CASE REPORT

Reversible severe bilateral visual loss in an unusual case of bilateral lateral geniculate myelinolysis during acute pancreatitis

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SUMMARY

A 40-year-old man was diagnosed with pancreatitis following cholecystectomy. During hospitalisation, he reported bilateral acute vision loss. His best-corrected visual acuity (BCVA) was counting fingers in the right eye and 20/200 in the left eye. Ocular fundus examination and optical coherence tomography revealed a slight alteration in the retinal nerve fibres in the nasal macular region. Automated perimetry revealed bilateral visual field defects affecting both temporal and nasal hemifields in a predominantly nasal distribution, and brain MRI confirmed symmetrical lesions within both lateral geniculate nuclei. BCVA was gradually recovered, reaching 20/20 within 6 weeks.

BACKGROUND

Central pontine myelinolysis (CPM) is a demyelinating disease, usually associated with alcoholism, rapid correction of hypernatraemia or hyponatraemia and other electrolyte disturbances. Extrapontine demyelination of both lateral geniculate nuclei (LGN) is an uncommon cause of acute bilateral vision loss, and might cause retrograde retinal alterations. Although MRI is the optimal imaging technique to confirm the diagnosis, optical coherence tomography (OCT) and visual field examination are able to detect subtle changes.

To the best of our knowledge, this is the first case reported where bilateral visual field defects in a predominantly nasal distribution from intrageniculate myelinolysis were induced by acute pancreatitis without fluid and electrolyte imbalance.

CASE PRESENTATION

A 40-year-old man presented to the emergency department reporting abdominal pain and vomiting, several days after being discharged from hospital after a laparoscopic cholecystectomy. He had no history of drug or alcohol abuse. Physical examination revealed a soft abdomen, depressible and painful to palpation at the right upper quadrant, with no signs of peritonism. Abdominal ultrasonography showed fluid collection in the gallbladder bed, suggestive of abscess in the surgical site. The patient's chemistry profile revealed elevated values of bilirubin (3.4 mg/dL) aspartate aminotransferase 556 U/L, alanine transaminase 636 U/L and γ glutamyl transferase 660 U/L, amylase of 4052 IU/L and blood count showed leucocytosis ($11.34 \times 10^3/\mu\text{L}$). Ion levels were within normal limits. The patient was admitted to the surgical service with a

diagnosis of acute pancreatitis. Blood pressure figures, heart rate, temperature and oxygen saturation were within normal parameters during his hospital stay. In successive analyses, the lowest sodium level detected was 135 mEq/L (normal range: 136–145 mEq/L) and the patient suffered no diminution of consciousness, had no language disturbance and had no difficulty walking.

The patient received antibiotic treatment and parenteral nutrition, as well as therapy to replace fluid losses, and to maintain volume and electrolyte balance; subsequently, the digestive symptoms improved. Nevertheless, during the third day of hospitalisation, he reported severe acute vision loss in both eyes. Neurological examination showed normality in meningeal exploration and in cranial pairs, except for visual acuity and perimetry, as mentioned below; there was no impairment in strength, sensitivity or coordination. Tendon reflexes were also normal. Complete neuro-ophthalmological examinations were performed for visual acuity, in addition to binocular slit-lamp examination, colour vision, pupillary reflexes and funduscopy.

The patient's best-corrected visual acuity (BCVA) was counting fingers in the right eye (RE) and 20/200 in the left eye (LE). Colour vision (Ishihara testing) was abnormal. Pupillary reflexes were normal. Biomicroscopy of the anterior pole was normal and intraocular pressure was within the normal range. Fundus examination revealed no pathological findings, except for a slight change in colour and brightness of the nasal macular region. OCT also showed slightly increased macular thickness in the nasal region of both eyes (320 μm in RE and 350 μm in LE) as well as altered reflectivity of the retinal layers (figure 1). The low visual acuity of the patient prevented the successful completion of the visual field, which also could not be assessed due to the low reliability of the test. However, a second automated perimetry test revealed bilateral visual field defects affecting both temporal and nasal hemifields in a predominantly nasal distribution 7 days after the patient's vision had become affected (figure 2), and subsequent brain MRI showed hyperintense symmetrical lesions confined within both LGN on fluid-attenuated inversion recovery (FLAIR) images and on T2-weighted images. Coronal T1WI after gadolinium (Gd) administration showed clearly visible enhancement within both LGN (figure 3A, B).

