High-Dose Steroid Treatment in a Patient With Balò Disease Diagnosed by Means of Magnetic Resonance Imaging

Nicholas J. Lanciano, DO*
Deborah S. Lyu, OMS III*
Carl Hoegerl, DO, MSc

Balò disease, also called concentric sclerosis, is a rare variant of multiple sclerosis that has been historically characterized as a monophasic, rapidly progressive, often fatal demyelinating disease. The diagnosis of Balò disease is based on the pathognomonic concentric ring pattern seen on magnetic resonance images. The authors describe the case of a 30-year-old woman with Balò disease who was initially treated with an unconventionally high dose of steroids and had a positive long-term clinical outcome with near-complete resolution of her neurologic symptoms. The authors also report MRI findings of concentric lesions before steroid treatment and resolution of the lesions shortly after and several months after steroid treatment. Given the outcome in this patient, we propose using high doses of steroids in the early treatment of patients with Balò disease. However, further studies are needed.

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Balò disease, or concentric sclerosis, was first described by József Balò in 1928.1 The case he described occurred in a young law student who presented with aphasia and right hemiplegia. This young male patient suffered progressive neurologic deterioration that was thought to be caused by a brain tumor. After his death, histologic examination of the affected tissue revealed bands of unmyelinated axons alternating with bands of preserved myelination. The axons were observed to be intact, with giant astrocytes containing multiple nuclei and lipid-filled macrophages. Balò termed the disease encephalitis periaxialis concentrica and considered it to be a variant of multiple sclerosis (MS) related to Marburg-type MS, or acute MS, which was first referenced in 1906.1,4

The pathogenesis of Balò disease remained somewhat of a matter of speculation until recently. It was originally believed that the concentric pattern observed in the disease was due to the axons undergoing demyelination from neuronal injury, followed by remyelination.5,6 A 2001 report6 supports the theory that the axons are not undergoing remyelination; rather, the areas of the axons that are myelinated at histologic examination have actually been spared from demyelination. The concentric layers may be spared from demyelination by protective preconditioning molecules induced in response to hypoxia-like tissue injury.2,6,7,8

Balò disease affects mostly young adults and appears to have a slight predilection for individuals of Asian descent.9,10 Because Balò disease can involve any area of the central nervous system, patients with this condition may present with a variety of neurologic symptoms.11 The most frequently reported clinical features include headache, aphasia, cognitive or behavioral symptoms, and seizures.10

The course of Balò disease was historically believed to be fulminant and fatal, but recent case reports have suggested that this characterization may not be entirely accurate, especially if Balò disease is diagnosed early and managed with high-dose steroids or immune modulation therapy.11,12 Since magnetic resonance imaging (MRI) became available to demonstrate lesions before death,5,9,12 some cases of Balò disease have been reported to resolve spontaneously.2,10,11

Report of Case
A 30-year-old right-handed white woman presented to the emergency department with a 2-day history of mild aphasia and apraxia. She was seen by a psychiatrist at that time and was sent home after being evaluated for depression. The patient returned to the emergency department 2 days later because of worsening symptoms; her aphasia and apraxia had increased to the point that it was interfering with her occupation as an accountant. She was feeling confused and having trouble saying and remembering words, and she was having difficulty performing tasks at work that she had performed daily for a couple of years. She had become particularly alarmed when she could no longer manage to use text messaging on her cell phone. Her family history was significant for the pres-
ence of MS in 3 family members: her father, a paternal uncle, and a maternal cousin.

In addition to the symptoms already noted, the patient also had symptoms of depression. She denied any change in vision, eye pain, sensory loss, focal or generalized weaknesses, or difficulty with gait or balance. Her vital signs at physical examination included a blood pressure of 137/88 mm Hg, a heart rate of 87 beats per minute, a tympanic temperature of 97.9°F, and a respiratory rate of 16 breaths per minute. She was well nourished and in no acute distress. On auscultation, her heart rate and rhythm were regular without an audible murmur, and both lungs were clear. Her abdomen was soft and nontender with normal bowel sounds in the right lower quadrant.

At examination, the patient was alert and oriented to person, place, and time. Her recent and remote memories were intact, and her attention span and concentration were normal. In testing for aphasia, she demonstrated mild to moderate impairment of fluency and comprehension, but she was able to repeat, recall, and name familiar objects.

A neurologic examination revealed that the patient’s cranial nerves II through XII were intact bilaterally. Her deep tendon reflex scores on the left side were 2/4 in the upper extremity and 3/4 for the patellar reflex. The patient also had a positive Babinski sign on the left side. In both upper and lower extremities, she exhibited mild muscle weakness (4/5) on the left side and normal muscle strength (5/5) on the right. The neurologic examination findings were otherwise unremarkable.

