

CASE REPORT

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

KEYWORDS

hyperemesis gravidarum, ophthalmoparesis, papilloedema, retinal hemorrhages, thiamine

1 | 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.¹ Operational 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 hemorrhages with visual loss.⁴ Papilloedema 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.

2 | CASE REPORT

A 22-year old lady presented with a history of blurring of vision since 6 days. The visual blurring was acute at onset and became 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

All authors were involved in the clinical care of the patient and in the preparation and revision of the manuscript.

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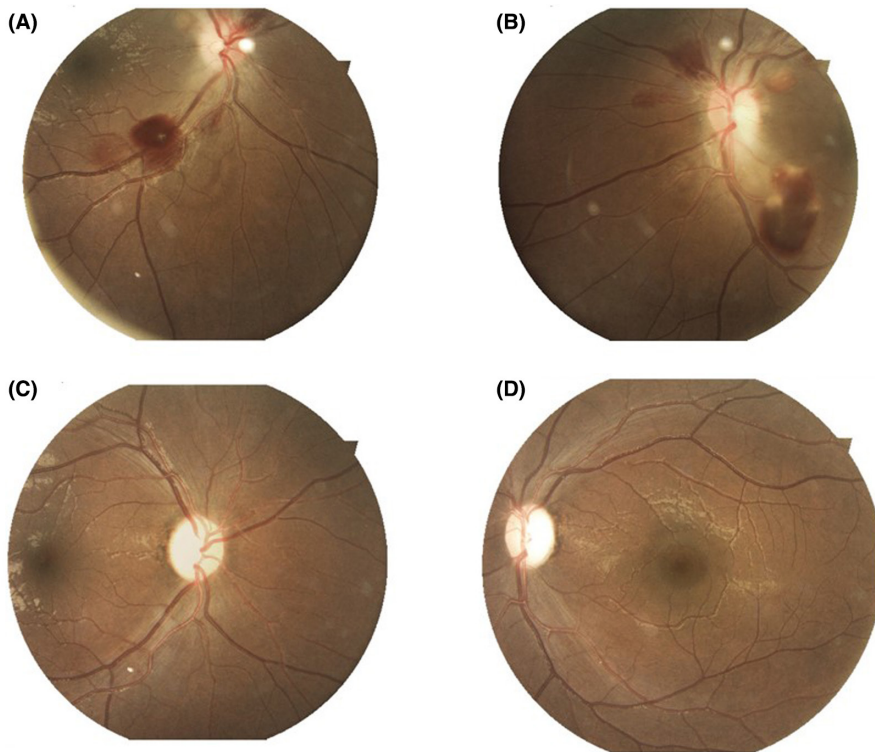


FIGURE 1 (A) and (B) show disc edema in the right and left eyes respectively, with retinal hemorrhages. (C) and (D) show resolution of disc edema, after treatment with thiamine in the right and left eyes respectively with disc pallor.