Autoimmunity, hypercoagulability and serologic tests were all negative. There were no pathological findings in the intracranial MR angiography,

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Figure 1 Optical coherence tomography showing a slight increase in nasal macular thickness in both eyes as well as alteration in the reflectivity of the nasal retinal layers.

transthoracic echocardiography and echocardiography of the supra-aortic trunks.

OUTCOME AND FOLLOW-UP

Colour vision slowly returned to normal and BCVA gradually recovered to 20/200 in the RE and 20/100 in the LE 7 days

after symptom onset, reaching 20/20 within 6 weeks. OCT image showed a decrease in nasal macular thickness in both eyes (292 μm in RE and 319 μm in LE) and a recovery of morphology in retinal layers compared to the first scan. Automated visual fields were improved, but bilateral nasal defects remained.

Figure 2 Automated visual fields showing bilateral incongruous nasal field defects.

Figure 3 (A) Axial T2 MRI showing symmetric hyperintense lesions affecting both lateral geniculate nuclei (LGN). (B) Coronal T1WI after gadolinium (Gd) administration. Enhancement within both LGN is clearly visible. (C) Control axial T2 MRI, where lesions are still visible, but where the extent and intensity of the abnormal high-signal intensity are decreased. (D) Coronal Gd-enhanced T1WI, in which contrast uptake is no longer evident.

Follow-up MRI studies were performed 6 months after visual loss. On axial and coronal T2 images, the lesions were almost unnoticeable, the extent and intensity of the abnormal signal intensity were greatly decreased and Gd-enhanced T1WI sequences returned to normal (figure 3C, D).

DISCUSSION

CPM was originally described by Adams *et al* in 1959.¹ The clinical syndrome has been described in numerous subsequent case reports as a rapidly evolving flaccid quadriplegia, with facial and lingual weakness and difficulty speaking or swallowing. CPM is a demyelinating disease of the central nervous system that is associated with a distinctive neuropathological finding in the central part of the pons; however, it may appear in extrapontine locations, even when the pons is normal.² Three patterns of demyelinated lesions were defined by Gocht *et al*³ after observing 58 cases with CPM and/or extrapontine myelinolysis (EPM). Tomlinson *et al*⁴ were the first to make the link between CPM and severe hyponatraemia. Afterwards, it was observed that the risk of CPM seemed greater when the hyponatraemia was corrected too quickly.⁵ The clinical consequences, ranging from asymptomatic CPM to coma, do not necessarily depend on the severity of degree of hyponatraemia, in fact, the disorder may occur with normal serum sodium levels.

Conditions associated with an increased risk of central pontine and EPM are alcoholism, chronic nutritional deficiency and many other systemic diseases with electrolyte abnormalities, including chronic lung disease, kidney and liver disease, diabetes mellitus, sepsis, liver transplantation and neoplasms.^{3 6}

Lateral geniculate myelinolysis is a unique pattern of CPM, reported in 1981 by Goldman and Horoupian.⁷ Cerebral MRI has been the optimal imaging technique to confirm its diagnosis since 1995, when Donahue *et al*⁸ reported neuroimaging and visual field findings.

We present a patient with a bilateral lesion in his LGN confirmed by MRI, which resulted in bilateral central visual loss affecting both temporal and nasal hemifields in a predominantly nasal distribution. The fact that this patient was being treated for pancreatitis, with the need for parenteral nutrition and fluid, suggests that this could have been the root cause. Osmotic demyelination should be suspected in patients receiving intravenous fluids during the management of underlying diseases; pancreatitis is one of the diseases associated with increased risk of CPM and EPM.⁶

Our patient suffered bilateral lateral geniculate demyelination even when serum sodium levels showed no significant variations. No hyponatraemia and no hypernatraemia were identified in any of his blood tests, and the patient did not present clinical manifestations of these disturbances. As already documented,

EPM may occur when the serum sodium level is normal⁹ or elevated,¹⁰ related in the first case, with the delayed reaccumulation of glycerophosphorylcholine in brain cells.

