

Journal of Clinical Neuroscience (1999) 6(1), pp. 46-48. Copyright © 1999 Harcourt Brace & Co. Ltd

Baló's Concentric Sclerosis In A Woman From Papua New Guinea

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Abstract

We report a case of Baló's concentric sclerosis (a variant of multiple sclerosis) from Papua New Guinea. A 42-year-old woman with a past episode of optic neuritis presented with a left hemiparesis. Magnetic resonance imaging revealed a solitary large tumour-like right cerebral lesion with a pattern of concentric bands of different signal intensities. The diagnosis was established by biopsy of the lesion. To our knowledge, this is the first reported case of Baló's concentric sclerosis in the indigenous population of Papua New Guinea.

Keywords

Baló; concentric sclerosis; magnetic resonance imaging; multiple sclerosis; Papua New Guinea

Introduction

Baló's concentric sclerosis is a rare variant of multiple sclerosis. It is characterized by lesions with an unusual concentric pattern of alternating demyelinated and partially myelinated bands in the cerebral white matter.^{1,2} In 1928, Baló reported this pattern in lesions found at autopsy in a 23-year-old male with right hemiparesis, aphasia and papilloedema³ but the first reported case was that described by Marburg in 1906 as 'acute multiple sclerosis'.⁴ Concentric sclerosis usually affects young adults and has an acute or subacute onset and progressive course.¹ There is one reported case of typical multiple sclerosis in the indigenous population of Papua New Guinea,⁵ but to our knowledge this is the first reported case of Baló's concentric sclerosis in this population.

Case Report

A 42-year-old woman from Papua New Guinea developed blurring of vision in the lower half of the visual field of her left eye in August 1992. This was associated with pain behind her left eye which was aggravated by eye movement. Over the following 2 days, the vision in her left eye gradually deteriorated to almost complete visual loss. Her past medical history included tuberculosis at the age of 15 years and recurrent bouts of malaria.

In September 1992 she was admitted to the Royal Brisbane Hospital. On examination of the right eye the visual acuity was 6/6 and colour vision was normal. On the left the visual acuity was less than 6/60 and there was a dense central scotoma. There was a left afferent pupillary defect. The right optic disc was normal but the left was slightly swollen. The eye movements and remainder of the neurological examination were normal. A computed tomography (CT) head scan showed thickening of the posterior two-thirds of the left optic nerve. Oligoclonal immunoglobulin G (IgG) bands were present in both the cerebrospinal fluid (CSF) and the serum. There were no acid-fast bacilli detected in the CSF.

A visual evoked response could not be elicited by stimulation of the left eye. A diagnosis of left optic neuritis was made. Her vision improved and the pain resolved, and she returned to Papua New Guinea. In December 1995 she was readmitted to the Royal Brisbane Hospital. Two and a half weeks prior to this, she had developed unsteadiness of gait, a heavy sensation in both

legs and numbness on the left side of her body and face, which was followed over the next few days by increasing weakness in her left leg and arm. There were no visual symptoms, speech problems or bladder or bowel disturbance. Examination revealed a mild left upper motor neurone facial palsy and weakness of elevation of the left shoulder. The optic discs were normal. There was no wasting and tone was normal. Apart from minimal left shoulder adduction, there was complete paralysis of the left upper limb. Power was moderately reduced in the left lower limb, especially distally. Apart from a mild reduction in hip flexion, power was normal on the right. She was unable to stand. The deep tendon reflexes were increased on the left and normal on the right. The left plantar response was absent and the right was flexor. There was a patchy left hemisensory disturbance. Coordination was intact.

There was a mild increase in the CSF total protein to 660 mg/litre (reference range: 150-600) and in the CSF IgG to 104 mg/litre (reference range: 10-60) without oligoclonal bands. The CSF leukocyte count was 4/gl. Cryptococcal antigen and acid-fast bacilli were not detected in the CSF. An autoantibody screen and syphilis serology were negative. Her erythrocyte sedimentation rate was 1 mm/h. Magnetic resonance imaging (MRI) of the brain showed a well defined lesion involving the deep white matter of the right parietal lobe extending across the midline through the corpus callosum. There was irregular peripheral enhancement after the intravenous administration of gadolinium, and minimal surrounding oedema (**Fig. 1**).

