Letters

reduced pupillary response to light in the right eye, as well as a pale optic disc in the right fundus. The visual field testing of the right eye demonstrated baring of the blind spot of Mariotte. Visual evoked response testing revealed the slight elongation of P100 latency. Fluorescein angiography revealed late staining of the right disc without other specific abnormalities. Exophthalmometric readings were 24 mm in the right eye and 20 mm in the left eye with a base of 111 mm. The orbital CT scan revealed severe hypertrophy of the extraocular muscles and orbital fat. Laboratory results showed an increase in thyrotropin binding inhibiting immunoglobulin (TB II) (23-7%) and triglyceride (188 mg/dl). Serum and cerebrospinal fluid tests for syphilis were negative. The antinuclear antibodies, anti-Ro (SS-A) antibodies, anti-La (SS-B) antibodies, rheumatoid factor, antinuclear antibodies were negative. After testing the patient was diagnosed as having endocrine ophthalmopathy. Radiation (20 Gy) was administered and the steroid therapy was continued. Although TBI became normal (5-9%), there was no beneficial effect. Sixteen months after the first attack, the colour of the optic disc was completely pale and visual acuity was zero in the right eye.

Six months later, the patient complained of a decrease in visual acuity in the left eye. Examination revealed the best corrected visual acuity to be 20/50 in the left eye. The visual field testing of the left eye demonstrated central scotoma within 10 degrees. A CT scan disclosed a thickening and enhancement of the dura mater, extending to the cavernous sinuses including the area of the optic foramina which appeared to be constricting the optic nerves. Severe hypertrophy of the extraocular muscles was unchanged (Fig 1). The chest radiograph was normal. Laboratory results showed an increase in C reactive protein (8.4 mg/dl); however, serum angiotensin II converting enzyme level was normal. An open biopsy revealed marked thickening of the temporal dura mater due to severe chronic inflammation, as well as fibrosis with small necrotic foci and perivasculitis with early granulomatous reaction. The reactive cells were mainly lymphocytes with some plasma cells and histiocytes. No evidence of tuberculosis or fungal infection was found (Fig 2). At this point, the diagnosis of idiopathic hypertrophic pachymeningitis was established. The patient experienced a complete loss of vision for 1 month.

COMMENT

We believe this case to be the first in the ophthalmic literature, presenting bilateral optic neuropathy associated with idiopathic hypertrophic pachymeningitis. Different pathological entities which can produce thickening of the dura mater, sarcoidosis, late syphilis, rheumatoid arthritis, tuberculosis, intracranial fibromatosis, fungal infection, and so on should be considered in the differential diagnosis.

In addition, Adler et al described a case of spinal aural pachymeningitis associated with pulmonary nodule, and concluded that this entity may be part of a systemic inflammation, like an autoimmune or connective tissue disorder. However, both clinical and laboratory findings were not suggestive for the existence of such conditions in the present case.

Initially, this case was treated as idiopathic optic neuritis and the steroid treatment was effective; however, owing to the development of hypertrophic pachymeningitis, the patient suffered a complete loss of vision. CT imaging and magnetic resonance imaging of the brain and orbit revealed no abnormalities at first, but subsequent neuroimaging enabled us to establish the diagnosis accurately. Although cases of hypertrophic cranial pachymeningitis are very rare, it may be added to the differential diagnosis of optic neuropathy.

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A case of traumatic retinal avulsion at the optic nerve head

EDITOR—We present an unusual case of blunt trauma to the optic nerve head. Following trauma the patient developed a retinal avulsion at the optic nerve head with subsequent total retinal detachment which resulted in a rubeotic eye.

CASE REPORT

A 37-year-old woman fell down a staircase and injured the left side of her face and brow directly against the edge of a radiator. She lost consciousness briefly and was admitted to the local district general hospital. A brow laceration was sutured and she was referred to the ophthalmic department because of reduced acuity.

Examination of the right eye was normal. There was no perception of light in the left eye. There was a left anterior uveitis: 2+ cells, 3+ flare. There was a left afferent pupillary defect present. Funduscopy findings showed a ring of haemorrhage at the optic nerve head and a pale swollen retina with a cherry red spot at the macula (Fig 1). A provisional diagnosis of anterior traumatic optic neuropathy was made. A B-scan ultrasound revealed some...
swelling around the left optic nerve head. A fluorescein angiogram (Fig 2) revealed a 360 degree section of retinal tissue from the edge of the optic disc. There was no filling of the retinal vasculature with copious leakage from the left optic nerve. Six months later she presented to the department with an acute development of rubetic glaucoma. A total retinal detachment was also noted at this stage.

