

# Optic Disc Edema in Syphilis: Lighting Up the Diagnosis

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## Introduction

- Syphilis = spirochete *Treponema pallidum* infection
- Known as “the great imitator” due to variable clinical manifestations in patients
- Early diagnosis is critical as antibiotic treatment is curative

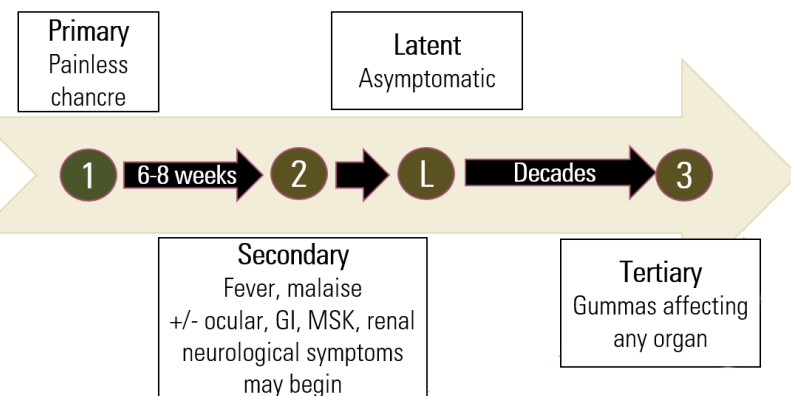


Figure: Stages of Syphilis and Disease Manifestations

- Ocular involvement = neurosyphilis** (prompts neuroimaging and CSF analysis)
- Most commonly manifests as uveitis, chorioretinitis, optic disc edema, optic neuritis, perineuritis, papilledema
  - May be present at any stage of disease

**Purpose:** We describe four patients who presented with swollen optic disc(s) without overt signs of intraocular inflammation, later determined to be secondary to syphilis.

## Methods

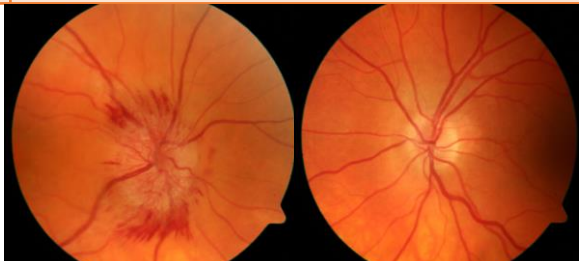
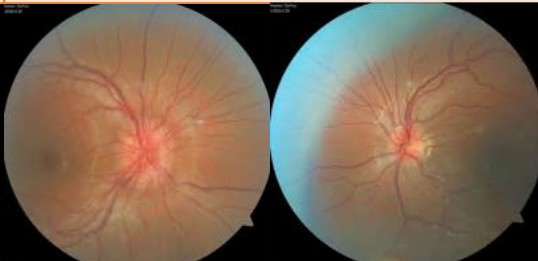


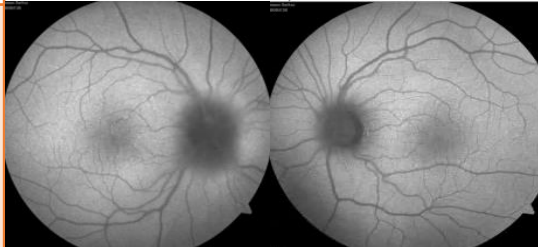
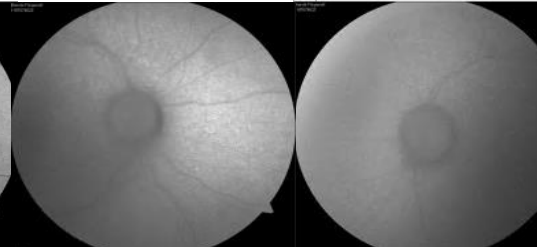

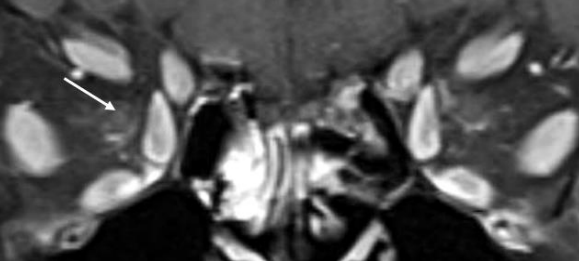
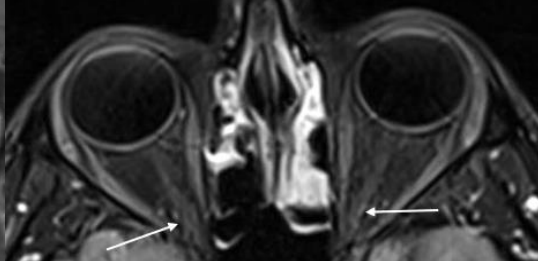
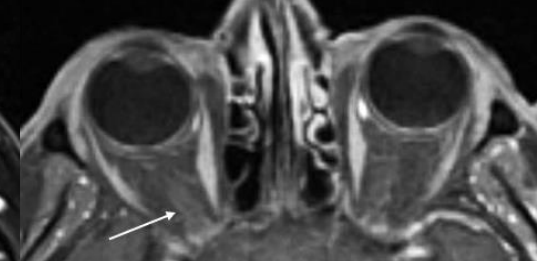

