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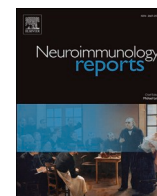
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Management of Baló Concentric Sclerosis with rituximab: A case study with long-term follow-up

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ABSTRACT

Background: Baló concentric sclerosis (BCS) is a rare demyelinating disorder that overlaps with other demyelinating diseases. BCS usually present in the context of multiple sclerosis (MS) or preceding typical MS. It rarely presents as an isolated lesion. IV methyl prednisolone (IVMP) is the mainstay in the treatment with various outcomes. Maintenance therapy is still not clearly defined.

Case presentation/case report: We present a case of isolated BCS that responded clinically and radiologically to long-term rituximab therapy.

Conclusion: A definite guideline for treating patient with BCS either with MS or as isolated entity is still controversial. Our case reflects a remarkable response to rituximab in achieving clinical and radiological long-term remission.

Introduction

BCS is a rare demyelinating disorder, pathologically characterized by large concentric lesions with circumferential rings of demyelination alternating with rings of relatively preserved myelin ("onion bulb" appearance) (Lucchinetti et al., 2000). It has often been considered as uncommon severe variant of multiple sclerosis (MS) (Lucchinetti et al., 2000, Popescu and Lucchinetti, 2012). Other hypothesis about BCS neuropathology suggest possible tissue preconditioning and hypoxia-like injury (Stadelmann et al., 2005). Features that makes BCS a distinct entity from MS are disease course, which is often monophasic and self-limiting, CSF profile, significantly lower frequencies of CSF-restricted oligoclonal bands (OCB) and disease severity that is often fatal at first occurrence (Hypoxia-like tissue injury and glial response contribute to Baló concentric lesion development | Neurology [Internet] 2022) (Ayrygnac et al., 2020 Jul 1). In this article we report a case of BCS managed successfully with Rituximab.

Case presentation

A 43-year-old female presented with two-week history of progressive onset of right-sided weakness and numbness, more prominent in the leg. Recently she started having memory problems and impaired computer

skills. No headache or fever. Medical history is remarkable for hypertension, depression, diabetes mellitus, asthma, and obesity. Examination revealed a conscious alert patient with impairment of recent memory, comprehension, and executive skills, right-sided upper motor neuron facial paresis with right hemiparesis, associated hyperreflexia, extensor plantar (Babinski) response and reduced sensitivity to pinprick and light touch on the right side. EDSS was 6.0. MRI revealed characteristic BCS appearance (Fig. 1A). There was no spinal cord involvement. CSF analysis revealed slightly increased protein 0.48 g/L in the absence of pleocytosis. Isoelectric focusing and immunofixation for oligoclonal bands (OCBs) in CSF and serum showed equivocal results which is defined by our lab of having less than 3 bands simultaneous in CSF and serum. Autoimmune screening was negative. Based on these findings the diagnosis of BCS was made. Therefore, 1 g of methylprednisolone for 5 days was administered. However, she still has incomplete recovery, persistent cognitive impairment. Maintenance therapy with off-label rituximab (two 1 g infusions 15 days apart followed by 1 g q6 monthly for 3 years) was initiated. Over the follow up period, she improved clinically (EDSS 2) and Radiologically (Fig. 1B).

Discussion

Baló's concentric sclerosis (BCS) is a form of pseudotumoral

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demyelination lesions. Patients present with mass effect symptoms similar to patients with tumefactive demyelination lesions (Hoang et al., 2021). Similarly, BCS can occur as a monophasic isolated lesion, in the context of MS. Large lesions are rare but smaller lesions (Baló-like lesions) may occur in MS as part of BCS spectrum. (Bhargava et al., 2015) Cases of BCS and tumefactive lesions were described together which may be referred to as an overlap, at least radiologically (Vakroukou et al., 2022).

BCS diagnosis is made depending on MRI findings of a concentric or lamellar lesion, yet many patients are unnecessarily biopsied (Arenas Vargas et al., 2020). CSF is usually bland, OCBs are uncommon compared with MS (BCS = 35%; MS = 98%) which support considering it a separate entity. (Jarius et al., 2018)

BCS has been considered to exhibit either a monophasic severe course or typical relapsing-remitting MS (RRMS) course as well. Clinical overlap among BCS, MS and RRMS was described in the literature (Aygnac et al., 2020, Hardy et al., 2016).

