# LETTER TO THE EDITOR

## To The Editor

### Torsional Nystagmus and Oscillopsia as Initial Presentation of Balo's Concentric Sclerosis

**Keywords:** Balo's Concentric Sclerosis, Multiple Sclerosis, Torsional Nystagmus

We describe a young man presenting with oscillopsia due to torsional nystagmus who had multiple concentric white matter lesions (WMLs) on brain neuroimaging and was eventually diagnosed with Balo's concentric sclerosis (BCS). We review recent literature on BCS which argues that this is a distinct clinical entity different from multiple sclerosis (MS).

A 40-year-old Caucasian man noticed vertigo, sudden onset binocular oblique diplopia, and oscillopsia for the past 2 weeks. He was healthy and did not take any medications but was a recreational cannabis user and smoked half-a-pack of cigarettes for over 20 years. He was diagnosed with benign paroxysmal positional vertigo in the local emergency department and prescribed betahistine. His visual symptoms persisted, and referral for neuro-ophthalmologic consultation was made. Afferent examination was normal. Efferent examination demonstrated slightly jerky smooth pursuit, normal saccadic velocity, and a high frequency torsional, nystagmus which worsened on right gaze. Head impulse test was negative. Detailed neurological examination was otherwise normal. Urgent magnetic resonance imaging (MRI) of the brain with contrast demonstrated one infratentorial and multiple supratentorial T2-hyperintense oval WMLs. Two lesions demonstrated concentric ring pattern with post-contrast enhancement (Figure 1). MRI spine was unremarkable. Cerebrospinal fluid (CSF) examination demonstrated normal composition and normal cytological examination, and there were no oligoclonal bands (OBs). Aquaporin-4 and myelin oligodendrocyte glycoprotein antibody testing were negative. Rheumatological and infectious workup was unrevealing. Abnormalities on brain MRI were suggestive of multiphasic demyelinating disease with evidence of dissemination in time and space; however, the size of the lesions, ring enhancement, and absence of OB in CSF were very unusual, thus diagnosis of BCS was made and torsional nystagmus was localized to enhancing right cerebellar lesion. Treatment with 1 g of intravenous solumedrol commenced. On follow-up 1 month later, oscillopsia resolved and previously seen torsional nystagmus was reduced in frequency and amplitudes. Follow-up MRI 2 months later showed decrease in the size of previously seen lesions and resolution of enhancement; however, a new non-enhancing lesion was now seen in centrum semiovale again with concentric ring appearance.

BCS is a rare demyelinating condition characterized by discrete radiological and pathological lesions of concentric ring morphology representing alternating regions of demyelinated and myelinated fibers.[1,2,3,4] It typically presents in fourth decade of life with female predominance and higher prevalence in Asians.[1,2] Clinical presentation varies from focal abnormalities seen in MS (focal paresis, sensory disturbance, and ataxia) to greater cortical dysfunction seen in patients with intracranial mass lesions (headache, seizures, cognitive difficulty, and behavioral change).[1,3] Apart from MS, differential diagnosis includes tumefactive demyelination, glioblastoma multiforme, primary central nervous system lymphoma, brain abscess, and infarction.[1,2] Historically, prognosis in patients with BCS was associated with high rate of significant disability or death. However, advent of MRI has facilitated earlier diagnosis enabling timely intervention with improved prognosis.[1,2,3] Classically, BCS is characterized by alternating isointense and hypointense concentric rings on T1 and hyperintense onion-like lamellae on T2-weighted sequences with peripheral post-contrast enhancement of the lesions indicating disease activity.[1,2] Unlike MS, supratentorial WNLs typically spare cortical U fibers.[1,2] OBs in CSF are typically absent in BCS in contrast to MS where they are present in over 95% of cases.[1,2,3,4]

It is uncertain whether BCS is a phenotypical variant of MS or a separate entity of atypical demyelinating disease.[1,2,3,4] Case