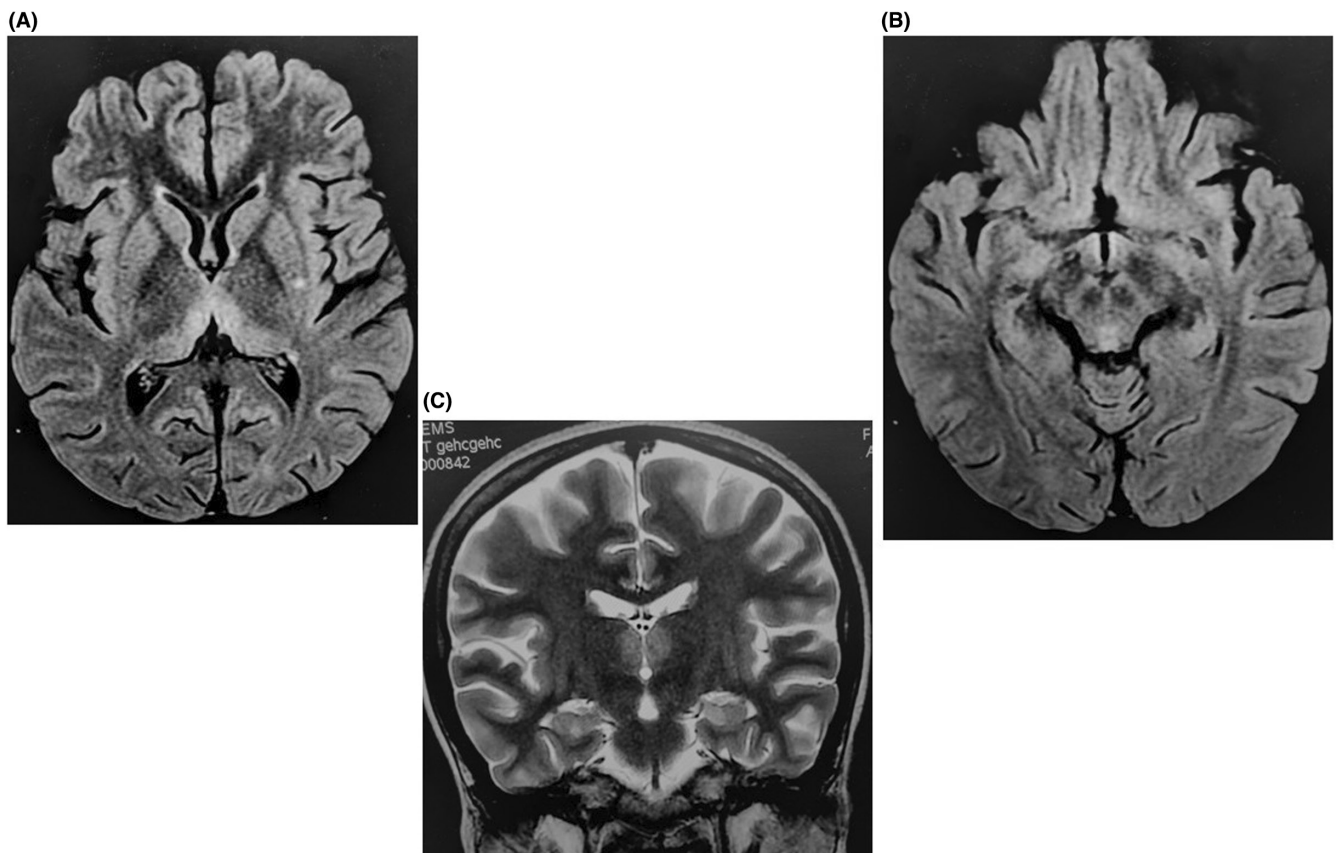


FIGURE 2 (A) showing hyperintensities on FLAIR images in bilateral medial thalami. (B) showing hyperintensities on FLAIR images in periaqueductal gray matter. (C) showing hyperintensities in bilateral mammillary bodies.

ataxia. Further, there were bilateral cerebellar signs on examination.

Funduscopy revealed bilateral disc edema with retinal hemorrhages (Figure 1A,B). Her blood investigations revealed a sodium level of 128 meq/l, and a

potassium level of 2.9 meq/l, and she tested positive for urine ketone bodies. An echocardiogram was performed, which revealed normal cardiac function. A nerve conduction study of all four limbs was done, which was normal.