Maintenance therapy in BCS was described in the literature, medications such as interferon beta-1a, mitoxantrone, natalizumab and

fingolimod have been used. Therapeutic benefit over a short follow up period was more prominent when there was BCS-MS overlap (Hardy et al., 2015, Capello and Mancardi, 2004). Alemtuzumab treatment was associated with one death, which emphasize the hypothesis that the humoral responses are not prominent in BCS lesions, where innate immunity plays a central role with lipid-laden macrophages and giant multiple nuclei astrocytes predominance. (Brown et al., 2013) The role of PLEX is controversial, there are reports about using it in patients responding poorly to corticosteroids and as a rescue therapy in more typical tumefactive lesions. (Hardy et al., 2015, Sekijima et al., 1997). To the best of our knowledge, there are no data reporting the use of rituximab in isolated BCS. Tzanetakos et al. conducted a retrospective study describing eight cases of BCS, two patients were treated with rituximab as a maintenance therapy, both had MS-like lesion as well. The first case was BCS and tumefactive lesion, responding with effective regression of the lesion on MRI, the patient was followed for 16 months without any disease activity. The second case had spinal cord lesions and was followed for 15 months and showed good response to rituximab. Among the reported cases, one case is like our patient and was managed

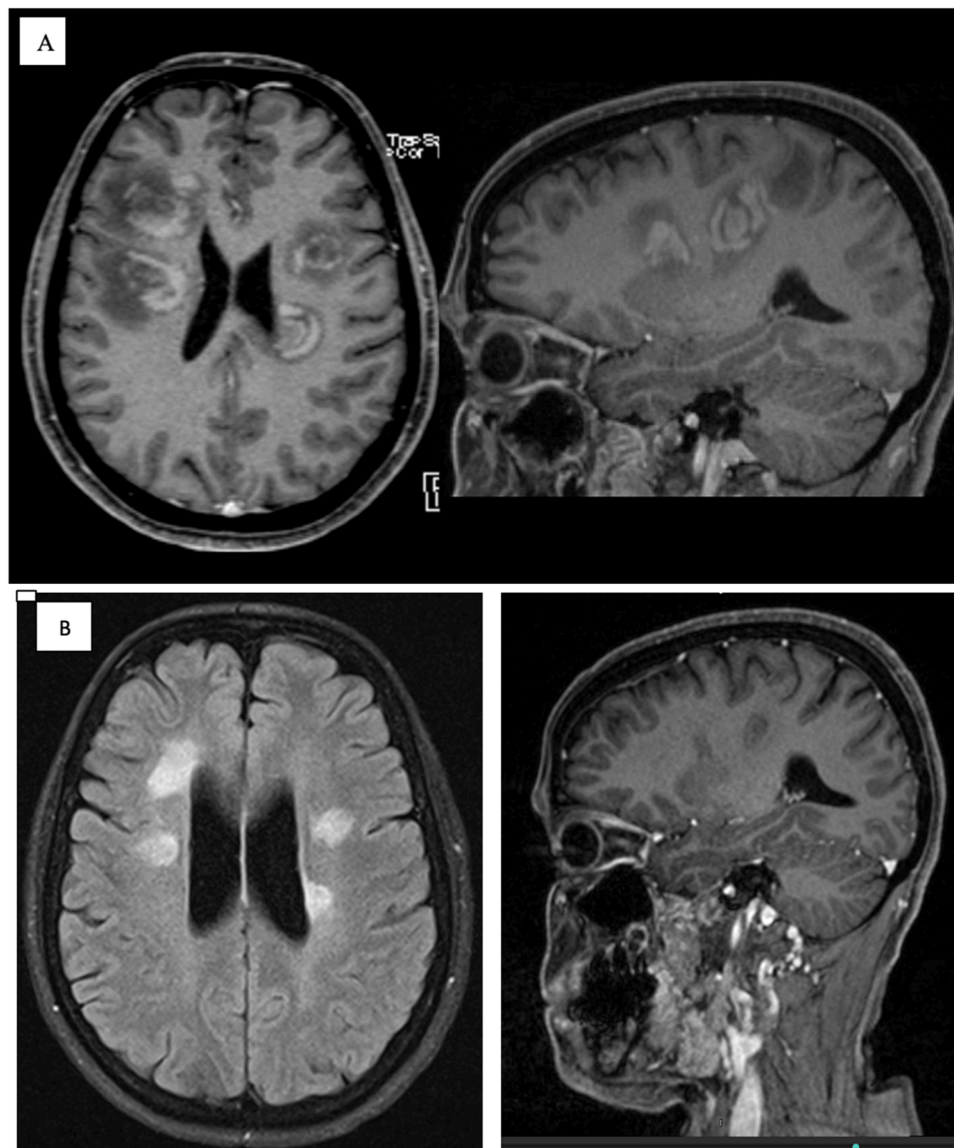


Fig. 1. 1A MRI at admission (January 2019): At admission (January 2019), Multifocal subcortical and periventricular lesions with irregular, ring like enhancement. 1B Follow up study (December 2022), Stable MR appearance of the previously described BCS lesions, no more signs of disease activity or newly developed lesions seen.

with IVMP initially with poor response, and subsequently responded to maintenance therapy with cyclophosphamide. However, In our case the patient had isolated BCS and no spinal involvement ([Heterogeneity of Baló's concentric sclerosis: a study of eight cases with different therapeutic concepts - PMC \[Internet\] 2022](#)).

Conclusion

Although long-term therapy for BCS was described in the literature, most cases had MS-like lesions along with the diagnosis of BCS. A definite guideline for treating patient with BCS either with MS or as isolated entity is still controversial. Our case reflects a remarkable response to rituximab in achieving disease regression clinically and radiologically and achieving long-term remission.

