Bilateral optic atrophy with hydrocephalus

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A 25-year-old woman was referred to us by an ophthalmologist to whom she had complained of gradual bilateral loss of vision over a period of several weeks, which had progressed to a near total blindness during the preceding two weeks. She did not complain of any other symptoms suggestive of raised intracranial tension. There was no history of head injury, cardiac disease, tuberculosis, sexually transmitted diseases or exposure to toxic substances. There was no consanguinity. Developmental milestones and intelligence were normal.

The clinical examination was essentially normal except for short stature (height 146 cm, head circumference 56 cm). Ophthalmological examination revealed bilateral symmetrically semidilated pupils, which did not react to light. The eye movements and tension were normal. Funduscopy revealed bilateral optic atrophy (figure 1). The rest of the central nervous system and other systemic examination was normal. Preliminary haematological, biochemical, immunological, and radiological investigations did not show any abnormality. The electroencephalogram revealed diffuse cortical dysfunction suggestive of raised intracranial tension. Computed tomography (CT) of the brain showed gross dilatation of the entire ventricular system with a markedly thinned-out cerebral cortex (figures 2 and 3).

Figure 1 Funduscopy

Figure 3 Brain CT scan

Figure 2 Brain CT scan

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Questions

- 1 What are the causes of bilateral optic atrophy in adults?
- 2 What is the diagnosis in this case?

Answers

QUESTION 1

Optic atrophy is the term usually applied to the condition of the disc when the end point of the optic nerve is degenerated. Injury to the nerve fibres along any part of their course from the retina to the lateral geniculate body, leads to their degeneration and atrophy. When optic atrophy occurs without local disturbances, unassociated with generalised disease of the central nervous system, and without any discoverable cause, the condition is described as primary optic atrophy. The causes of secondary optic atrophy are given in the box. It may often not be possible to determine whether optic atrophy was primary or secondary in a particular patient, especially when there is a delay in presentation.

QUESTION 2

The diagnosis is Dandy Walker complex, characterised by cystic dilation of the fourth ventricle and an enlarged posterior fossa with upward displacement of lateral sinuses, tentorium and torcular herophili associated with varying degrees of vermian aplasia or hypoplasia. It may be caused by failure of development of the anterior medullary vellum (embryonic roof of the 4th ventricle), atresia of the 4th ventricular outlet foramina, or delayed opening of the foramen of Magendie.

The diagnosis was made in this patient on the basis of the CT scan, which revealed a grossly dilated ventricular system with severe cerebral atrophy and the presence of a posterior fossa cyst. Bilateral optic atrophy could be explained on the basis of chronic

Causes of secondary optic atrophy

- extensive disease of the retina, eg, pigmentary retinal dystrophy, occlusion of the central retinal artery or thrombosis of the vein
- a break in the fibre continuity as a result of their strangulation, eg, papillitis, neuroretinitis, multiple sclerosis
- papilloedema (choked disc) due to elevated CSF pressure producing axoplasmic stasis as a result of raised intracranial tension (due to infections such as meningitis or encephalitis, cerebrovascular accidents, tumours, head injuries, or CSF circulation blocks), papillitis and pseudopapillitis, intracranial tumours, oedema due to uveitis, glaucomatous atrophy, or toxic atrophy (eg, exposure to methanol, heavy metals, or drugs)

raised intracranial tension. In view of the relative short history of a few weeks of visual deterioration, a neurosurgeon's opinion was solicited and the patient underwent a ventriculoperitoneal shunt operation. There was, however, no improvement in her vision when she was last evaluated, six months after surgery. This is the first documented case of Dandy Walker complex from India.

Final diagnosis

Dandy Walker complex with bilateral optic atrophy.

Keywords: Dandy Walker complex; bilateral optic atrophy; blindness

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