Incipient Syphilitic Papillitis

John J. Chen^{a,b}, M. Tariq Bhatti^{a,b}, Elizabeth Bradley^a, James Garrity^a, and Matthew J. Thurtell^c

^aDepartment of Ophthalmology, Mayo Clinic, Rochester, MN, USA; ^bDepartment of Neurology, Mayo Clinic, Rochester, MN, USA; ^cDepartment of Ophthalmology and Visual Sciences, University of Iowa, Iowa, IA, USA

ABSTRACT

Patients with syphilis can present with optic disc oedema (ODE) without visual compromise, which has been primarily attributed to papilloedema from raised intracranial pressure or optic perineuritis from optic nerve sheath inflammation. We report four cases of ODE in the setting of syphilis with preserved visual function, normal intracranial pressure, and no enhancement of the optic nerve or sheath on magnetic resonance imaging. We propose the term “incipient syphilitic papillitis” for cases presenting with ODE, preserved vision and absence of optic nerve sheath enhancement, which is likely a more common presentation than syphilitic optic perineuritis.

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Introduction

Syphilis is a sexually transmitted infection caused by the spirochete *Treponema pallidum*, which can affect any part of the eye. Ocular manifestations can include interstitial keratitis, uveitis, chorioretinitis, retinitis, retinal vasculitis, optic neuropathy, and other cranial neuropathies.^{1,2} Optic nerve involvement can result from optic neuritis, optic perineuritis, neuroretinitis, papilloedema, or papillitis with or without retinal vasculitis. Optic nerve involvement and retinitis are considered synonymous with neurosyphilis, and therefore require long and high-dose treatment with intravenous antibiotics, typically penicillin.^{3,4}

Optic disc oedema (ODE) due to syphilis without visual compromise has been primarily attributed to papilloedema from raised intracranial pressure or optic perineuritis from optic nerve sheath inflammation.⁵ However, these terms were proposed before magnetic resonance imaging (MRI) was largely available. We report four cases of ODE due to syphilis with normal visual function and no MRI enhancement of the optic nerve sheath.

Methods

Retrospective chart review of four patients with serology-positive syphilis, ODE, and preserved visual function.

Results

Three males and one female with median age 33 years (range 30 to 63 years) presented with ODE from syphilis (Table 1). Unilateral ODE was present in all of the patients (Figures 1 and 2). None of the patients had symptoms to suggest raised intracranial pressure. Three were diagnosed with syphilis because of the eye findings while one patient was found to have ODE after he was diagnosed with neurosyphilis due to symptoms of back pain and numbness. Although three of the four patients reported subjective blurring of vision or transient visual obscurations, visual acuity was unaffected, except for one patient with visually significant posterior capsular cataracts (Figure 1 and Table 1). In all cases colour vision was normal as were visual fields other than enlarged blind spots. No patient had a relative afferent pupillary defect, iritis, or vitritis. MRI showed no enhancement of the optic nerve or optic nerve sheath in any of the patients. Three patients had isolated enhancement of the optic disc consistent with papillitis (Figures 1 and 2). Rapid plasma reagin (RPR) was positive in all patients. The cerebrospinal fluid (CSF) opening pressure was normal (≤ 20 cmCSF) in two patients, but was not

Table 1. Demographics, clinical findings, neuroimaging manifestations, paraclinical test results, and visual outcome.

Patient	Age	Gender	Presenting symptoms	Affected Eye(s)	Ocular findings	Serum serologies	CSF studies	Orbit MRI findings	Time to resolution of disc edema	Initial visual acuity	Final visual acuity
1	36	M	Transient visual obscurations OS for 1 week before presentation	OD	ODE OD	Positive syphilis IgG RPR 1:64 HIV positive	OP not obtained WBC 3/mm ³ Protein 31 mg/dL CSF VDRL neg	Trace optic nerve head enhancement	4 weeks	20/20 OU	20/20 OU
2	30	F	Gradual progressive bilateral blurred vision over 12 months	OS	ODE OS PSC cataracts OU	Positive syphilis IgG RPR 1:64 Reactive TPPA	OP 12.5 cmCSF WBC 2/mm ³ Protein 35 mg/dL CSF VDRL n/a	Optic nerve head enhancement	3 weeks	20/80 OU	20/20 OU (pseudophakic)
3	30	M	Subjective blurred vision and transient visual obscurations for 2.5 months before presentation	OS	ODE OS	Positive syphilis IgG RPR 1:64	OP 20 cmCSF WBC 2/mm ³ Protein 48 mg/dL CSF VDRL neg	Normal	3 weeks	20/20 OU	20/20 OU
4	63	M	Progressive low back pain and numbness over 8 weeks. No visual symptoms.	OS	ODE OS	Positive syphilis IgG RPR 1:128	OP not obtained WBC 27/mm ³ Protein 111 mg/dL CSF VDRL pos	Optic nerve head enhancement	3 weeks	20/20 OU	20/20 OU

