eSELF ASSESSMENT QUESTIONS

Visual disturbance in a patient with progressive limb weakness

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A 38 year old man presented in with a three month history of progressive weakness of all four limbs. He also had numbness and tingling sensation in his arms and legs for the same period of time. The weakness in his leg had progressed to an extent that he was unable to walk unsupported and recently had a few falls. Over the last previous months he had shown deterioration in visual acuity especially of the left eye. There was no significant past medical history. He was a non-smoker and denied taking alcohol in excess.

Power was grade 3–4 in all four limbs. The tendon reflexes were absent and the plantar responses were down going. There was diminution in all modalities of sensation in both upper and lower limbs in a glove and stocking distribution. His retinal photograph is shown in fig 1. Rest of the cranial nerves were normal.

Investigations

Full blood count, urea and electrolytes, glucose, serum vitamin B12, thyroid and liver function tests were normal. Autoantibodies, syphilis serology, and viral titres were negative. Tests for HIV was also negative. Plasma protein electrophoresis was normal. Nerve conduction studies revealed severe delay in sensory and motor nerve conduction velocities. Computed tomography and magnetic resonance imaging of the brain were normal. After excluding a space occupying lesion, lumbar puncture was performed. The albumin content of the

Figure 1 Retinal photograph of left eye showing papilloedema. There is severe swelling of the optic disc associated with dilated retinal veins and peripapillary haemorrhages.

cerebrospinal fluid (CSF) was 4.5 g/l (normal range 0.15–0.40 g/l) with polyclonal hypergammaglobulinaemia. CSF cell counts were normal. The CSF pressure was 40 cm (normal range 5–15 cm).

Questions

- (1) What does the retinal photograph show?
- (2) What is the complete diagnosis?
- (3) What is the management of this condition?

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Answers

QUESTION 1

The retinal photograph of the left eye shows florid papilloedema with disc swelling, dilated veins, and peripapillary haemorrhages.

QUESTION 2

The complete diagnosis is chronic inflammatory demyelinating polyneuropathy (CIDP) causing benign intracranial hypertension.

QUESTION 3

High dose corticosteroids, plasmapheresis, and intravenous immunoglobulin are beneficial in CIDP. Immunosuppressive agents (azathioprine and cyclosporin) have also been used with variable success. For benign intracranial hypertension repeated lumbar puncture is usually effective. High dose steroids and acetazolamide have also been used with variable success. If these measures fail and there is a threat to vision then lumbthecoperitoneal shunt may be required. Occlusion of the shunt due to raised CSF protein is a potential complication and a barrier to early neurosurgical intervention.

Discussion

CIDP is characterised by progressive weakness of the limbs over several weeks or months. It has several different presentations such as pure motor syndrome, sensory ataxic variant, mononeuritis multiplex pattern, relapsing type, and paraparetic subtype.¹ Patients typically present with progressive stepwise or relapsing weakness.² To fulfil the diagnostic criteria for CIDP, weakness must be progressive for at least two months, which distinguishes it from Guillain-Barré syndrome. Weakness can vary in severity but is generally symmetric and involves proximal and distal muscles of upper and lower extremities. Sensory complaints usually consist of numbness and tingling but painful paraesthesia is not uncommon. Autonomic and respiratory insufficiency occurs less frequently than Guillain-Barré syndrome. The precise aetiopathogenesis of CIDP is not clear but autoimmune mechanisms involving autoantibodies against myelin proteins and glycolipids have been implicated.³

The three laboratory studies for support of diagnosis of CIDP are the CSF examination, electrophysiological studies, and nerve biopsy. CSF shows albuminocytological dissociation. Of the three laboratory studies, the CSF may be the most useful, as 94% of patients have raised CSF protein.⁴ Nerve conduction studies in CIDP are suggestive of demyelination, with slowed nerve conduction velocities, prolonged or absent F-waves, conduction blocks, and temporal dispersion. Nerve biopsy shows interstitial and perivascular inflammatory infiltration causing segmental demyelination. Demyelination is affected by T-cells and macrophages within the endoneurium. Myelinated fibres are lost to a certain degree and many of the remaining ones are seen to be undergoing demyelination-remyelination. Onion bulb formations are conspicuous in the recurrent and relapsing cases. Nerve biopsy, however, is the least useful in supporting the diagnosis of

CIDP.⁴ In clinical practice it is rarely undertaken and is more of a research procedure.

Randomised control trials have shown that corticosteroids, plasmapheresis, and intravenous immunoglobulin are beneficial in CIDP.⁴ High dose prednisolone is effective in the treatment of CIDP. The dose is 100 mg daily for several weeks. When the improvement plateaus the dose can be slowly reduced by 5 mg every 2–3 weeks.

Recent trials have shown that intravenous immunoglobulin has a role in treatment of CIDP.⁵ It has now superseded corticosteroids as the treatment of choice. The dose is 2 g/kg over five days. It works by modulation of Fc receptors or T-cell function. Response may be temporary and the treatment might have to be repeated.

Plasmapheresis also has beneficial effect on both neurological disability and nerve conduction. Intermittent plasmapheresis can reduce the use of steroid and allow it to be discontinued.⁶ Immunosuppressive agents (azathioprine and cyclosporin) have been used with variable success.

CIDP can be associated with benign intracranial hypertension. The precise mechanism is not clear but raised CSF protein causing impaired reabsorption may be one possible explanation. Repeated lumbar puncture can help reduce the CSF pressure. Drug treatment consists of high dose steroids and acetazolamide. None of them are consistently effective.

Regular visual acuity and field testing is important in detecting early visual affection. If the conservative measures are unsuccessful then shunt procedures (lumbthecoperitoneal) may be required. However there is a high risk of shunt occlusion due to high protein content of the CSF.

Our patient was treated with intravenous immunoglobulin and high dose corticosteroids and over ensuing months recovered remarkably from polyneuropathy. However despite repeated therapeutic drainage of CSF the pressure remains high. He has been referred to neurosurgeons for further management to prevent visual deterioration.

Final diagnosis

Chronic inflammatory demyelinating polyneuropathy causing benign intracranial hypertension.

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