



Case Report

Balo's concentric sclerosis a rare variant of multiple sclerosis in a Nigerian adult male: A case report

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Received : 15 July 2022
Accepted : 09 September 2022
Published : 28 October 2022

DOI
[10.25259/SNI_636_2022](https://doi.org/10.25259/SNI_636_2022)

Quick Response Code:



ABSTRACT

Background: Balo's concentric sclerosis (BCS) is a rare variant of multiple sclerosis (MS) that usually runs a benign course. Typically, brain magnetic resonance imaging (MRI) shows concentric lamella of areas of demyelination alternating with normal zones of myelin preservation. Clinically, it may be mistaken for other intracranial space-occupying lesions, especially in our subregion, where neuroimaging facilities are scarce or not affordable. The authors report a rare case of BCS in a young Nigerian male diagnosed and managed in Enugu, Nigeria. This is possibly the first reported case in sub-Saharan Africa.

Case Description: The patient presented with behavioral changes, focal seizures, cranial neuropathy, long tract signs, and cerebellar dysfunction causing severe disability and functional impairment with a Modified Rankin Score (mRS) of 4. Brain MRI showed multiple heterogeneous lesions in the right cerebral hemisphere, brainstem, and cerebellum with features in keeping with BCS. Cerebrospinal fluid oligoclonal bands were negative and C-reactive protein was normal. He had high-dose steroids and immunosuppressive therapy. He made a gradual neurological improvement and significant symptom resolution and was able to return to work after a year (mRS-1).

Conclusion: BCS is a rare variant of MS in sub-Saharan Africa. The mainstay of diagnosis is MRI which shows the typical concentric demyelinating pattern. Steroids and immunosuppressive therapy are effective in the treatment of BCS.

Keywords: Balo's concentric sclerosis, Demyelination, Intracranial space-occupying lesion, Multiple sclerosis

INTRODUCTION

Balo's concentric sclerosis (BCS) is a rare neuroinflammatory disease characterized by concentric rings of white matter demyelination. It is a variant of multiple sclerosis (MS) initially described by Joseph Balo, in 1928, which runs a benign course with a fairly good outcome when compared to other demyelinating lesions.^[2] It is associated with cerebral white matter oligodendrocyte loss, astrocytopathy, and neuronal injury similar to what is seen in MS.^[1,5,8] The neuroinflammation seems to arise from a perivenular zone located deep in the white matter and spread centrifugally

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outward.^[5,8] The lesion may be solitary or multiple and often mimic intracranial space-occupying lesions (ICSOL) and other differential diagnoses.

Brain magnetic resonance imaging (MRI) is the diagnostic investigation of choice. A collaborative report in the literature confirmed the characteristic concentric lamella of demyelination alternating with normal zones of myelin preservation seen in both FLAIR, T1- and T2-weighted images which give the lesion an onion peel appearance.^[1,3,5,7] Other complex patterns have been described and there may be variable postcontrast enhancement dependent on the degree of the neuroinflammation present.^[1,8]

The authors report a rare case of BCS in a young Nigerian male diagnosed and managed in Enugu Nigeria. This is probably the first reported case in sub-Saharan Africa.

CASE DESCRIPTION

The patient is a 31-year-old man who presented with a history of recurrent throbbing headaches of 10 weeks duration (Visual analog score 7-8/10). There were associated focal seizures involving the left upper limb which resolved with the administration of antiepileptic medications, personality changes characterized predominately by aggressive and irrational behavior as well as gradual onset but progressive weakness of the left side of the body.

Examination revealed that he was fully conscious with dysarthria. He had left facial nerve palsy upper motor neuron

type, but vision, hearing, and other lower cranial nerves were normal. He also had left-sided spastic hemiparesis with a power of 3 to 4-5 (medical research council scale) and a significant left hemisensory deficit. The gait was grossly ataxic with associated recurrent falls predominately to the left. His Modified Rankin Score (mRS) was 4 at presentation. He had a course of low-dose oral steroids in the referral hospital before presentation to our center without any improvement.

Brain MRI showed multiple heterogeneous lesions in the right cerebral hemisphere, brainstem, and cerebellum. The largest lesion was in the right capsuloganglionic area extending into the deep white matter and displaying alternating isointense and hypointense concentric rings/bars on T1WI [Figure 1]. The T2WI [Figures 1a-c and 2a-f] images showed similar isointense and hyperintense lamellae with a patchy enhancement pattern on the administration of gadolinium contrast [Figures 2g and h]. Similar lesions of varying intensities were noted in the right frontal, left occipital, and left cerebellar regions [Figures 1c-d and 2e-f]. Cervical MRI was normal [Figures 3a and b].