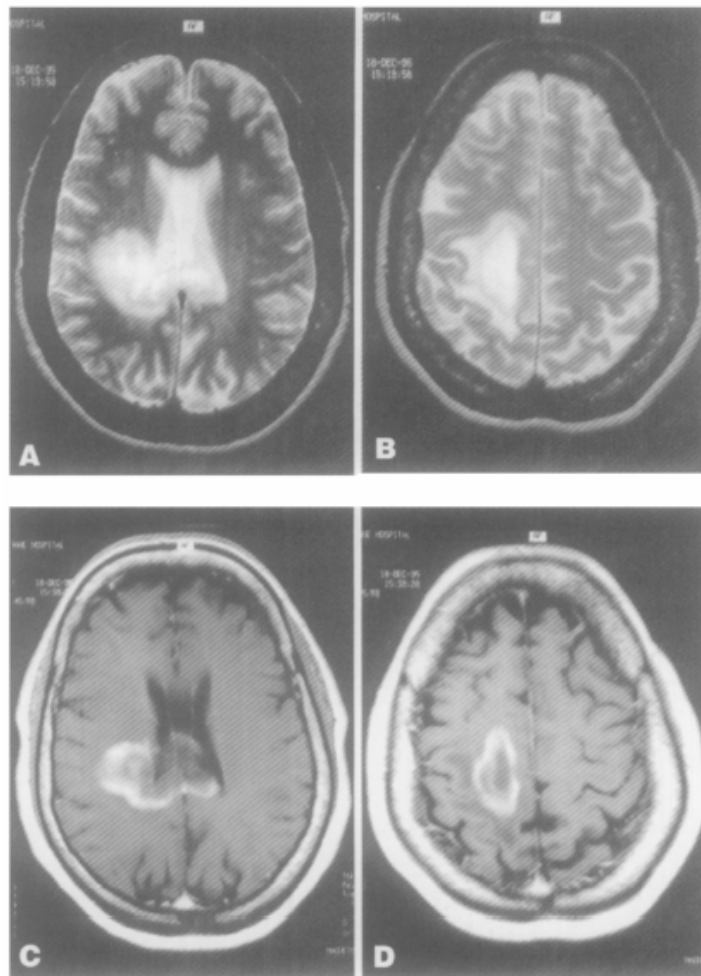


Fig. 1 Axial MRI scans showing the right cerebral lesion adjacent to the lateral ventricle. (A,B) T2-weighted. (C,D) T1-weighted after the intravenous injection of gadolinium.

A tentative diagnosis of intracranial tumour was made and she was commenced on oral dexamethasone. Because of her past history of tuberculosis and a positive Mantoux test, isoniazid and pyridoxine were given as prophylaxis. A stereotactic biopsy of the lesion revealed reactive changes with gliosis and lipid-laden phagocytes and no evidence of tumour. Special stains showed loss of myelin and relative preservation of axons within the lesion consistent with primary demyelination. She was therefore given intravenous methylprednisone 500 mg daily for 5 days. An MRI brain scan in January 1996 showed that the lesion was slightly smaller and did not enhance with gadolinium. Her strength had improved, with residual moderate left upper limb weakness and mild left lower limb weakness.

After discharge from hospital she attended outpatient rehabilitation with physiotherapy emphasising stance and gait training. An MRI brain scan in February showed that the lesion had enlarged laterally, anteriorly and superiorly with new areas of contrast enhancement. In March 1996 a repeat stereotactic brain biopsy again revealed demyelination with associated accumulation of foamy macrophages and reactive astrocytosis. Furthermore, a thin band of relatively preserved myelin was seen within the demyelinated region consistent with a diagnosis of Baló's concentric sclerosis (**Fig. 2**).

