Unusual association of diseases/symptoms

A rare potentially treatable cause of bilateral optic disc swelling

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Summary

Rapid onset bilateral optic disc swelling generally indicates an intracranial problem—that is, papilloedema. However, when there is also visual loss, disease affecting the optic nerves themselves must be considered.

We present the diagnostic problem of a patient with optic disc swelling and progressive visual loss.

Investigations finally revealed hypocalcaemia secondary to primary hypoparathyroidism. With treatment a marked improvement in vision occurred.

This reversible rare cause of optic disc swelling should not be forgotten.

BACKGROUND

It is very rare for a patient to present to an eye department with symptoms due to hypocalcaemia. Consequently, this patient had many investigative tests and, in addition, received high-dose methyl prednisolone before the correct diagnosis was made.

Raising awareness of sight threatening complications of hypocalcaemia might enable future clinicians to treat earlier and perhaps avoid some costly investigations.

CASE PRESENTATION

A 62-year-old Caucasian man presented to the eye department with blurred vision in his left eye, which he had noticed 5 days previously.

There was no history of headache, eye pain or diplopia and no other new neurological symptoms.

There was a complicated past medical history. He had had mastoid surgery many years before and was deaf in the left ear. Eleven years previously he had developed staph-

Figure 1 Visual fields at presentation.

Figure 2 Optic disc appearances at presentation.

Figure 3 ECG prior to treatment.

lyococcal septicaemia and meningitis complicated by the development of extradural infection involving the caudal equina. His recovery was incomplete and resulted in weakness of both legs and limited bladder and bowel control.

Current medication was omeprazole and quinine for cramps. He was an ex-smoker for the last 12 years and did not drink alcohol. There was no family history of eye diseases.

On examination the patient appeared generally well. He was afebrile and normotensive. Visual acuities with correction were 6/12 right and 6/60 left. Pupils were equal but there was a marked left relative afferent pupil defect. Eye movements were full with no ptosis.

Colour vision testing by Ishihara method showed impairment mild on the right and severe on the left. Automated perimetry showed a superior nasal arcuate field loss on the right and gross generalised field loss on the left (figure 1).

Fundus examination revealed bilateral disc swelling with absent spontaneous venous pulsation (figure 2). The disc swelling was more marked on the left side. In both eyes the rest of the retinal examination was normal.

Temporal arteries were palpable and non-tender. Chest and abdominal examination was unremarkable. General neurological examination revealed no new neurological signs just the longstanding mild weakness of both legs and left-sided deafness.

The patient was admitted under the physicians and vision progressively deteriorated for the first week to only hand movement vision on the left and maintained acuity but loss of upper hemi field on the right.

INVESTIGATIONS

Full blood count, clotting function, B₁₂ and folate were all normal. Erythrocyte sedimentation rate was 8 mm/h, C reactive protein 1 mg/dl, cholesterol 5.7 mmol/l, triglycerides 2.9 mmol/l and low-density lipoprotein 3.4 mmol/l. Thyroid function was normal as was urea, electrolytes and glucose. Auto antibodies were not raised and paraneoplastic antibodies were not present.

Chest x-ray showed only a small focus of right-sided atelectasis. MRI of the brain with venogram showed no abnormality and abdominal ultrasound was normal.

Figure 4 Post-treatment visual fields.

Figure 5 Post-treatment optic discs.

Cerebrospinal fluid opening pressure was normal. There was a slight increase in protein level (0.45 g/l) but no bacterial growth after 48 h culture. Serological testing for syphilis, Lyme disease and HIV was negative.

Bone profile showed a corrected calcium of 0.95 mmol/l, phosphate of 2.28 mmol/l and alkaline phosphatase of 88 IU/l. Twenty-four hour urinary calcium was 0.6 mmol with a normal range of 2.5–7.5. Parathormone level was 0.5 pmol/l (normal range is 1.6–6.9) and synacthen test was normal.

ECG showed a prolonged corrected QT interval at 0.532 s (figure 3). Electro-diagnostic tests revealed a normal electroretinogram but markedly reduced and delayed responses in the visual evoked potential recordings for the left eye.