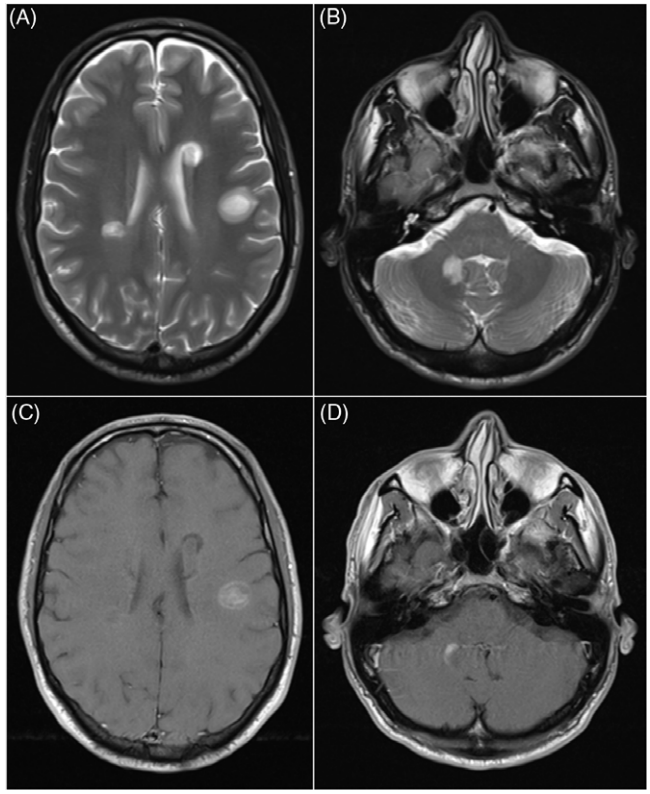


***Figure 1:*** *Brain MRI demonstrating multiple T2-hyperintense lesions with concentric ring pattern within supratentorial region (A) and one within the infratentorial right cerebellum (B); corresponding brain MRI post-contrast T1-enhancing lesions demonstrating active disease (C and D).*



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reports describing this entity refer to it either as BCS or "Balo-like lesions in MS" interchangeably[1,3] creating ambiguity resulting in indistinguishable disease profiling. While there have been reports of BCS or Balo-like lesions presenting concomitantly with MS or known to later progress to typical MS indicating possible overlap, there is some evidence based on lesion size, morphology, presentation, and severity distinguishing BCS as a separate entity.[4] Jarius et al. performed a retrospective analysis of 146 CSF profiles in 132 patients with BCS features and reported absence of OBs in two-thirds of patients ($p < 0.000001$ compared with MS) with even higher difference in the subgroup with both radiological and histopathological evidence of BCS. Since OBs represent a qualitative marker of intrathecal IgG synthesis and are almost universally present in patients with MS, this study suggested that BCS is an immune-pathogenetically distinct entity.[4]

Treatment guidelines in BCS are limited due to its rarity. Current literature supports the use of high-dose corticosteroids for active disease which may also improve prognosis.[1,2] For cases with high disease burden and those with poor steroid response or relapses, intervention is case-dependent. Plasma exchange may be considered as a form of rescue therapy, while other treatments such as intravenous immunoglobulin, cyclophosphamide, and other disease-modifying immunosuppressants have also been used in case reports. Currently, there is insufficient evidence to decide which of these therapies are most beneficial and whether implementation of maintenance therapy is efficacious in preventing relapse.[1,2]

In summary, our case aims to raise awareness of BCS as a potentially distinct entity and highlights the value of CSF investigations in patients with concentric WMLs. Patients with BCS presentation should be referred to tertiary care centers in order to improve clinical, radiological, immunochemical, and pathological profiling as well as document treatment outcome to better define BCS as a disease entity.

## Disclosures

The authors have no conflicts of interest to declare.

## Statement of Authorship

Both authors (EM and TJP) have contributed equally to data aquisition, manuscript writing, figure preparation, and revision of the manuscript.

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