Abbreviations: ODE = optic disc oedema; MRI = magnetic resonance imaging; CSF = cerebrospinal fluid; WBC = white blood count; OP = opening pressure; RPR = rapid plasma reagin; TPPA = treponema pallidum antibody; VDRL = venereal disease research laboratory test; HIV = human immunodeficiency virus; PSC = posterior subcapsular; n/a = not available

Figure 1. 30-year-old female with unilateral ODE in the left eye and bilateral posterior subcapsular cataracts (patient 2 in Table 1). Fundus photograph demonstrates hyperaemic disc oedema in the left eye. The view was hazy due to significant posterior subcapsular cataracts. T1-weighted magnetic resonance imaging of the orbits with contrast and fat suppression demonstrates enhancement of the optic nerve head on the left (red arrow), but no enhancement of the optic nerve or optic nerve sheath.

Figure 2. 63-year-old male with unilateral ODE in the left eye (patient 4 in Table 1). Fundus photograph shows mild ODE with a few nasal peripapillary haemorrhages. Humphrey visual field in the left eye is full. T1-weighted magnetic resonance imaging of the orbits with contrast and fat suppression demonstrates enhancement of the optic nerve head on the left (red arrow), but no enhancement of the optic nerve or optic nerve sheath.

measured in the other two patients. One patient had a CSF pleocytosis, elevated CSF protein, and positive CSF venereal disease research laboratory (VDRL) testing, while the other three patients had normal CSF constituents (Table 1). One

patient was previously known to be human immunodeficiency virus (HIV) positive. The rest of the infectious and inflammatory workup was negative in these patients, including ANA, ACE, ANCA, Lyme, and quantiferon. All

patients were treated with intramuscular or intravenous penicillin for 14 days with complete resolution of the ODE.

Discussion

We describe four cases of unilateral ODE from syphilis with preserved visual function. There was no optic nerve or optic nerve sheath enhancement on MRI in any of the patients. The opening pressure was normal in the two patients in which it was measured. The primary site of pathology appeared to be inflammation of the optic disc, as demonstrated by isolated enhancement of the optic disc on MRI in three of the patients. This case series highlights the importance of considering syphilis in the differential diagnosis of a patient presenting with unilateral ODE with preserved visual function, even in the absence of vitritis. It is unclear how long the ODE would last in these patients if untreated or whether this would progress to vision loss because these patients were treated with IV or IM penicillin upon diagnosis with fairly fast resolution of the ODE. One patient did have ODE for 2.5 months

before being seen in the neuro-ophthalmology clinic and maintained 20/20 vision, and therefore the ODE can be longstanding in at least some instances without degrading vision.

The terms papillitis, optic perineuritis, and optic neuritis were applied to syphilitic involvement of the optic nerve prior to the advent of MRI.⁵⁻⁷ Therefore, it was not possible to determine the precise region of optic nerve involvement in the past. Walsh distinguished optic perineuritis and optic neuritis based on the examination findings and visual status, where optic perineuritis was defined by ODE without loss of vision.⁵ In the era of MRI, the term optic perineuritis has been carried over to recent descriptions of preserved vision in the setting of ODE despite a lack of dedicated neuroimaging of the orbits to confirm or refute the presence of optic nerve sheath enhancement,⁸⁻¹³ and some cases have been erroneously used to describe this clinical entity despite a lack of optic nerve sheath enhancement.^{14,15} In contrast, the term papillitis has typically been used in the setting of patients with ODE and vision loss,^{16,17} although there are recent reports of

Figure 3. Algorithmic approach to ODE from syphilis.

papillitis similar to our patients with preserved vision.^{18,19} There is a single prior report of a patient with radiological evidence of optic nerve sheath enhancement from neurosyphilis resulting in bilateral ODE and preserved vision.²⁰ However, based on our case series and a review of the literature, this may represent the minority of cases of asymptomatic ODE in the setting of neurosyphilis.

Instead of speculating on the anatomical region of syphilitic optic nerve involvement, Graveson stated in 1950 that 'in the absence of precise pathological details it has, therefore, been felt desirable to describe the condition as one of optic neuritis.'⁶ In addition, he stated that 'papillitis may possibly prove to be a more exact description, but it suggests a restriction of the disease process which is, as yet, unsupported by pathological findings.' However, with MRI to evaluate for retrobulbar and optic nerve sheath enhancement and based on our findings we believe the term 'incipient syphilitic papillitis' should be used to describe cases of ODE with preserved vision in the setting of normal intracranial pressure and lack of optic nerve sheath enhancement on MRI. We propose a new algorithmic approach to ODE from neurosyphilis that includes MRI in elucidating the underlying anatomical region of the anterior visual pathway (Figure 3).

Disclosure of Interest

The authors report no conflict of interest.

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