



## Case Report

## Balo's Concentric Sclerosis with monophasic course: A report of 2 cases

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## ABSTRACT

**Introduction:** Balo's Concentric Sclerosis (BCS) is a rare demyelinating disease sometimes considered a variant of multiple sclerosis. It is characterized by an acute or subacute neurological symptoms with characteristic MRI "onion-like" white matter lesions. BCS has a wide range of presentations but is mostly self-limiting. Steroids are indicated in patients with aggressive disease.

**Case presentation:** We report 2 cases of BCS with monophasic course of stroke-like symptoms and single periventricular concentric lamella with onion-like appearance on MRI without inflammatory reaction in the CSF. They were treated with corticosteroids achieving clinical improvement and without neurological deficit or relapse over the following years.

**Clinical discussion:** A number of cases of BCS are described in the literature that show marked recovery with early diagnosis and treatment with steroids.

**Conclusion:** BCS appears to have a good prognosis when treated early in its diagnosis with steroids.

## 1. Introduction

Balo's Concentric Sclerosis (BCS) is a rare demyelinating disease considered a variant of multiple sclerosis (MS). It presents acutely with focal neurological deterioration. Magnetic Resonance Imaging (MRI) is the diagnostic modality, which usually reveals concentric lamella in cerebral white matter with "onion-like" lesions. BCS, also known as the Marburg variant, is regarded as a variant of MS. BCS was first described by Marburg in 1906 as "acute MS" and then by Barré in 1926 [1]. In 1928 Balo reported a young law student with a progressive neurological disorder who died 3 months after the onset and the postmortem gross examination revealed bilateral large lesions of concentric lamella of alternating demyelinated and partially myelinated tissues involving the corpus callosum and right hemisphere, the white matter of the left hemisphere was also spotted [2].

BCS often affects young adults and three different clinical courses have been described; including an acute and self-limiting, remitting-relapsing variant and rapidly progressive primary disease [3]. These last 2's clinical and radiological characteristics appear to be more strongly related to MS. MRI has facilitated diagnostic due to its ease to detect

characteristic lesions, current diagnosis relies on imaging and clinical presentation. CSF analysis often shows few mononuclear cells, proteins and occasionally oligoclonal bands (OCB). BCS usually courses with solitary lesions, which may persist for years. These lesions may regress and resemble typical demyelinating plaques [1,3–6].

BCS relation with MS is still unclear, there is controversy on whether BCS is a variant of MS or separate entity [3–6]. Prior authors have hypothesized increased risk of progression to MS when multiple bands and/or oligoclonal bands are found. In this report we describe two patients presenting at an academic institution with BCS, which presented with a monophasic course, and characteristic Balo lesions in MRI that disappeared during a long term follow up.

This case report has been reported in line with the SCARE Criteria [7]. Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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## 1.1. Case reports

### 1.1.1. Case 1

A 44-year-old man presented to the emergency room in September 2010 with sudden onset of dysarthria and dysgraphia mimicking stroke. No history of cerebrovascular risk factors. No relevant family history, past surgical or medical history as well as allergies were reported by the patient. An enhanced MRI showed a periventricular concentric demyelinating lesion in left periventricular area (Fig. 1A and B). Clinical and imaging findings raised suspicion for isolated MS clinical syndrome. A lumbar puncture revealed one OCB, 3 mononuclear cells and normal proteins in CSF. Somatosensory evoked potentials were compatible with demyelinating disease. The patient received IV methylprednisolone (1000 mg/day for 5 days) showing clinical improvement and was discharged 5 days later without any neurological deficit. His neurologist prescribed interferon, once a week subcutaneously that was suspended one month later due to side effects. Case reassessment from a neurologist consulted for a second opinion suggested BCS. The patient has remained asymptomatic and 10 years later (September 2020). Follow-up MRI revealed small gliosis in the left periventricular area (Fig. 1C).