COMMENT

To our knowledge this a unique case. In the literature there are several cases of traumatic anterior optic neuropathy but none described where the retina is completely severed around its insertion to the optic nerve head. The incidence of indirect traumatic optic neuropathy varies following closed head trauma (0.5–5%). Classically, injuries that occur anterior to where the central retinal artery enters the optic nerve produce abnormalities in the retinal circulation in association with visual loss. Further, avulsions of the optic nerve as it enters the globe produce a distinct picture with a partial ring of haemorrhage at the optic nerve head. We postulate that the central retinal artery and vein were consequently severed during the original injury which resulted in the absence of blood supply to the inner retina. The precise mechanism whereby this may have occurred is speculative. One possibility could be that a violent rotational injury occurred with sudden deformation of the globe. A pattern of central retinal occlusion was seen (pale ischaemic retina with a cherry red spot at the macula). The resulting ischaemia which ensued either from the former or from the total retinal detachment was the stimulus for neovascularisation with the development of rubetic iris and rubetic glaucoma. The development of this complication is unusual but is recognised (5%–20% of central retinal artery occlusion developing rubetic as a complication). 

Werner's syndrome

EDIToR,— Werner's syndrome, known also as paneraria or adult progeria, is an autosomal recessive inherited disease which displays the features of accelerated aging.

Werner's syndrome has been reported with diverse ocular changes such as juvenile cataract, nystagmus, blue sclera, proptosis, keratoconjunctivitis, cloudy corneas, iris telangiectasia, retinitis pigmentosa, macular degeneration, and choriotiretinitis. Cataract is the hallmark of Werner's syndrome, appearing usually between 10 and 20 years of age. Involvement is generally bilateral and visual acuity deteriorates rapidly. The recommended treatment in these cases is extracapsular cataract extraction. In Werner's syndrome, after cataract surgery, bullous keratopathy is a common complication and penetrating keratoplasty is suggested in these cases. Here, we describe a patient with Werner's syndrome and incomplete Leopard syndrome who had successful penetrating keratoplasty. No clinical signs of corneal graft failure were noticed after a 1 year follow up.

CASE REPORT

A 48-year-old Bedouin man, whose parents were first cousins, was followed in our centre. The patient's medical history included hoarseness, hearing difficulties, and marked sclerodactyly. He appeared older than his age, small in stature, with a typical bird-like facies, thin high pitched voice, and irregular teeth with malocclusion. The adipose tissue was reduced over all his body. The skin was tense, shiny, and adherent with innumerable hyperpigmented macules varying in size that resembled ephehides and simple lentigines.

At 30 years of age, he underwent uneventful bilateral extracapsular cataract extraction. Subsequently, a secondary glaucoma developed in both eyes and responded to antiglaucoma treatment. In 1992, aphakic bullous keratopathy appeared in the left eye while failing to respond to topical treatment.

In 1994, the patient complained of severe pain and a decrease in visual acuity in the left eye. His best corrected visual acuity with spectacles was 6/15 right eye with a refraction of +12.0 dioptres and hand motion with full light perception left eye with +10.0 dioptres. Slit-lamp examination in the left eye showed a severe bullous keratopathy with diffuse corneal opacities and deep corneal neovascularisation. In the right eye, the cornea was clear. Both eyes were aphakic. Intraocular pressure was normal in both eyes. Fundus examination in the right eye showed a total disc excavation. The left fundus could not be seen, and ultrasound did not reveal any posterior segment abnormalities.

He underwent uneventful penetrating keratoplasty in his left eye. Histopathological examination of the corneal showed bullous formation and extensive subepithelial, fibrous tissue that had replaced Bowman's layer. Twelve months later, the corneal graft was clear and visual acuity had improved.

COMMENT

The patient described displayed the classic features of Werner's syndrome. He had 12 of the 17 criteria defined by Thannhauser¹ and completed by Salt⁵ (Table 1). In addition to the classic features of Werner's syndrome, our patient had some features that are not found in Werner's syndrome but are, however, found in Leopard syndrome (that is, ECG changes and sensorineural deafness)³ (Table 2). He also displayed some features that are common to Werner's syndrome and to Leopard syndrome (growth retardation, abnormal genitalia, and endocrine disorders). The histological and ultrastructural findings of some lentigines showed the presence of melanosome in Langerhans cells as reported recently in Leopard syndrome.⁷

The possible existence of a generalised mesenchymal defect had been suggested in Werner's syndrome.⁵ Our patient may be an example of an association of genetic defects affecting both tissues of neuroectodermal and...
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