Patients who presented to a tertiary neuro-ophthalmology practice with a swollen optic nerve head(s) but no overt signs of intraocular inflammation which was eventually determined to be secondary to syphilis were included.

## Summary and Conclusions

### Summary of Cases

Characteristic	# of cases
Systemic manifestations	3 of 4 (2 with rash, 1 with abdominal pain and elevated liver enzymes)
High-risk group status	3 of 4 (two MSM, 1 sex worker); <b>0 patients disclosed this at first visit</b>
<b>Visual function</b>	
Severely decreased	2 of 4
Normal	2 of 4 (both had photopsias)
<b>Ocular inflammation (vitritis/chorioretinitis)</b>	
Obvious at presentation	0 of 4
Subtle chorioretinitis seen on autofluorescence	3 of 3
Developed later	4 of 4
Perineuritis on imaging	3 of 3

- Systemic symptoms are common in patients with syphilitic optic neuropathy.
- Early recognition is critical as antibiotic treatment is curative.
- Index of suspicion should be high for all patients, even without the presence of risk factors.
- Patients with optic disc edema complaining of photopsia should all be investigated for syphilis.
- Fundus autofluorescence is a very useful test and is expected to be consistent with subtle chorioretinitis.
- Optic perineuritis is common in patients with syphilitic optic neuropathy, its pathophysiology likely similar to meningitis seen in neurosyphilis.

	Case 1	Case 2	Case 3	Case 4
<b>Age, gender</b>	58, male	30, male	62, female	25, female
<b>Laterality</b>	Unilateral	Bilateral	Bilateral	Unilateral
<b>Vision</b>	20/20 both eyes	20/20 both eyes	Counting fingers, 20/400	20/400, 20/20
<b>Symptoms</b>	Right eye photopsias, truncal rash	Bilateral photopsias, rash on palms and soles	Bilateral progressive decreased vision, abdominal pain	Blurry vision OD
<b>RAPD</b>	Mild	None	Present	Present
<b>Other ocular findings</b>	OD mild vitreitis	Chorioretinitis both eyes, punctate hyper-autofluorescence	OS anterior chamber cells, keratic precipitates, bilateral vitreitis, punctate hyperautofluorescence	OD vitreitis and chorioretinitis, punctate hyper-autofluorescence
<b>Visual fields</b>	Right blind spot enlargement, left normal	Right blind spot enlargement, left normal	OD diffuse depression, OS nasal	OD central scotoma
<b>RNFL thickness</b>	362 um OD, 91 um OS	304 um OD, 104 um OS	120 um OD, 121 um OS	283 um OD, 103 um OS
<b>HIV serology</b>	Negative	Negative	Negative	Negative
<b>Lumbar puncture</b>	Not done	Indeterminate serology	Positive serology	Negative serology
<b>Outcome after treatment</b>	Visual symptoms and edema resolved	Visual symptoms and edema resolved	Vision improved to OD hand motions, OS 20/50, developed retinal detachment in OD	Vision improved to 20/50, 20/20
<b>Fundus photos</b>				
	Right disc edema with peripapillary hemorrhages	Asymmetric disc edema	Bilateral temporal pallor and mild elevation	Right optic disc edema
<b>Fundus autofluorescence</b>	N/A			
	N/A	Small punctate foci of hyperautofluorescence bilaterally	Hyperautofluorescence in the peripapillary region bilaterally	Multiple hyperautofluorescence dots scattered in OD
<b>Neuro-imaging</b>				
	Coronal T1-fat sat post contrast MRI showing subtle enhancement of the right optic nerve sheath.	Axial T1-fat sat post contrast MRI showing subtle enhancement of the bilateral optic nerve sheaths.	Axial T1-fat sat post contrast MRI showing enhancement of the right optic nerve sheath.	Coronal T2 MRI showing increased signal in the right optic nerve sheath. No contrast-enhanced study performed.

**Table:** Detailed description of cases. OD: right eye, OS: left eye, ON: optic nerve, MRI: magnetic resonance image, RNFL: retinal nerve fiber layer, RAPD: relative afferent pupillary defect