### 1.1.2. Case 2

A 25-year-old female without past family or medical history presented to the emergency room with left sided hemiparesis and left facial palsy in September 2020. On admission, the patient was hemodynamically stable, afebrile and the neurological examination revealed in the upper and lower left limbs weakness 2/5 in proximal and distal limb sectors, Hoffman and Trömmer signs in the left side without Babinski response. Deep tendon reflexes were normal, as well as sensorial examination, the rest of cranial nerves did not revealed abnormalities. Stroke protocol MRI revealed a right sided periventricular concentric demyelinating lesion with a concentric ring pattern that showed enhancement after endovenous contrast injection. The typical pattern of alternating hypointense-isointense and hyperintense rings on T2-weighted images were present (Fig. 1 D-F). The CSF showed normal proteins, no leukocytes and the OCB were negative. The diagnosis of BCS was established and the patient received IV methylprednisolone. (1000 mg/day for 5 days) and a remarkable clinical improvement was observed and one week later the patient was discharged without

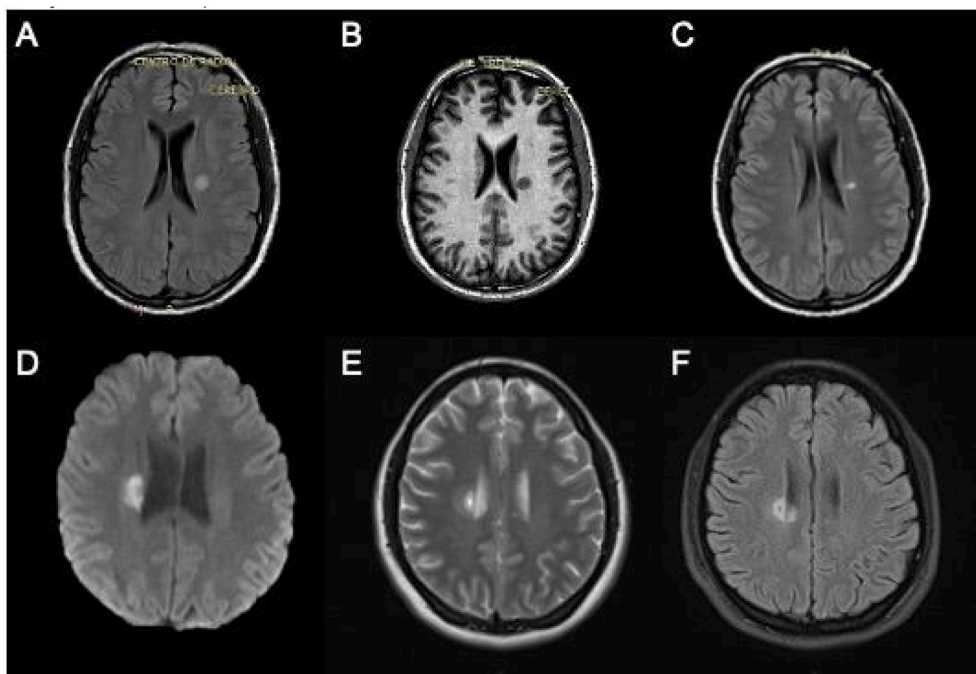
neurological deficit. She remains asymptomatic eight months after diagnosis.

## 2. Discussion

In this report we describe two patients with BCS, that showed monophasic course characterized by a clinical onset mimicking stroke with no inflammatory changes in CSF, one case with a single OCB and both cases showing a single Balo lesions in MRI. These two cases improved after steroid treatment and during the long term follow up (8 months–10 years) they remain in normal clinical condition and no active brain lesions were observed on MRI (Fig. 1C).