Cerebrospinal fluid (CSF) oligoclonal bands were negative, and CSF chemistry and cell count were normal. CSF culture was negative; however, the cytology showed fluffy cotton wool exudates in keeping with the presence of a possible degenerative or neuroinflammatory lesion. CRP was normal (<2.50 mg/dl.)

He was treated with a combination of steroids and immunosuppressive agents. The first course was high-dose steroids (intravenous methylprednisolone 1 g daily for 5 days

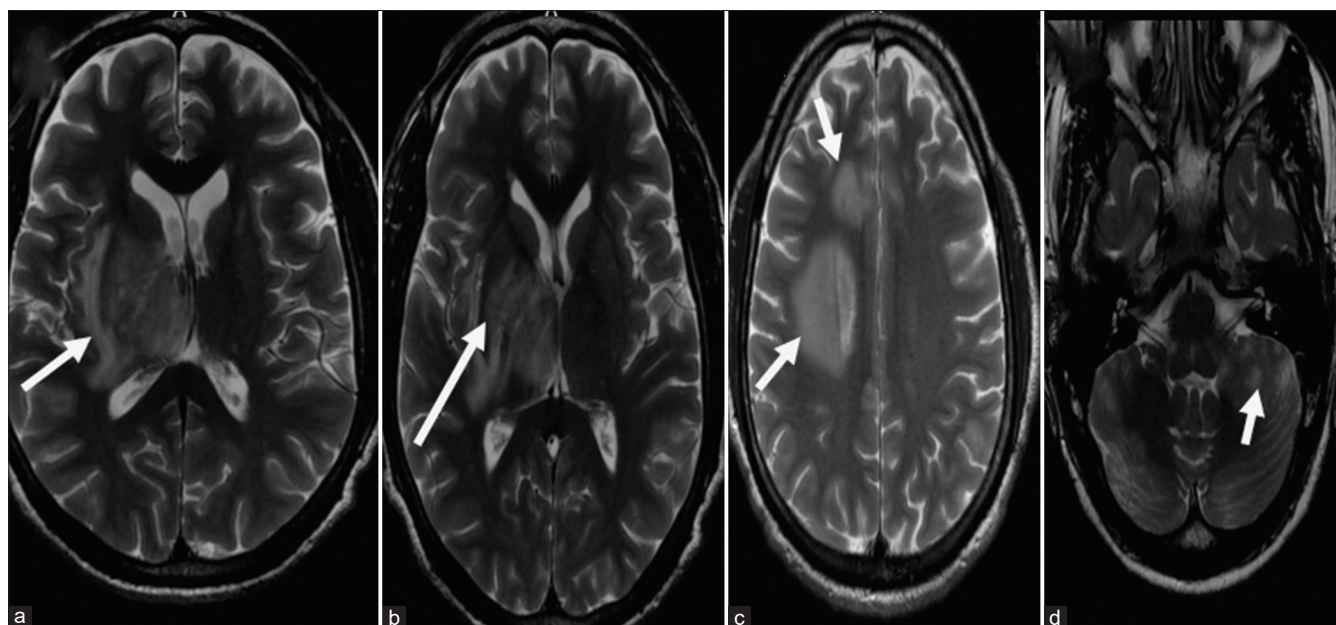


Figure 1: Brain magnetic resonance imaging (MRI) of the patient at presentation (a-d) axial cuts of T2-weighted image of brain MRI of index patient showing multiple hyperintense lesions largest in the right capsuloganglionic area and adjacent white matter displaying concentric rings/bars (a-c). (d) Shows a similar hyperintense lesion in the left cerebellar hemisphere (white arrow depicts the lesion).

