

Wernicke's encephalopathy: Is visual loss a red herring?

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Abstract

Predominantly visual loss, is very rare in Wernicke's encephalopathy. A 22 year old lady, in her 28th week of gestation, presented with a confused mental state, bilateral papilloedema with retinal hemorrhages, ophthalmoparesis, and cerebellar signs. Her MRI brain was suggestive of Wernicke's encephalopathy and she recovered with intravenous thiamine.

Title Page

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Key Words:

Papilloedema, Retinal Hemorrhages, Hyperemesis gravidarum, ophthalmoparesis, Thiamin

Introduction:

“Polioencephalitis hemorrhagica superioris” was the original term coined by Carl Wernicke in 1881 to describe the classical syndrome, with a triad of ophthalmoparesis, ataxia and mental status changes. Typically, it is encountered in clinical situations involving poor nutrition such as alcoholics, post gastrointestinal surgery, hyperemesis gravidarum, malignancy and other such states.¹Operational criteria proposed for Wernicke’s encephalopathy need any two of the following: Dietary inadequacies, eye movement abnormalities, ataxia and either an altered mental state or memory disturbances.² Thiamine is the mainstay of treatment. Although

in the initial series of Adams, 100 mg per day was recommended, many subsequent studies recommend 500 mg, initially, parenterally.^{3,4}

Visual loss is an unusual and rare manifestation of Wernicke's encephalopathy. In the original series of Adams et al, only 3 of 232 patients had retinal haemorrhages with visual loss. ⁴Papilloedema with Wernicke's encephalopathy has been described only rarely in a few case reports. We describe a patient of Wernicke's, who presented with predominantly visual loss.

Case Report:

A 22 year old lady, presented with a history of blurring of vision since 6 days. The visual blurring was acute in onset and progressive over 3 days. She was 28 weeks pregnant and had a history of vomiting since 3 months. Clinical examination revealed that, although conscious, she was confused and not oriented to time, place or person. Her bilateral corrected visual acuity was 6/9 bilaterally. There was a bilateral sixth nerve palsy. She was able to stand with minimal support and had stance/ gait ataxia. Further, there were bilateral cerebellar signs on examination.

Funduscopy revealed bilateral disc edema with retinal hemorrhages (Figure 1: A and B). Her Blood investigations revealed a sodium of 128 meq/ l, potassium of 2.9 meq / l and urine ketone bodies were positive. An echocardiogram done revealed normal cardiac function. A nerve conduction study of all 4 limbs was done, which was normal.

MRI brain done showed flair hyperintensities in bilateral medial thalami, periaqueductal grey matter and mamillary bodies (figure 2). Visual fields showed severe constriction bilaterally (Figure 3 A and B: Right and Left eyes respectively). Thus, a diagnosis of Wernicke's encephalopathy was made.

The patient was treated with Thiamine parenterally, 500 mg iv on the first day followed by 100 mg intravenously thrice a day for 7 days. Her vision improved dramatically with treatment. The bilateral sixth nerve palsy and cerebellar signs also resolved in 1 week. She was continued on oral thiamine 100 mg twice daily for a month.

The disc edema resolved completely in 1 month (Figure 1 C and D) and visual fields improved too (Figure 3 C and D, Right and Left eyes respectively)

Discussion:

Wernicke's encephalopathy is a serious nutritional disorder with potentially life- threatening consequences, if unrecognized. Although, the triad of ophthalmoparesis, changes in higher mental functions and ataxia are easily recognized, there may be confounding clinical features such as blurring of vision, which is rarely seen, thus delaying the diagnosis. Among the ocular findings, nystagmus is most common, followed by bilateral 6th nerve paresis, conjugate gaze palsy, scotomata, pupillary abnormalities, retinal haemorrhages, ptosis, and disc edema. ⁵

Disc edema is a rare finding in Wernicke's encephalopathy.⁶ Retinal haemorrhages are seen more frequently, sans disc edema. These manifestations have been described with bariatric surgery too. In the patients described, disc edema with retinal hemorrhages accompanied the vision loss, along with other characteristic manifestations. The patients showed a good response to intravenous thiamine. ^{7,8}

Mumford described a similar case of disc edema with retinal hemorrhages in a case of hyperemesis gravidarum, with other features of Wernicke's encephalopathy, who responded well to parenteral thiamine administration.¹

The mechanisms underlying the visual manifestations are not clear. It is likely that an optic neuropathy associated with Wernicke's encephalopathy, due to nutritional deficiency of thiamine could result in the disc edema. This is consistent with quick resolution of fundoscopic findings with thiamine administration. Another plausible explanation is the possible necrosis of nerve cells with edema, which underlies neuropathology of Wernicke's.¹

Conclusions:

Early treatment with parenteral thiamine can prevent the cognitive complications of Wernicke’s encephalopathy. Our case illustrates that, in a setting such as pregnancy, where there is an enhanced metabolic requirement of thiamine, one must have a high index of suspicion in patients presenting with visual loss and other features of Wernicke’s encephalopathy. This would prevent a potential delay in diagnosis and facilitate urgent treatment with parenteral thiamine.

Author contributions:

Sujit Kumar: Clinical management of the case and preparation of manuscript

Abdul Rawoof Bolar: Neuroophthalmological evaluation of the case and preparation of manuscript

Rohit Shetty: Neuroophthalmological evaluation of the case and preparation of manuscript

Sharath Kumar Goddu Govindappa: Neuroradiological assessment and preparation of manuscript

Manithody Narayana Bhat Pramod: Clinical management of the case and preparation of manuscript

Jagadish Basavaraj Agadi: Clinical management of the case and preparation of manuscript

Lakshminarayanapuram Gopal Vishwanathan: Clinical management of the case and preparation of manuscript

Chaitra Prameshwara Adiga: Neuroradiological assessment and preparation of manuscript

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Legends:

Figure 1: A and B showing disc edema in the right and left eyes respectively, with retinal haemorrhages. C and D showing resolution of disc edema, after treatment with thiamine in the right and left eyes respectively with disc pallor.

Figure 2: A showing hyperintensities on Flair images in bilateral medial thalami. B showing hyperintensities on Flair images in periaqueductal grey matter. C showing hyperintensities in bilateral mamillary bodies.

Figure 3: A and B represent Visual fields of right and left eyes respectively with severe field constriction.

C and D represent Visual fields of right and left eyes respectively, a month after treatment with centrocecal scotomas.



