorsed having a blurry vision for a few months along with bilateral tinnitus. She also endorsed bilateral wrist pain, and right sided wrist swelling. She reported rapid hair loss for 2 to 4 weeks. She reported being monogamous with her boyfriend of three years.

Physical exam was remarkable for a papular rash on the chest, abdomen, back, and upper extremities with sparing of her lower extremities and palms. She also had enlarged right anterior cervical lymph nodes. There was non-scarring diffuse alopecia on the scalp, but more pronounced over the temporal bone. A slit-lamp exam revealed fine pigment on the anterior lens capsule bilaterally. There were trace cells in anterior and vitreous chambers. Fundoscopic exam revealed bilateral frisen grade 3 disc edema with 360-degree elevation and blurring of small vessels at the margin. Visual fields and extraocular move-

ments were full. Visual acuity through pinhole without correction was 20/30 in the right eye and 20/30 in the left eye. Labs were remarkable for ESR 72 mm/h (0-20 mm/h) and CRP 14 mg/L (0-10 mg/L). HIV Ag/Ab and hepatitis panel were negative. RPR titer was 1:1024, and treponemal total antibody was >8. A lumbar puncture was performed. CSF showed 9 WBCs, 63% lymphocytes, glucose 62 mg/dl, protein 26 mg/dl. CSF VDRL was reactive. The opening pressure was 57cm H2O (normal <25 cm H20). CT scan of the orbits showed no abnormality.

She clinically improved with continuous high dose intravenous penicillin following desensitization and was discharged home to complete a 14-day course.

Discussion

Syphilis, caused by infection with Treponema pallidum, is a sexually transmitted disease with rising prevalence over the last few years. If untreated, the disease will progress through four stages of infection (primary, secondary, latent, tertiary). 47% of primary and secondary syphilis cases occur among men who have sex with men (MSM) only and men who have sex with men and women [1]. Although neurosyphilis is referred to as tertiary syphilis, Central Nervous System (CNS) involvement can occur at any stage. Early neurosyphilis usually occurs few weeks after infection and primarily involves the meninges and CNS vasculature compared to parenchymal involvement in late syphilis. In our case, serologic tests and the presence of rash and patient history indicate the secondary stage of syphilis.

Patients with ocular syphilis typically have anterior uveitis unless they are coinfected with the Human Immunodeficiency Virus (HIV), where there is the involvement of the posterior segment [4]. Optic nerve involvement, in the form of papilledema, perineuritis, or optic neuritis, is the second most common type of syphilitic ocular impairment [5]. Other syphilitic ocular manifestations include retinitis, vitritis, chorioretinitis, vasculitis, and panuveitis with or without change in visual acuity [6]. Ocular involvement in syphilis has a higher incidence in patients with HIV infection. In HIV-negative patients, the incidence is higher in women, and they also tend to present with more severe forms of visual impairment and complications such as chorioretinitis.

Papilledema refers to optic disc edema caused by Increased Intracranial Pressure (ICP). Patients with papilledema usually present with symptoms such as headache, nausea, vomiting, ataxia, diplopia or altered mental status. Usual causes of papilledema include idiopathic intracranial hypertension, subarachnoid hemorrhage, intracranial tumors, subdural hematoma, and intracranial inflammation. Optic disc edema, sometimes termed as papillitis, may also sometimes occur in conditions without increased ICP, such as a central retinal artery or central rein vein occlusion, congenital structural anomalies, and optic neuritis [7].

Rarely, papilledema or papillitis has been known to occur in immunocompetent patients with neurosyphilis [8-10]. Giant cell arteritis was considered in the differential, although this was unlikely given minimally elevated CRP and the absence of classic symptoms such as jaw claudication and scalp tenderness. Optic perineuritis (involvement of nerve sheaths without the involvement of the nerve itself) usually spares visual acuity and visual fields except for an enlarged blind spot [11]. Perineuritis was considered less likely given raised ICP and lack of radiologic findings. Given minimal inflammatory cells in the anterior and vitreous chamber, uveitis was considered unlikely. Optic disc swelling in the absence of signs of optic neuropathy supported the diagnosis of papilledema due to increased ICP and ruled out optic neuritis and papillitis. The etiology of papilledema is unclear, although it is likely related to early CNS involvement and meningeal infection-causing secondary intracranial hypertension.

The CDC guidelines recommend doing both non-treponemal and treponemal tests in order to confirm a diagnosis of syphilis [12]. All patients with ocular syphilis must undergo a neurologic exam and CSF evaluation via lumbar puncture. To diagnose neurosyphilis, the patient must have CNS or ophthalmic signs or symptoms, serologic evidence for syphilitic infection, plus one of the following: positive CSF-VDRL, increased CSF protein (>40 mg/dl), increased CSF WBC count (>5 mononuclear cells/uL). In our case, the patient had CNS and ophthalmic symptoms, positive CSF-VDRL, CSF pleocytosis, and positive serum treponemal test.

The treatment of ocular syphilis is the same as that of neurosyphilis. The mainstay of treatment is usually with 3 to 4 million units of intravenous penicillin G given every four hours or a continuous intravenous infusion of 18 to 24 units of penicillin G daily for 10 to 14 days [12]. In our first case, due to a reported Penicillin allergy, the patient had to be penicillin desensitized before initiating full dose treatment. The general guidelines for intravenous penicillin desensitization are double the dose at each step, starting with an initial dose that is 1/10,000 of the desired total dose over 15 minutes of continuous infusion. If there is no adverse reaction, then the next dosage is given. Alternate treatment regimens include 200 mg twice daily doxycycline for 28 days or ceftriaxone 2 g intravenously or intramuscularly daily for 10–14 days if desensitization to penicillin is not possible. However, the efficacy is not clearly known [13].

Conclusion

In conclusion, patients with syphilis and ocular symptoms must undergo prompt ophthalmic assessment, neurologic exam, and CSF evaluation via lumbar puncture. Treatment should not be delayed while awaiting tests as severe outcomes, including permanent vision impairment, have been reported. It is also essential to eliminate secondary etiologies before considering a diagnosis of idiopathic intracranial hypertension. Physicians and ophthalmologists must consider ocular and neurosyphilis as a differential diagnosis in patients presenting with papilledema to promptly diagnose and treat this condition and prevent the occurrence of blindness.

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