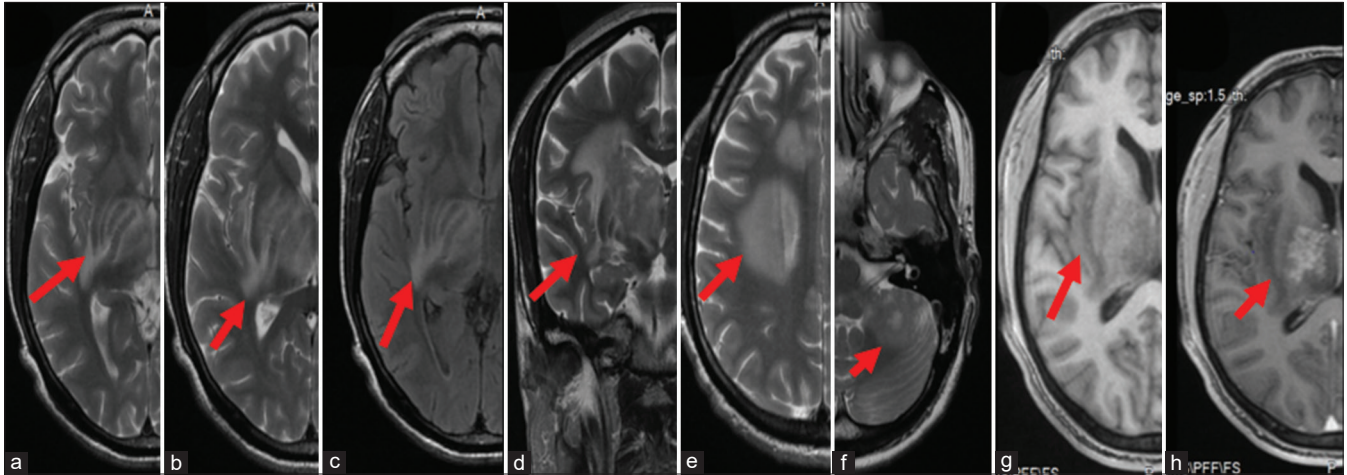


Figure 2: Brain MRI of index patient before treatment (a-h): Brain MRI showing T2, FLAIR, T2, T1, figures 2a-f and postcontrast images of BCS depicting the alternating concentric lamella of demyelination plaques and punctate contrast enhancement (g and h) Red arrow shows the lesion.

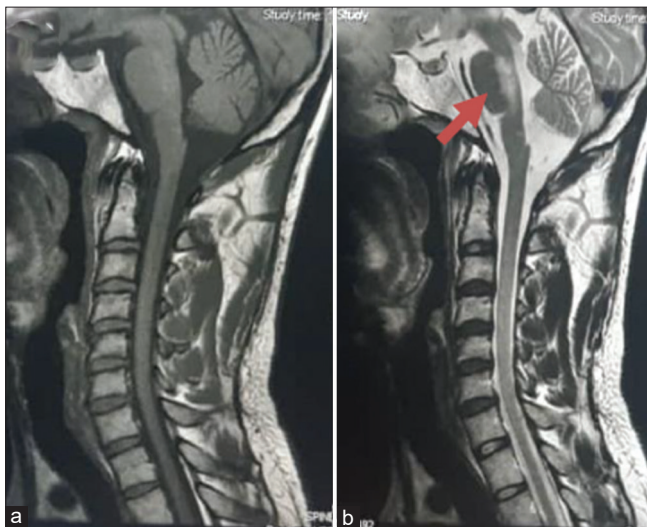


Figure 3: Cervical spine MRI (a and b) shows normal T1WI and T2WI of the cervical spine. Arrow (b) points to brainstem hyperintensity suggestive of demyelinating lesion.

followed by a maintenance dose of oral prednisolone). He made remarkable improvement after treatment with resolution of the headaches, speech deficits, and hemibody weakness. He had significant improvement in his personality; however, the gait and coordination problems persisted causing some limitations in his ability to carry out activities of daily living (mRS of 2).

Five months after the first course of steroid therapy, he had progressive worsening of speech, gait, and limb weakness that necessitated a repeat course of treatment and maintenance with oral azathioprine. He made a gradual but progressive improvement and almost complete resolution of the speech deficits, hemibody weakness, gait, and coordination problems for 2–3 months. Brain MRI repeated 2 months after the second course showed almost complete resolution of the

lesions with no associated edema or contrast enhancement [Figures 4 and 5].

He had regular physical therapy and rehabilitation and was able to return to work with mRS of 1 at 18 months follow-up.