As in our cases, BCS often affects young adults and three different clinical manifestations are described including an acute and self-limiting, remitting-relapsing variant and rapidly progressive primary disease [3–5]. The cases presented, as those in the majority of the literature were acute-self limiting. A number of BCS cases have been described in the literature corresponded to the remitting-relapsing or rapidly progressive primary disease, that we considered that these cases are MS associated with Balo lesions or inclusive Marburg variant of MS and these patients died early in the follow up. Moreover, the autopsy reported by Balo, described bilateral large lesions of concentric lamella involving the corpus callosum, right and left hemispheres, and the white matter of the left hemisphere was also spotted [2]. The Balo description suggests that the concentric lamella were associated with rapidly progressive demyelinating disorder [1,2]. Overall 40 cases have been described in the literature included the 2 herein. In this cohort, 5 recurrences have been recorded, suggesting an overall good prognosis. Only one death has been recorded and most make full or partial recovery. Further effort to determine if full recovery can be achieved with variations on treatment is worth pursuing. All cases were treated with steroids, however dosage and timing still need to be defined and explore its possible relation with outcomes. Our scheme of 1000 mg IV methylprednisolone for 5 days achieved full recovery in our 2 patients. Also longer observation periods are needed to confidently define relapse rate. Additionally future publications could focus on BCS cases with aggressive course and further explore its possible relation to MS.

The diagnosis of BCS is actually based on clinical and MRI findings [1,3–8T]. The CSF often shows proteins, few mononuclear cells, and



**Fig. 1.** Case 1: A) Axial T2 weighted obtained in 2010 showing the typical pattern of alternating isointense and hyperintense rings and B) Axial T1 weighted image obtained in 2010 displays a hypointense rounded Balo lesion. C) FLAIR axial view obtained in 2020 revealing gliosis where the Balo lesion was observed. Case 2: D) Axial-Diffusion weighted image, E) Axial T2 weighted image and F) FLAIR image are showing the typical pattern of alternating hypointense-isointense and hyperintense rings in the right periventricular region.

occasionally OCB. In the preset series, the CSF was normal one OCB was detected in one patient. A monophasic neurological expression of symptoms usually suggests the presence of single Balo concentric lesion and in BCS is rare to find multiple lesions. Serial MRI may reveal the outcome of a Balo in the long term, our cases revealed small gliosis where Balo lesion was previously observed ten years after the onset (Fig. 1A–C). In this report, we describe two young patients mimicking a stroke-like syndrome and the MRI showed the typical pattern of alternating hypointense-isointense and hyperintense rings on FLAIR, T2 and T1 weighted images. No abnormalities in the CSF were observed and both patients presented a remarkable good response to steroid treatment. As in our cases, 39 BCS patients are described in the literature that presented similar monophasic clinical course with no other demyelinating lesions compatibles with MS in the MRI and showing complete recovery in most of them and without neurological deficit after steroids treatment (Table 1). A number of patients with BCS with additional demyelinating lesions including Marburg variant in MRI and without monophasic clinical course, were usually associated bad prognosis and death.

While evidence provided by case reports is limited, we provided a comprehensive review of key data from similar cases and added our own experience to the available pool. Our experience further suggest the benign course of BCS and its responsiveness to steroids.

### 3. Conclusion

We presented cases suggestive of monophasic BCS with an acute self limiting presentation, with matching imaging findings. Methylprednisolone 1000 mg IV for 5 days as treatment allowed for full recovery in both patients. Current literature suggests most cases of BCS are self limited and respond well to steroids. Further studies are needed to determine if BCS is a disease on its own or a variant of MS.

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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### 4. Annals of medicine and surgery

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All authors must disclose any financial and personal relationships with other people or organisations that could inappropriately influence (bias) their work. Examples of potential conflicts of interest include employment, consultancies, stock ownership, honoraria, paid expert testimony, patent applications/registrations, and grants or other funding.

None to disclose.

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All sources of funding should be declared as an acknowledgement at the end of the text. Authors should declare the role of study sponsors, if any, in the collection, analysis and interpretation of data; in the writing of the manuscript; and in the decision to submit the manuscript for publication. If the study sponsors had no such involvement, the authors should so state.

No funding received.

#### Ethical approval

Research studies involving patients require ethical approval. Please state whether approval has been given, name the relevant ethics committee and the state the reference number for their judgement.

