

# Baló's concentric sclerosis: imaging findings and pathological correlation

Maxine Darke<sup>1\*</sup>, Farshad M Bahador<sup>2</sup>, Douglas C Miller<sup>3</sup>, Norman S Litofsky<sup>4</sup>, Humera Ahsan<sup>2</sup>

1. School of Medicine, University of Missouri, Columbia, MO, USA

2. Department of Radiology, University of Missouri Health Care, Columbia, MO, USA

3. Department of Pathology, University of Missouri Health Care, Columbia, MO, USA

4. Department of Neurosurgery, University of Missouri Health Care, Columbia, MO, USA

\* **Correspondence:** Maxine Darke, 803 East Green Meadows Road #204, Columbia, MO, 65201, USA  
(✉ [med9qc@mail.missouri.edu](mailto:med9qc@mail.missouri.edu))

Radiology Case. 2013 Jun; 7(6):1-8 :: DOI: 10.3941/jrcr.v7i6.1251

## ABSTRACT

Baló's concentric sclerosis is a primary inflammatory central nervous system demyelinating disease that is considered a rare, radiographically and pathologically distinct variant of multiple sclerosis. Baló's concentric sclerosis is characterized by alternating rings of demyelinated and myelinated axons, and it is most frequently diagnosed postmortem by autopsy or, more recently, by magnetic resonance imaging without pathologic verification. This report is of a case of Baló's concentric sclerosis in which the patient presented with left-sided focal sensorimotor deficits. The patient's lesion demonstrated characteristics of Baló's concentric sclerosis by magnetic resonance imaging, but since a neoplastic process was also suspected initially, the patient underwent a surgical biopsy. This pathology sample now provides the opportunity to correlate the tissue diagnosis of demyelination with characteristic magnetic resonance imaging findings; this comparison is infrequently found in the literature.

## CASE REPORT

### CASE REPORT

A 55 year-old right-handed African American male presented to his primary care physician because of the new onset of a headache, left sided numbness, weakness, and clumsiness. He reported going to sleep with a headache and waking up the next day with left arm and leg numbness as well as left hand weakness and clumsiness. These symptoms persisted for several days, so the patient decided to seek care. The patient's primary care provider then referred him to our institution for further work-up. The patient presented to our institution 10 days following his symptom onset and reported that he still had some numbness and tingling on the left side of his body as well as left hand weakness and clumsiness. Physical examination at that time revealed left pronator drift, left Hoffmann's sign, decreased muscle strength in the left upper and lower extremities, and increased reflexes in the left upper and lower extremities. Sensation was intact bilaterally. The patient reported having a separate but similar episode of

left sided weakness and numbness that occurred 3-4 months previously and that lasted for 3 days before spontaneously resolving.

The patient's MRI showed a round lesion with concentric ring enhancement in the right precentral gyrus with surrounding edema and no mass effect. The concentric rings were hypointense and isointense on T1-weighted images, and the hypointense regions showed complete enhancement after contrast administration (Images 1-2 and 3-4, respectively). The concentric rings were hyperintense and isointense on T2-weighted images (Images 5 and 6). The 1.9 x 1.6 x 1.1 cm lesion also showed restricted diffusion (Images 7 and 8). The location of this mass in the right precentral gyrus/primary motor cortex correlated with the patient's symptoms. A few tiny T2 hyperintense foci were also identified in the periventricular region perpendicular to the long axis of the ventricles (Image 9).

Following imaging, the differential diagnosis included a demyelinating lesion or a neoplastic process. For further evaluation, the patient underwent a right frontoparietal craniotomy, and no solid tumor was identified. An intra-operative ultrasound was used to localize the mass and guide a tissue biopsy. Frozen specimens were diagnostic of a demyelinating lesion rather than a neoplasm, and this diagnosis was confirmed in the final pathology examination of fixed tissues.

Following the pathology report, cervical and thoracic spine MRIs were subsequently performed; these images showed non-enhancing plaques in the cervical region (Image 10). CSF analysis was not done due to recent neurosurgery, but the diagnosis of multiple sclerosis (MS), and specifically of Baló's concentric sclerosis, was strongly suggested based on the clinical history, pathology report, and imaging findings. Thus, the patient was prescribed a 3-day course of methylprednisolone 500 mg IV BID (twice daily). He showed improvement in his left sided muscle strength during his 5-day hospital stay with this treatment.

The patient returned for a follow-up appointment 2 months later at which time he had regained considerable strength on his left side. During this clinic visit, the patient recalled having several episodes of left sided weakness and numbness in the past; these episodes typically lasted several weeks before resolving spontaneously. He was diagnosed with MS and started on Interferon beta-1a. The patient has been doing well on Interferon beta-1a and has not reported any exacerbations of MS as of his visit in the neurology clinic 24 months after his hospitalization. The patient opted to not perform any follow up imaging studies despite phone calls from neurosurgery and radiology.