CASE DISCUSSION

BCS is described as a variant of MS and usually presents with single or multiple concentric tumefactive-like lesions which causes widespread neurologic dysfunction.^[5] In the subregion, where the common etiologies of ICSOL are tuberculosis, toxoplasmosis, and primary central nervous system lymphoma among others, especially in the immunocompromised, BCS though rare, should be considered a differential diagnosis. The increasing availability of MRI in sub-Saharan Africa has made this diagnosis possible despite the suspected rarity of MS and its tumefactive variants in Africans.^[10,11] Karaarslan *et al.* reported five cases based on clinical, pathological, and MRI features and inferred that BCS may be a self-limiting disease that is not always fatal.^[7]

The MRI striking features of BCS are reminiscent of histopathology comparison of this rare demyelinating disease. These characteristic MRI imaging findings from the high tesla images allow antemortem diagnosis of BCS when performed at the onset of the disease. This helps to solve the diagnostic dilemma associated with this disease which before now was made postmortem or following biopsy of a suspected ICSOL.

Our index patient had behavioral changes, cranial neuropathy, long tract signs, and cerebellar dysfunction that significantly affected his quality of life and MRI features showing multiple demyelinating plaques in the subcortical, deep white matter, and the cerebellum with features in keeping with BCS. On treatment with high-dose steroids, he showed remarkable

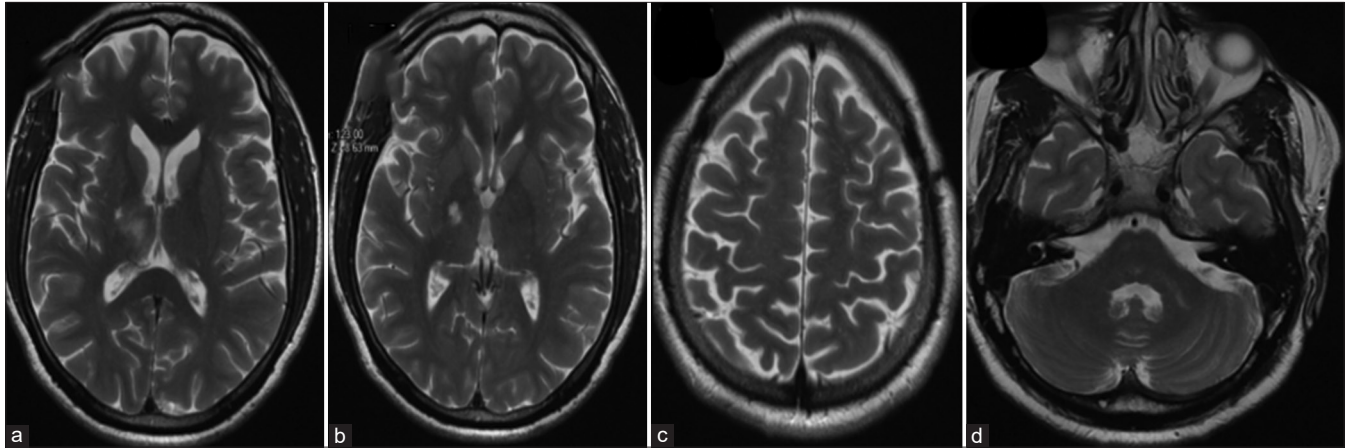


Figure 4: Brain MRI after two courses of high-dose steroids and immunosuppressive agents (a-d) shows similar cuts in Figure 1 of the same patient 7 months after treatment. Note almost complete resolution of inflammatory plaques.