The laboratory tests ordered for hematologic, metabolic, and liver function testing had normal results. The rapid plasma regain test was nonreactive, and results of the antinuclear antibody test and serologic testing for Lyme disease were normal. The patient’s vitamin B12 level and erythrocyte sedimentation rate were also normal. The MRI ordered at initial presentation and performed the same day showed a concentric lesion with alternating hypo- and hyperintense rings corresponding to bands of myelinated and demyelinated tissue, respectively. The concentric patterns seen on MRI resemble the rings of a tree and thus give Balò disease its hallmark appearance. Given the patient’s presentation, her family history of MS, and the pathognomonic concentric pattern seen on the MRI results13 (Figure 1 and Figure 2A), it was determined that Balò disease was the most likely diagnosis; this diagnosis was later confirmed by physicians at the Johns Hopkins Hospital, where she went for a second opinion.

Figure 1. Pathognomonic brain magnetic resonance imaging findings in a 30-year-old woman with Balò disease. T2-weighted axial image obtained at admission shows alternating concentric zones of demyelinated and myelinated white matter in the right frontal region, consistent with a diagnosis of Balò disease.

Figure 2. Contrast-enhanced T1-weighted sagittal brain magnetic resonance imaging (MRI) showing course of Balò disease in a 30-year-old woman before (A), during (B), and after (C) treatment with high-dose methylprednisolone. The MRI results obtained on admission (A) shows a concentric ring with active enhancement. By day 4 (B), ring of enhancement is resolved. After approximately 1 year (C), MRI results show a single patchy lesion without enhancement.
After the diagnosis of Baló disease, the patient was treated with intravenously (IV) administered methylprednisolone, 2000 mg/d for 3 days followed by 1000 mg/d for another 6 days. After the first 3 days at 2000 mg/day, she showed substantial improvement in her abstract reasoning and aphasias. This improvement was confirmed by a second MRI study at 4 days after admission that showed resolution of the previously enhanced ring (Figure 2B). At 9 days, the fluency of the patient’s speech had returned to normal, she no longer appeared to be aphasic, and her strength on the left side had returned to baseline. She still showed signs of mild apraxia, but her condition was deemed stable enough for her to be discharged. She was discharged on a treatment regimen of oral steroids (prednisone), with tapering doses over 4 weeks starting at 60 mg.

Neuroimaging studies performed during the first year after initial presentation demonstrated no additional lesions. The patient’s latest MRI (Figure 2C) revealed stabilization of the lesion with no signs of enhancement, and the patient reported that she has not experienced additional flares and was able to return to work shortly after her discharge from the hospital. Because of patient noncompliance, the patient did not follow up for almost 1 year after discharge from the hospital. However, at her latest physical examination, her neurologic deficits were nearly completely resolved.

Comment
Because of the rarity of Baló disease, little has been published on the preferred management of this condition. In most reported cases, the general treatment plan was similar to that used for acute MS attacks. Cases for which successful outcomes were reported received a treatment regimen of high-dose steroids, immunosuppressive therapy, or both; steroids were used more commonly. For case reports that identified steroid treatment have also varied widely. Many patients showed improvements within days to months, followed by complete recovery, relapse, or failure to recover. To our knowledge, no previously reported case of Baló disease has been managed with methylprednisolone IV at a dosage of more than 1000 mg/d. In the current report, we present a patient with Baló disease whose diagnosis was based on the pathognomonic concentric ring pattern seen at MRI. She showed a clinically significant recovery with a 9-day course of high-dose steroids (methylprednisolone, given IV, 2000 mg/d for 3 days followed by 1000 mg/d for 6 days), followed by tapering doses of oral steroids. After only 3 days of methylprednisolone treatment at 2000 mg/d, the patient began to show marked improvements in neurologic symptoms—findings supported by the improved MRI image obtained on day 4. Repeated MR imaging during the first year after initial presentation demonstrated no additional lesions, and the patient’s latest physical examination findings showed almost complete resolution of her neurologic symptoms.

Owing to its rapid progression and often fatal and unpredictable clinical outcomes, we believe that the inflammatory demyelination in Baló disease should be managed early with very high-dose steroids once the diagnosis is confirmed with MRI. Patients are generally extremely tolerant of short-term treatment with high-dose methylprednisolone administered IV; the most common adverse effects are psychiatric and gastrointestinal disturbances and an increased risk of infection and fracture.

Conclusion
Although further studies of early treatment for Baló disease are warranted, we propose here that treatment with unconventional high doses of steroids, administered IV, could be lifesaving and more effective than treatment with standard dosages.

References