## DISCUSSION

Baló's concentric sclerosis (BCS) is a rare entity that is characterized by alternating bands of myelinated and partially demyelinated axons. It is hypothesized that these bands either represent remyelination of previously demyelinated regions or early stages of acute demyelination. These rings are thought to develop from an initial core region of demyelination around which a protective inflammatory ring develops in an attempt to isolate and prevent further expansion of demyelination. The central core eventually breaks through, but the ring is spared from damage, leaving a myelinated band between the areas of demyelination. Rings continue to form in this manner [1, 2].

BCS can present clinically as a single manifestation or any combination of neurological symptoms including headache, numbness, weakness, seizures, aphasia, and cognitive or behavior dysfunction. This variety of symptoms can be attributed to the numerous locations that BCS lesions can be found; potential sites include the cerebral hemispheres, cerebellum, brain stem, spinal cord, and optic chiasm. Roughly 85 cases of BCS have been reported in the literature, and most of those were reported in relatively young males, ranging from 4 to 56 years old [1, 3].

The clinical course of BCS can be classified into the following types: a single and self-limited event, relapsing-remitting, and primary progressive. Early reports indicated that the clinical course was often primary progressive with death coming in weeks to months, but more recent reports show extended survival, spontaneous remission, and asymptomatic cases. Improved outcomes are partially due to MRI detection and early treatment with corticosteroids [4].

MRI is often used to diagnose BCS, and irregular, concentric rings are thought to be pathognomonic for the disease. These concentric rings represent alternating areas of demyelinated and myelinated axons, shown by characteristic intensities on various MRI sequences. Acute disease activity involving demyelination, gliosis, and perivascular lymphocytic infiltration is hyperintense on T2-weighted images; these hyperintense areas alternate with isointense rings, which represent areas of preserved myelination. Isointense rings on T1-weighted images also correspond to areas of myelination, but demyelinated zones appear as hypointense rings. Moreover, post-gadolinium contrast T1-weight images demonstrate concentric ring enhancement at sites of increased blood brain barrier permeability and inflammation responsible for demyelination [5, 6].

Acute BCS lesions have also been reported to have a characteristic MR spectroscopy pattern of decreased N-acetylaspartate peak (reflecting neuronal loss), elevated lactate peak (suggesting impaired aerobic metabolism), and elevated choline and lipid peaks (consistent with increased lipid membrane turnover, inflammation, and/or gliosis) [7, 8].

Diffusion-weighted imaging (DWI) along with diffusion coefficient maps can help to confirm the presence of BCS. Restricted diffusion, which is shown as high signal intensity on DWI, has been described in areas of active demyelination, while areas of facilitated diffusion are represented by low signal intensity. Thus, BCS lesions demonstrate alternating rings of high and low signal intensity, representing demyelinated and myelinated areas, respectively. Sequential imaging has shown that restricted diffusion is initially present around the central core of the lesion and then spreads outwardly, which supports the theory that acute demyelination moves concentrically from the central core. The diffusion coefficient map can further confirm the presence of restricted diffusion, which is demonstrated as low signal intensity, while relatively unrestricted diffusion is shown as high signal intensity. DWI typically shows fewer rings compared to T2-weighted images because of lower spatial resolution [2, 7].

The concentric rings visualized on imaging correlate with the alternating rings of myelin preservation or remyelination and demyelination demonstrated in tissue specimens, especially with the assistance of myelin stains. This pattern is demonstrable in autopsy specimens, but is unlikely to be appreciable in biopsies which will usually lack large samples of the entire lesions, as was the case for our patient's biopsy. A standard myelin stain, Luxol Fast Blue (LFB), shows intact myelin as blue whereas demyelinated areas appear unstained, or, when combined with Hematoxylin and Eosin, pink. Thus, the classic autopsy pathology specimens of BCS lesions

consist of alternating rings of blue (normal well-myelinated white matter) and pink (hypercellular, demyelinated areas). When the biopsy is done within about two weeks of the onset of demyelination, myelin debris can also be visualized as LFB-blue spots within macrophages. Bielschowsky silver stains can be used to demonstrate relative preservation of axons [8, 9]. The demyelinated (pink) bands in the tissue section can be correlated with the hyperintense areas on T2-weighted images, the hypointense rings on T1-weighted images, the areas of enhancement on the post-gadolinium contrast T1-weight images, and the rings of restricted diffusion or high signal intensity on DWI. Further, these areas of demyelination represent active disease.

Sequential imaging has shown that BCS lesions lose their characteristic ring appearance over variable durations of time and often coalesce into typical demyelinated plaque-like MS lesions [7]. Some concentric patterns are no longer evident on imaging seven weeks after symptom onset while some are still present over a year later [10].