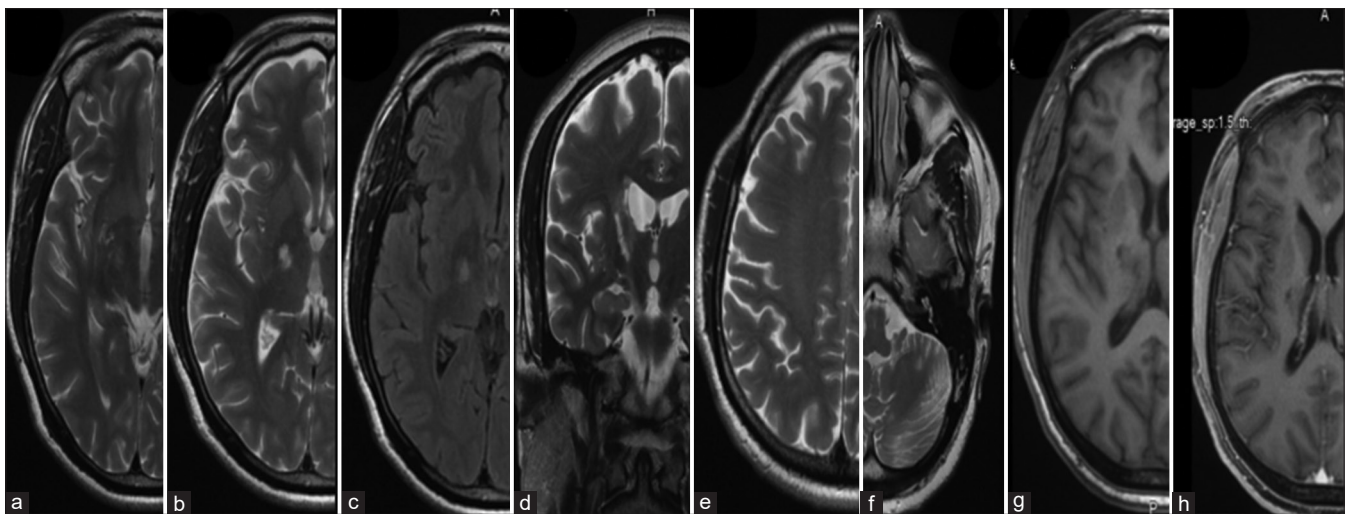


Figure 5: Brain MRI patient after treatment (a-h) T2, FLAIR, T2, T1WI, and postcontrast images show almost complete resolution of the inflammatory plaques after treatment with high-dose steroids and immunosuppressive. Compare with findings in Figure 2.

clinical response with an improvement of mRS from 4 to 1 and almost complete disappearance of the lesions on repeat brain MRI [Figures 4 and 5]. This clinical and radiologic response supports an inflammatory demyelinating lesion.^[1,5,8]

The cause of the neuroinflammation in BCS is not clear, but it is believed that unknown toxins or substances are released from the perivenular zone at the geometric center of the lesion and spread outward.^[5,8] These substances activate T-cells, macrophages, and microglia to release cytokines and chemokines that spread outward in a wave-like pattern from the focal center interacting with the oligodendrocytes nearby causing a varying degree of demyelination of the axons and subsequent remyelination giving the characteristic onion peel appearance. High-dose steroids in most cases suppress this inflammation reaction and lead to its resolution over time.^[7,8] The judicious use of

high-dose steroids and other immunosuppressive remains the mainstay of treatment and response is often remarkable as noted in many reports,^[1,3,5] that is similar to the finding in our index patient.

Although the usual concentric lamella of demyelinating plaques alternating with areas of myelin preservation is the predominant findings of BCS, other studies have noted varied presentation patterns on MRI including distorted rings, rosettes, carnation pattern, and even parallel and rectangular bars.^[1,7,8] This bizarre pattern may occur due to prior administration of steroids as noted in our index case.

About half of BCS coexists with MS and this may explain the varying expression of oligoclonal bands in CSF of patients with BCS.^[5,12] Studies have shown that patients with positive oligoclonal bands and MRI features suggestive of BCS will

likely progress to remitting and relapsing forms of MS on long-term follow-up.^[5,9] Some other authors have argued that BCS and MS are the same spectra of diseases with different expressions, and both are characterized by predominately T-cells mediated inflammation with B-cells response commonly found in MS.^[1,4-6] In addition, the absence of oligoclonal bands in BCS is attributed to the paucity of B-cell and plasma cell clonal response.

Although BCS runs a benign course, it is essential to counsel patients about the risk of recurrence which may occur anytime during care and follow-up. This may arise from a preexisting lesion(s) or *denovo* from previously uninvolved sites typically disseminated in space and time as observed in MS.^[4]

BCS, although rare, requires a high index of suspicion during clinical and neuroimaging evaluation of patients suspected to have multiple intracranial lesions. Resource-constrained regions with limited MRI facilities may miss this potential variant of MS that runs a rather benign course resulting in a diagnostic dilemma, unnecessary biopsies, and delay to institute appropriate management.

CONCLUSION

BCS, a rare variant of MS, exists in sub-Saharan Africa. The mainstay of diagnosis is MRI which shows the typical concentric demyelinating pattern in FLAIR, T1-, and T2-sequences. Its course is often benign but can present with multiple recurrences and relapses. Steroids and immunosuppressive agents are often effective in the management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Campbell FC, Oti B, Ndafia NM, Hart I, Okwunodulu O, Ndubuisi CA, *et al.* Balo's concentric sclerosis a rare variant of multiple sclerosis in a Nigerian adult male: A case report. *Surg Neurol Int* 2022;13:486.