

# Syphilitic Optic Neuropathy: Diagnosis Not To Be Missed

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## Abstract

**Purpose of the study:** To describe two cases of syphilis presenting with optic neuropathy and to highlight the importance of considering syphilis in the evaluation of optic neuropathy.

**Method:** Complete physical and ophthalmologic examination was performed in patients along with the necessary laboratory investigations including cerebrospinal fluid (CSF) analysis, syphilis serology in serum and CSF and HIV serology.

**Results:** Both patients presented with unilateral optic perineuritis evident as swollen optic discs, normal visual acuity and normal pupillary reactions to light and accommodation. Complete recovery of optic neuropathy was achieved in both patients following treatment with procaine penicillin which was administered with oral probenecid and a short course of steroids.

**Conclusion:** Early diagnosis and prompt treatment with penicillin is essential to prevent visual impairment in syphilitic optic neuropathy.

## INTRODUCTION

The incidence of sexually transmitted diseases (STD) including syphilis has been rising in recent years [1, 2]. The ability of this illness to imitate different ocular disorders often results in misdiagnosis or delayed diagnosis. We report two cases of syphilis presenting with optic neuropathy. As it is not commonly encountered by clinicians, the most important aspect of diagnosing ocular syphilis is having a high index of clinical suspicion.

## CASE HISTORY

### CASE 1

A 40 year old man presented to the eye clinic with intermittent painless visual blurring in his left eye for 2 weeks. Six months earlier, he had had a retinal detachment of his left eye and was concerned that he might be developing a recurrence. He denied other neurological or systemic symptoms.

On examination visual acuity was 6/6 in both eyes with an enlarged blind spot in the left eye. Pupillary reactions and colour vision was normal in both eyes. Fundoscopy revealed a markedly swollen left optic disc (Figure 1.1). The right optic disc was normal. There were no signs of uveitis and

retinal vasculitis. A working diagnosis of optic neuritis was made. He presented to the genitourinary medicine clinic several days later with a history of feeling tired for a few weeks following several unsafe sexual contacts. On direct questioning, he admitted to a history of skin rash on his torso and the inner aspects of his arms six months earlier which lasted for 4-6 weeks.

On investigations, his full blood count and ESR were normal. Serum treponemal EIA and TPA serology was positive with a VDRL titre of 64. MRI of brain and orbit was normal. CSF examination showed 6 white cells/mm<sup>3</sup>; protein of 0.28g/L and glucose of 4mmol/L (serum glucose of 5.2 mmol/L). No organisms were seen in the CSF. CSF TPA and VDRL were positive thereby confirming the diagnosis of neurosyphilis. He was treated with intramuscular procaine penicillin 2.4 grams once daily and probenecid 500 mg 6 hourly for 17 days along with a short course of prednisolone. Following treatment, the left optic disc swelling had almost fully resolved six weeks later (Figure 1.2).

### Figure 1

Figure 1.1: Left eye optic disc swelling at presentation (case 1).



### Figure 2

Figure 1.2: Optic disc swelling had fully disappeared six weeks following treatment.



### CASE 2

A 27 year old man presented with a 4 months history of generalised headache. Five days prior to presentation he awoke with left sided facial and limb weakness which improved spontaneously though incompletely over the subsequent days. No visual disturbance was reported. Six weeks earlier he noticed a generalised skin rash. On examination visual acuity was 6/6 in both eyes with mild enlargement of the right blind spot. The right optic disc appeared swollen and haemorrhagic (figure 2.1). Pupillary reactions to light and accommodation were normal with no

evidence of a relative afferent pupillary defect. Colour vision was normal in both eyes. A mild left lower motor neuron facial weakness was noted but neurological examination was otherwise unremarkable. General examination revealed a maculopapular skin rash over the torso, arms and legs with involvement of palm and soles (figure 2.2). Inguinal and axillary lymphadenopathy was also noted. A systemic multifocal inflammatory disturbance was suspected. Blood tests showed normal full blood count and, an elevated CRP of 28 and ESR of 27 mm/hr. MRI of the brain and orbits revealed abnormal enhancement of both facial nerves following gadolinium. Lumbar puncture showed opening pressure of 20 cm of CSF, a white cell count of 80/mm<sup>3</sup> (90% lymphocytes); protein of 1.15 g/L (normal < 0.40 g/L) and glucose of 3.7mmol/L (serum glucose of 3.8mmol/L). Serum Treponemal EIA was positive. Serum TPA was positive at a titre greater than 5120 and VDRL titre was 64. Elevated antisyphilis reactivity was concurrently shown in the CSF with a TPA titre of 320 and VDRL titre of 2. Treatment was the same as above. Four months later the right optic disc swelling had fully resolved (figure 2.3).

### Figure 3

Figure 2.1: Right eye optic disc swelling at presentation (case 2)



**Figure 4**

Figure 2.3: Right eye following treatment



**Figure 5**

Figure 2.2: Maculopapular rash of secondary syphilis



## DISCUSSION

These two cases highlight the importance of considering syphilis in the differential diagnosis of optic neuropathy.

Ocular involvement primarily occurs in the secondary stage and less commonly in the tertiary stage of the disease. Numerous ocular manifestations include uveitis, scleritis, episcleritis,

dacroadenitis, interstitial keratitis, vitritis, chorioretinitis, retinal vasculitis, serous retinal detachment, optic neuropathy, gumma of the optic disc and cranial nerve palsies [3].

Optic nerve involvement in syphilis can manifest as papillitis, anterior or retrobulbar optic neuritis, papilloedema, and optic atrophy [4]. The clinical presentation in our two patients is in keeping with papillitis, also known as optic perineuritis. Typically the acute optic disc swelling which occurs in papillitis causes minimal if any visual symptoms and does not show any typical characteristics to distinguish it from other causes of optic nerve disease. If a high index of suspicion is not maintained, the underlying diagnosis of syphilis could easily be missed.

On the background of the its recent resurgence, syphilis should be considered in the differential diagnosis of optic neuropathy and investigated accordingly. Acute optic disc swelling may occur on its own as in first case or as a manifestation of the systemic inflammation that can accompany secondary syphilis as was seen in the second case. In either situation it is important to confirm involvement of the CNS by examining the CSF. The recommended treatment for ocular syphilis is the same as for neurosyphilis and involves intravenous penicillin G or intramuscular procaine penicillin for 10-14 days along with oral probenecid [3].

## References

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