Statement of ethics

Consent was obtained from the patients.
Case approved by HMC Medical Research center

Authors contribution

All authors contributed equally to writing the manuscript.

Declaration of Competing Interest

The authors report no conflicts of interest.

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Supplementary materials

Supplementary material associated with this article can be found, in the online version, at [doi:10.1016/j.nerep.2023.100177](https://doi.org/10.1016/j.nerep.2023.100177).

References

- Lucchinetti, C, Brück, W, Parisi, J, Scheithauer, B, Rodríguez, M, Lassmann, H., 2000. Heterogeneity of multiple sclerosis lesions: implications for the pathogenesis of demyelination. *Ann. Neurol.* 47 (6), 707–717. Jun.
- Popescu, BFG, Lucchinetti, CF., 2012. Pathology of demyelinating diseases. *Annu. Rev. Pathol.* 7, 185–217.
- Stadelmann, C, Ludwin, S, Tabira, T, Guseo, A, Lucchinetti, CF, Leel-Ossy, L, et al., 2005. Tissue preconditioning may explain concentric lesions in Baló's type of multiple sclerosis. *Brain J. Neurol.* 128 (Pt 5), 979–987. May.
- Hypoxia-like tissue injury and glial response contribute to Balo concentric lesion development | *Neurology [Internet]*. [cited 2022 Jul 31]. Available from: <https://n.neurology.org/content/87/19/2000.short>.
- Ayrignac, X, Letourneau-Guillon, L, Carra-Dallière, C, Duquette, P, Girard, M, Poirier, J, et al., 2020. From Baló's concentric sclerosis to multiple sclerosis: a series of 6 patients. *Mult. Scler. Relat. Disord.* 42, 102078. Jul 1.
- Hoang, VT, Trinh, CT, Van, HAT, Nguyen, TTT, Chansomphou, V, Pham, NTT, et al., 2021. Baló's concentric sclerosis mimicking tumor on magnetic resonance imaging in a young patient. *Clin. Med. Insights Case Rep.* 14, 1179547621989673. Jan 19.
- Bhargava, A, Pujar, GS, Banakar, BF, Shubhkaran, K, Hemant, J., 2015. Recurrent tumefactive demyelination: an unusual presentation. *J. Pediatr. Neurosci.* 10 (1), 55. Mar.
- Vakrakou, AG, Brinia, ME, Svolaki, I, Argyrakos, T, Stefanis, L, Kilidireas, C., 2022. Immunopathology of tumefactive demyelinating lesions-from idiopathic to drug-related cases. *Front. Neurol.* 13, 868525. Mar 15.
- Arenas Vargas, LE, Bedoya Morales, AM, Rincón Carreño, C, Espitia Segura, OM, Penagos, N, 2020. Baló's concentric sclerosis: an atypical demyelinating disease in pediatrics. *Mult. Scler. Relat. Disord.* 44, 102198. Sep 1.
- Jarius, S, Würthwein, C, Behrens, JR, Wanner, J, Haas, J, Paul, F, et al., 2018. Baló's concentric sclerosis is immunologically distinct from multiple sclerosis: results from retrospective analysis of almost 150 lumbar punctures. *J. Neuroinflamm.* 15 (1), 22. Jan 18.
- Hardy, TA, Reddel, SW, Barnett, MH, Palace, J, Lucchinetti, CF, Weinshenker, BG., 2016. Atypical inflammatory demyelinating syndromes of the CNS. *Lancet Neurol.* 15 (9), 967–981. Aug.
- Hardy, TA, Beadnall, HN, Sutton, IJ, Mohamed, A, Jonker, BP, Buckland, ME, et al., 2015. Baló's concentric sclerosis and tumefactive demyelination: a shared immunopathogenesis? *J. Neurol. Sci.* 348 (1–2), 279–281. Jan 15.
- Capello, E, Mancardi, GL., 2004. Marburg type and Baló's concentric sclerosis: rare and acute variants of multiple sclerosis. *Neurol. Sci. Off. J. Ital. Neurol. Soc. Ital. Soc. Clin. Neurophysiol.* 25 (4), S361–S363. NovSuppl.
- Brown, JW, Coles, AJ, Jones, JL., 2013. First use of alemtuzumab in Baló's concentric sclerosis: a case report. *Mult. Scler. Houndmills Basingstoke Engl.* 19 (12), 1673–1675. Oct.
- Sekijima, Y, Tokuda, T, Hashimoto, T, Koh, CS, Shoji, S, Yanagisawa, N., 1997. Serial magnetic resonance imaging (MRI) study of a patient with Baló's concentric sclerosis treated with immunoadsorption plasmapheresis. *Mult. Scler. Houndmills Basingstoke Engl.* 2 (6), 291–294. Jan.
- Heterogeneity of Baló's concentric sclerosis: a study of eight cases with different therapeutic concepts - *PMC [Internet]*. [cited 2022 Jul 31]. Available from: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7604966/>.