The differential diagnosis for a suspected BCS lesion includes acute disseminated encephalomyelitis (ADEM), "ordinary" MS, and a primary neoplasm [4]. In this case a primary neoplasm was initially suspected. Depending on the type of brain tumor, CT and MRI findings vary significantly. The most common primary tumors of concern here are high grade gliomas. MRI of such tumors will typically show an enhancing mass around which there is surrounding abnormal signal, hypo- or iso-intense on T1-weighted images and bright on T2-weighted (and FLAIR) images. This abnormal signal around the enhancing mass is often regarded as "edema" but for gliomas can represent brain tissue invaded by tumor cells, whereas with metastatic tumors (such as carcinomas and melanomas), it usually represents edematous brain without tumor cells [11].

MRI is more useful than CT to identify demyelinating lesions, including BCS, MS, and ADEM. With MRI, acute or active MS lesions are usually smaller and more sharply demarcated than BCS or ADEM lesions, and ADEM lesions are more extensive. ADEM lesions appear as increased signal intensity on T2-weighted MRI and decreased signal intensity on T1-weighted MRI; some lesions may enhance after contrast administration. MS and ADEM can look very similar by MRI, but serial studies can be useful to distinguish them because findings of new lesions would suggest MS. Post-contrast images in MS also show a mixture of enhancing and non-enhancing lesions due to the temporal dissemination in MS. Similar to BCS, active lesions in both MS and ADEM can demonstrate restricted diffusion (high signal intensity) on DWI [4, 12, 13].

CT and CSF findings can serve as adjuncts to MRI. CT scans in ADEM are often normal at onset and abnormal 5-14 days later, showing low attenuation, multifocal lesions in the subcortical white matter. CT findings in MS and BCS are variable; some patients have normal CT scans and some show low attenuation lesions [13]. CSF studies in BCS typically reveal a mononuclear inflammatory reaction and occasional oligoclonal bands compared to more numerous oligoclonal

bands typically found in the CSF of classical MS patients. CSF findings in ADEM commonly show pleocytosis and elevated protein levels [4].

Similar to the findings in this case report, other authors have described BCS lesions along with more typical MS lesions [3, 6, 14]. However, BCS lesions have been shown to disappear or shrink on follow-up MR imaging while MS lesions typically do not [12]. Follow-up imaging has not yet been done for the patient in this case report as he decided not to perform any follow-up imaging, but the patient is doing well without clinical evidence of recurrent disease after 2 years.

Early diagnosis with characteristic MRI findings for BCS is important because patients have responded favorably to treatment with corticosteroids [4]. Since tissue biopsy is not routinely used to diagnose BCS, this case report provides a rare opportunity to correlate pathology findings to MR images.

#### TEACHING POINT

Baló's concentric sclerosis (BCS) is a rare, pathologically and radiographically distinct variant of multiple sclerosis, and it is characterized by alternating rings of demyelinated and myelinated axons. The myelinated rings correlate with the isointense areas on T2- and T1-weighted images and low signal intensity on DWI; these areas alternate with the demyelinated rings, which appear as hyperintense areas on T2-weighted images, hypointense rings on T1-weighted images, areas of enhancement on post-gadolinium contrast T1-weight images, and rings of restricted diffusion (high signal intensity) on DWI.

#### REFERENCES

1. Karaarslan E, Altintas A, Senol U, et al. Baló's concentric sclerosis: clinical and radiologic features of five cases. *AJNR Am J Neuroradiol*. 2001 Aug;22(7):1362-7. PMID: 11498428
2. Kavanaah EC, Heran MK, Fenton DM, Lapointe JS, Nugent RA, Graeb DA. Diffusion-weighted imaging findings in Balo concentric sclerosis. *Br J Radiol*. 2006 Jul;79(943):e28-31. PMID: 16823051
3. Li Y, Xie P, Fan X, Tang H. Baló's concentric sclerosis presenting with benign clinical course and multiple sclerosis-like lesions on magnetic resonance images. *Neurol India*. 2009 Jan-Feb;57(1):66-8. PMID: 19305082
4. Mowry, EM, Woo JH, Ances BM. Baló's concentric sclerosis presenting as a stroke-like syndrome. *Nat Clin Pract Neurol*. 2007 Jun;3(6):349-54. PMID: 17549061
5. Caracciolo JT, Murtagh RD, Rojiani AM, Murtagh FR. Pathognomonic MR imaging findings in Balo concentric

sclerosis. *American Journal of Neuroradiology*. 2001 Feb;22(2):292-293. PMID: 11156771

6. Wengert O, Siebert E. Baló's concentric sclerosis. *N Engl J Med*. 2011 Aug;365(8):742. PMID: 21864168

7. Mowry, EM, Woo JH, Ances BM. Technology insight: can neuroimaging provide insights into the role of ischemia in Baló's concentric sclerosis? *Nat Clin Pract Neurol*. 2007 Jun;3(6):341-8. PMID: 17549060