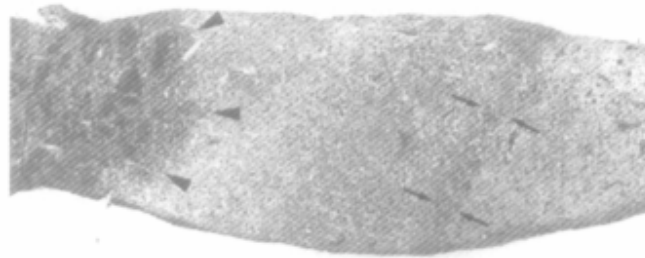


Fig. 2 Biopsy of the right cerebral lesion showing the sharp demarcation (arrowheads) between the normally myelinated white matter and the demyelinated area and showing a relatively preserved band of myelin (arrows) within the demyelinated region. Luxol Fast Blue. (x 23 magnification).

Although some axons were preserved in the demyelinated region, the number of axons was greatly reduced.

Discussion

Baló's concentric sclerosis is now generally considered to be a rare variant of multiple sclerosis.^{1,2} Baló suggested that 'encephalitis periaxialis concentrica' might represent a disease entity distinct from multiple sclerosis, partly because of the absence of spinal cord lesions.³ However, more recently Moore et al have reported a case demonstrating lesions of the Baló type in the lumbar spinal cord as well as in the medulla oblongata⁶ and Yao et al have described the histopathological findings of two patients with concurrent typical multiple sclerosis plaques and concentric sclerosis lesions and of another patient with these two lesion types in continuity.² The prior episode of left optic neuritis in our patient is consistent with the concept that Baló's concentric sclerosis is a variant of multiple sclerosis. In the present case, the diagnosis of Baló's concentric sclerosis was made on the basis of the second biopsy of the cerebral white matter lesion and supportive MRI findings. The first antemortem diagnosis of this condition was reported in 1986 by Garbem et al and was based on a needle biopsy.⁷

Antemortem diagnosis has also been made on the basis of characteristic MRI findings.^{8,9} The initial provisional diagnosis in the present case was intracerebral tumour. It has been reported that multiple sclerosis plaques may occur as single large lesions that resemble a tumour radiologically.¹⁰ In particular, the presentation of Baló's concentric sclerosis not infrequently mimics that of an intracerebral tumour.^{8,9,11} On review of our patient's initial MRI scan, a pattern of concentric bands of different signal intensities was noted (**Fig. 1B**), as previously described in

Baló's concentric sclerosis.^{8,9,11} Our case highlights the importance of considering inflammatory demyelination in the differential diagnosis of a solitary intracerebral lesion. Although the prognosis of the large focal tumourlike demyelinating lesions of the brain reported by Kepes¹⁰ was usually good, Baló's concentric sclerosis usually follows a progressive course.¹

The pathogenesis of the concentric nature of the demyelinating lesions is not clearly understood. In an ultrastructural study, Moore et al found abnormally thin myelin sheaths and numerous oligodendrocytes, and proposed that the partially myelinated bands resulted from remyelination at the edges of demyelinated zones.⁶ On the other hand, Yao et al found reduced oligodendrocyte numbers and lower levels of messenger RNA for myelin-related proteins in the partially myelinated areas compared with the normal-appearing white matter, and concluded that ongoing myelin breakdown, rather than remyelination of previously demyelinated areas, was responsible for the thin myelin sheaths.²

It is possible that the concentric alternating banded pattern represents repeated immune-mediated attacks against myelin antigens, as there is increasing evidence that multiple sclerosis is an autoimmune disease.¹² Serial MRI has demonstrated that the lesions of multiple sclerosis expand in a centrifugal pattern,¹³ but in typical multiple sclerosis there are many smaller lesions rather than a single large rapidly expanding lesion. Many of the recently reported cases of concentric sclerosis have come from China and the Philippines^{1,2} which have a low incidence of multiple sclerosis.

It is possible that, in Papua New Guinea, Baló's concentric sclerosis may also constitute a significant proportion of the cases of multiple sclerosis, as there has been only one reported case of typical multiple sclerosis in the indigenous population.⁵ There may be genetic and/or environmental factors in the Oriental and Melanesian populations that tend to focus the autoimmune attack in multiple sclerosis in a limited number of rapidly enlarging lesions, as occurs in Baló's concentric sclerosis.

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