The onset of symptoms in CMP and EPM is typically delayed from 2 to 7 days, as was seen in our patient.⁶

The LGN is the major relay station between the retina and visual cortex, since the vast majority of retinal ganglion cell axons terminate in them. Nevertheless, damage to both LGN is an uncommon cause of acute bilateral visual loss, hence neuroimaging studies of the LGN in the context of vision loss are rare. In our case, the imaging techniques had to overcome some anatomical challenges owing to the small size of the LGN and their proximity to other adjacent grey matter thalamic nuclei. Nowadays, MRI is an indispensable method for visualising thalamic nuclei such as LGN, and it is the optimal imaging technique to diagnose osmotic demyelination lesions and to perform subsequent follow-up.¹¹

Regarding MRI, acute demyelinating lesions usually appear symmetrical and hypointense on T1-weighted images, become hyperintense on T2 and on FLAIR images in the subacute stage, and intense enhancement images are observed on T1WI after Gd administration during the first week from the onset of symptoms, as we saw in our patient. Similar pathological T2 images and Gd sequences and their improvement in subsequent control MRI examinations have been described previously.^{8 11–15}

Bilateral lateral geniculate myelinolysis causes bilateral painless visual loss with unusual visual field patterns. The first bilateral visual field defects reported in a patient with presumed lateral geniculate myelinolysis were described by Donahue *et al*,⁸ these resembled an hourglass, with near total depression superior and inferior to fixation. Barton¹² also reported a similar case, finding bilateral congruous sectoranopias in keeping with the patterns of bilateral LGN damage described before. However, the visual fields from a case of possible lateral geniculitis, reported by Greenfield *et al*,¹³ showed left homonymous wedge-shaped defects and an incongruous right hemianopia described as hourglass-shaped field preservation, the opposite to the previous patient's defects. Likewise, Baker *et al*¹⁴ described incongruous homonymous bilateral visual field defects in a bow-tie configuration, in isolated bilateral acute infarctions of the lateral geniculate bodies.

Imes *et al*¹⁵ reported almost pure bilateral nasal hemianopias from presumed intrageniculate myelinolysis after emergency caesarean section and hysterectomy. Their findings on MRI and visual field were similar to ours.

They provided a logical explanation for hemianopic nasal field defects in intrageniculate myelinolysis, based on the uncrossed retinal projections (nasal hemifields) reaching the geniculate layers in the central portion of the LGN. However, lateral geniculate demyelinating lesions, due to their anatomical peculiarities, could produce very different visual field defects, depending on the affected area.

Therefore, if field defects are detected after a vision loss and not attributable to ophthalmological pathology such as glaucoma or optic neuritis, and once ischaemic and haemorrhagic strokes have been ruled out, we should consider involvement of the LGN.

The contribution of OCT in the diagnosis of visual loss arising from LGN is not yet known. This is the first description of an altered level of the retinal layers, possibly due to retinal oedema favoured by retrograde source from lesions of the LGN. In our patient, spontaneous OCT recovery coincided with the recovery of the patient's visual acuity. We believe that using OCT, a rapid and non-invasive imaging technique employed in

ophthalmology, could be useful in future cases of osmotic geniculate myelinolysis.

To the best of our knowledge, this is the first case report of bilateral central visual loss affecting both temporal and nasal hemifields in a predominantly nasal distribution, due to intrageniculate myelinolysis induced by acute pancreatitis without fluid and electrolyte imbalance, describing findings in both, magnetic resonance imaging (MRI) and OCT imaging techniques. However, without neuropathological confirmation, we can only speculate about the cause of these LGN lesions and visual loss, although the clinical evolution ruled out septic shock as a possible cause, as well as ruling out infectious disease and ischaemia.

Learning points

- Damage of both lateral geniculate nuclei is an uncommon cause of acute bilateral visual loss.
- Osmotic demyelination should be suspected in patients receiving intravenous fluids during the management of underlying diseases.
- Bilateral lateral geniculate myelinolysis causes bilateral painless visual loss with unusual visual field patterns.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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