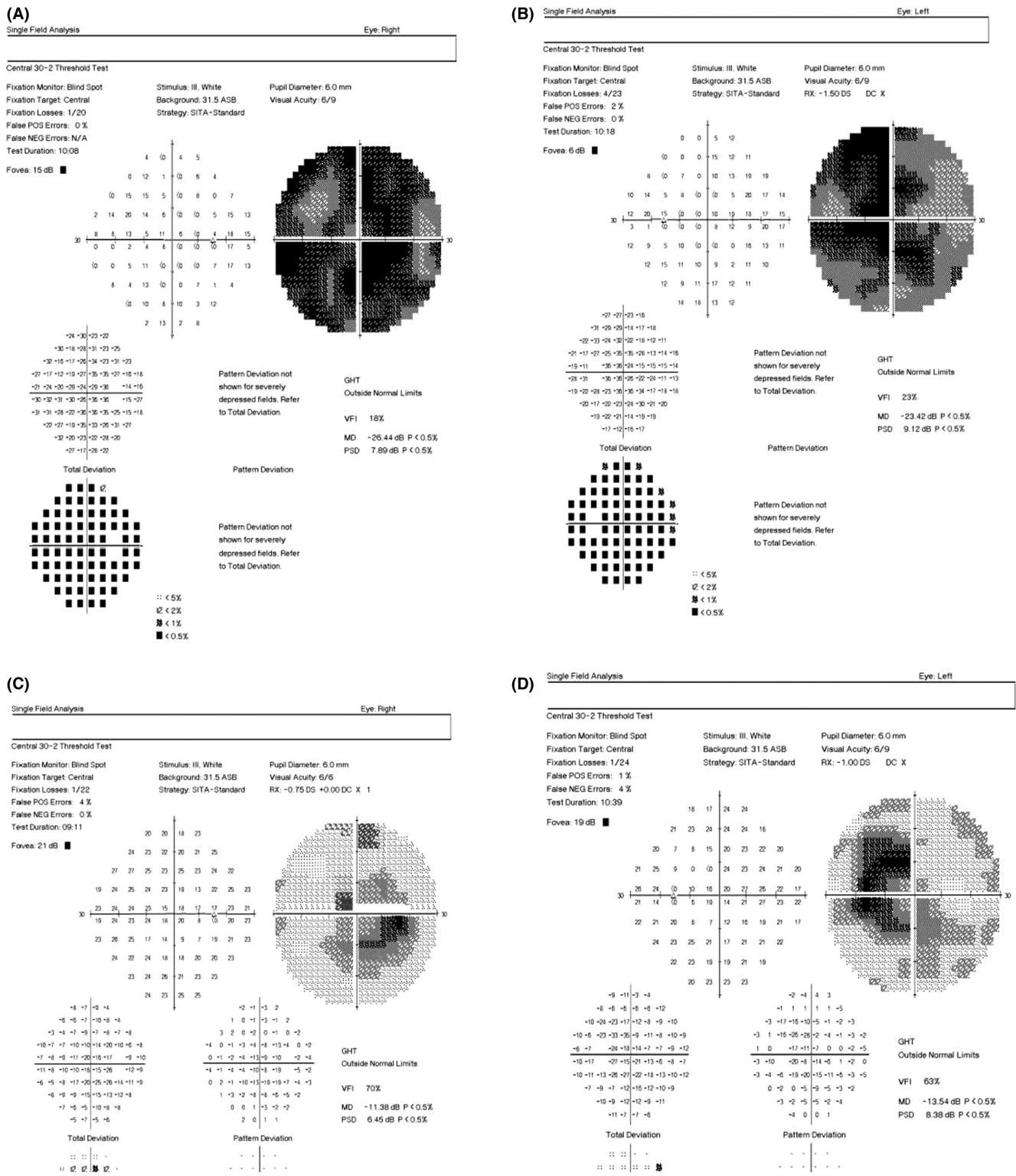


FIGURE 3 (A) and (B) represent visual fields of right and left eyes respectively with severe field constriction. (C) and (D) represent visual fields of right and left eyes respectively, a month after treatment with centrocecal scotomas.

MRI brain scan was done and the results showed Fluid attenuated inversion recovery (FLAIR) hyperintensities in bilateral medial thalami, periaqueductal gray matter, and mamillary bodies (Figure 2). Visual fields showed severe constriction bilaterally (Figure 3A,B: Right and left eyes, respectively). Thus, a diagnosis of Wernicke's encephalopathy was made.

The patient was treated with thiamine parenterally, with 500 mg iv on the first day, followed by 100 mg intravenously thrice a day for 7 days. Her vision improved dramatically with the 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 1C,D) and visual fields improved too (Figure 3C,D, Right and Left eyes respectively).

3 | 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 hemorrhages, ptosis, and disc edema.⁵

Disc edema is a rare finding in Wernicke's encephalopathy.⁶ Retinal hemorrhages are seen more frequently, without 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

Chitra et al.⁹ described a 25-year old lady in her 20th week of pregnancy, who presented with visual loss, disc edema, retinal hemorrhages, ataxia, and ophthalmoparesis. The clinical features improved rapidly with parenteral thiamine.

Thiamine prophylaxis in patients with hyperemesis gravidarum has also been recommended, starting from 50 mg per day to 200 to 1200 mg per day parenterally for 2 weeks followed by oral thiamine of 60 mg per day till the end of gestation.¹⁰

The initial treatment dose of 500 mg thiamine three times daily has been recommended by the Royal college of physicians and European federation of neurological societies, which is believed to result in a quick resolution of neurological symptoms.¹¹

Visual loss as a presenting clinical feature of Wernicke's encephalopathy is very rare and must be considered in an appropriate clinical setting, in order to facilitate early treatment.

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

4 | 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 with hyperemesis gravidarum, where there is an enhanced metabolic requirement of thiamine, one must have a high index of suspicion in patients presenting primarily with visual loss and other features of Wernicke's encephalopathy. This is especially important as disc edema and retinal hemorrhages are rare in Wernicke's encephalopathy. This would prevent a potential delay in diagnosis and facilitate urgent treatment with parenteral thiamine. Thus, visual loss may be reversible in Wernicke's encephalopathy, if treated early with 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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CONFLICT OF INTEREST

The authors have no conflict of interest to disclose.

DATA AVAILABILITY STATEMENT

Data available on request from the authors

CONSENT

A written consent has been obtained from the patient. It can be provided if requested.

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