DIFFERENTIAL DIAGNOSIS

Bilateral anterior ischaemic optic neuropathy; intracranial space occupying lesion; benign intracranial hypertension; toxic, inflammatory compressive or infective optic neuropathy; Devic's disease; Leber's hereditary optic neuropathy.

TREATMENT

The patient's vision deteriorated rapidly during the week he was admitted with progressive loss of acuity and field in his better right eye.

All initial investigations failed to provide the diagnosis and, therefore, a course of high-dose intravenous methyl prednisolone 1 g daily was given for 5 days to treat a

possible inflammatory, demyelinating or vasculitic process that might be affecting the optic nerves.

Considering underlying malignant disease a bone profile was requested, which surprisingly revealed extreme hypocalcaemia (corrected calcium 0.95 mmol/l). After confirming this unexpected result intravenous calcium gluconate was given. Primary hypoparathyroidism was diagnosed and so subsequent oral treatment has been with alphacalcidol and sandocal.

OUTCOME AND FOLLOW-UP

A gradual improvement in vision has occurred in this patient with normalisation of his serum calcium. At last review, visual acuities were 6/24 left and 6/4 right. The left field of vision is markedly improved and the right now normal (figure 4). The right optic disc is healthy and the left appears pale but no longer swollen (figure 5). The patient feels generally much better and mentally more alert.

Discussion Hypocalcaemia as a cause of papilloedema was first described by Luttwig in 1903.¹ It is listed in many short notes style medical textbooks but because it is extremely rare the diagnosis was not considered here and discovered fortuitously.

Ayuk *et al*² described a patient whose carpopedal spasm was attributed to panic attacks for 2 years until visual loss occurred and hypocalcaemia was finally diagnosed.

A number of authors have described complications of hypocalcaemia but generally in patients where there is a history of other endocrine problems and thyroid or parathyroid surgery.

Our patient did not offer any pointers to hypocalcaemia but on retrospective questioning he had noticed a spasm developing in his fingers during sphygmomanometry.

The basis of optic disc swelling in hypocalcaemia is uncertain. In some patients there is raised intracranial pressure even up to 46 cm of water.³ Raised intracranial pressure may be due to reduced cerebrospinal fluid absorption⁴ or hyper secretion. In other situations, as here, the cerebrospinal fluid pressure was normal.

Calcium affects nerve conduction. In particular, fast axonal transport is affected by calcium. A secondary effect

of this may be to cause axonal and optic nerve swelling. This could in turn result in ischaemic optic neuropathy in those with vascular risk factors as postulated by Maclean *et al*⁷ in 1998.

In our patient, correction of serum calcium has produced gradual improvement in vision but the left optic disc is atrophic. A secondary ischaemic effect may be responsible.

Learning points

- ▶ Hypocalcaemia is a very rare cause of optic disc swelling.
- ▶ Disc swelling may be true papilloedema with raised intracranial pressure or an optic neuropathy with visual loss as in this case.
- ▶ Checking the patient's serum calcium at an earlier stage in his work-up would possibly have avoided more costly and invasive investigations.
- ▶ Treatment of hypocalcaemia can reverse progressive visual loss.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. Luttwig G. Ein Fall von chronischer tetanie. Inaugural Dissertation, Munich, 1903.
2. Ayuk J, Matthews T, Tayebjee M, *et al*. A blind panic. *Lancet* 2001;**357**:1262.
3. Grant DK. Papilloedema and fits in hypoparathyroidism, with a report of three cases. *Q J Med* 1953;**22**:243–59.
4. Sambrook MA, Hill LF. Cerebrospinal fluid absorption in primary hypoparathyroidism. *J Neurol Neurosurg Psychiatr* 1977;**40**:1015–17.
5. Nathanson JA. Beta-adrenergic sensitive adenylate cyclase in choroid plexus: properties and cellular localisation *Mol Pharmacol* 1980;**18**:199–209.
6. Breuer AC, Atkinson MB. Calcium dependent modulation of fast axonal transport. *Cell Calcium* 1988;**9**:293–301.
7. McLean C, Lobo R, Brazier D. Optic disc involvement in hypocalcaemia with hypoparathyroidism: papilloedema or optic neuropathy? *Neuro-ophthalmology* 1998;**20**:117–124.

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