Research approved by institutional ethical regulations and

**Table 1**

Cases reported in literature with monophasic course [8–15].

References	# of Cases	Relapse	# of lesions on MRI	Oligoclonal Bands in CSF	Treatment	Follow-Up	Outcome
Chen	5	None	1 Pt with 9L 4 Pt 3–6 L	+1 Pt - 1 Pt,3NP	Not specified	1–3 years	100 % survival
Chen, Cj.	4	None	3-5 Lesions in 4 cases	NP	Not specified	2–23 months	100 % survival
Karaaslan E.	5	None	1 Pt with 9L 4 Pt 3–6 L	+1 Pt –2 Pt 2NP	MTP 1000 mg 7–10 days	6–47 months	100 % survival
Gu JI	3	None	1 Pt 3–5L 2 Pt with several lesions	+1 Pt – 2 Pt	DXM 15 mg/15 days, 1 Pt brain abscess	1 month-3 years	67 % survival, 1 Pt died lung infection
Khiat A.	2	None	1 Pt with 1L 1 Pt with several lesions	Not specified	MTP 1000 mg 7–10 days DXM 4 mg QID	21 days - 1 month	100 % survival with nil deficit
Chaodong W.	7	3	2 Pt with 2L 2 Pt with 5L 3 Pt with 3L	Nos specified	DXM 20–30 mg/7 days-1 month or MTP 1000 mg 5 days and 2–3 weeks	4–13.5 years	100 % survival with nil or mild deficit
Wallner-Blazek	10	1	3 Pt with several lesions 7 Pt with 1L	Not specified	High-dose steroids (doses and days of treatment not specified)	0–2 years	100 % survival and 83 % with nil or mild deficit
Agarwal, M [16]	3	1	2 Pt with 1L 1 Pt with 3L	- 2 Pt 1 Pt not specified	High dose steroids (not specified); biopsy in 2 pt diagnosis and treatment/ followed by steroids	3–4 years	100 % survival with moderate neurological deficit
Martinez H.	2	None	2 Pt with 1L	+1 Pt (1 band) –1 Pt	MTP	8 months-11 years	100 % survival with no neurological deficit

CSF: Cerebrospinal Fluid. L: Lesions. MRI: Magnetic Resonance Imaging. NP: Not Presented. Pt: Patient. L: Lesions. MTP: methylprednisolone, DXM: dexamethasone.

performed in accordance to institutional, national (Mexican Law 17 in Human Matter) and International (Helsinki declaration).

### Consent

Studies on patients or volunteers require ethics committee approval and fully informed written consent which should be documented in the paper.

Authors must obtain written and signed consent to publish a case report from the patient (or, where applicable, the patient's guardian or next of kin) prior to submission. We ask Authors to confirm as part of the submission process that such consent has been obtained, and the manuscript must include a statement to this effect in a consent section at the end of the manuscript, as follows: "Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request".

Patients have a right to privacy. Patients' and volunteers' names, initials, or hospital numbers should not be used. Images of patients or volunteers should not be used unless the information is essential for scientific purposes and explicit permission has been given as part of the consent. If such consent is made subject to any conditions, the Editor in Chief must be made aware of all such conditions.

Even where consent has been given, identifying details should be omitted if they are not essential. If identifying characteristics are altered to protect anonymity, such as in genetic pedigrees, authors should provide assurance that alterations do not distort scientific meaning and editors should so note.

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

### Author contribution

Please specify the contribution of each author to the paper, e.g. study concept or design, data collection, data analysis or interpretation, writing the paper, others, who have contributed in other ways should be listed as contributors. HRM: Case management, drafting, writing, editing. CIRG: Case management, writing JE: Case management, writing, editing JAFS: Writing, editing ACG: Writing, editing DEHG: Writing, editing.

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ISRCTN or numerous other registries.

No experimental treatment nor procedure.

### Guarantor

The Guarantor is the one or more people who accept full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

Hector R. Martinez.

### Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

### Appendix A. Supplementary data

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.amsu.2021.102602>.

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