8. Kim MO, Lee SA, Choi CG, Huh JR, Lee MC. Baló's concentric sclerosis: a clinical case study of brain MRI, biopsy, and proton magnetic resonance spectroscopic findings. *Journal of Neurology, Neurosurgery, and Psychiatry*. 1997 Jun;62(6):655-8. PMID: 9219760

9. Bitsch A, Bruhn H, Vougioukas V, et al. Inflammatory CNS demyelination: histopathologic correlation with in vivo quantitative proton MR spectroscopy. *American Journal of Neuroradiology*. 1999 Oct;20(9):1619-1627. PMID: 10543631

10. Ng SH, Ko SF, Cheung YC, Wong HF, Wan YL. MRI features of Baló's concentric sclerosis. *The British Journal of Radiology*. 1999 Apr;72 (856):400-403. PMID: 10474505

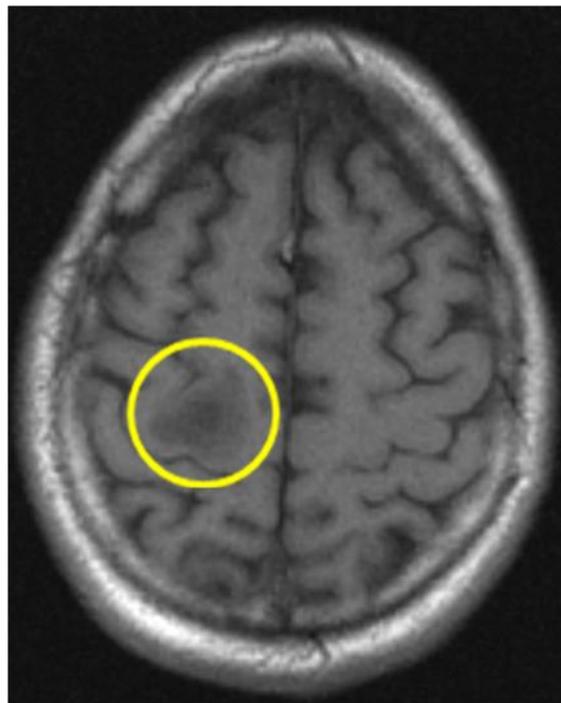
11. Popescu BF, Lucchinetti CF. Pathology of demyelinating diseases. *Annu Rev Pathol*. 2012 Feb;7:185-217. PMID: 22313379

12. Given CA, Stevens BS, Lee C. The MRI appearance of tumefactive demyelinating lesions. *AJR Am J Roentgenol*. 2004 Jan;182(1):195-9. PMID: 14684539

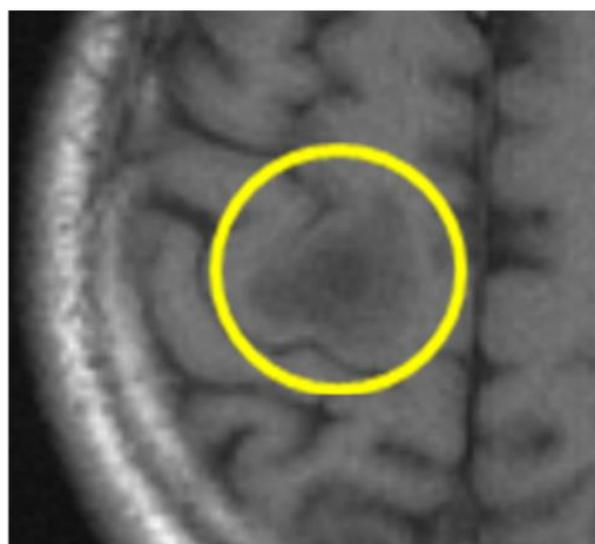
13. Garg RK. Acute disseminated encephalomyelitis. *Postgrad Med J*. 2003 Jan;79(927):11-7. PMID: 12566545

14. Kishimoto R, Yabe I, Niino M, et al. Baló's concentric sclerosis-like lesion in the brainstem of a multiple sclerosis patient. *J Neurol*. 2008 May;255(5):760-1. Epub 2008 Feb 25. PMID: 18293025

## FIGURES



**Figure 1:** 55 year-old male with Baló's concentric sclerosis. This axial T1-weighted MR image shows a non-uniform, round mass (circled) with irregular alternating bands of hypointense and isointense signal, representing demyelinated and myelinated areas, respectively. There is surrounding edema and no mass effect. The lesion measures 1.9 cm x 1.6 cm x 1.1 cm in AP, transverse, and craniocaudal directions. This mass is located in the right precentral gyrus, corresponding to the region of the motor strip and hand area. (Protocol: 1.5 tesla magnet strength, 5 mm slice thickness, TR: 468, TE: 12)

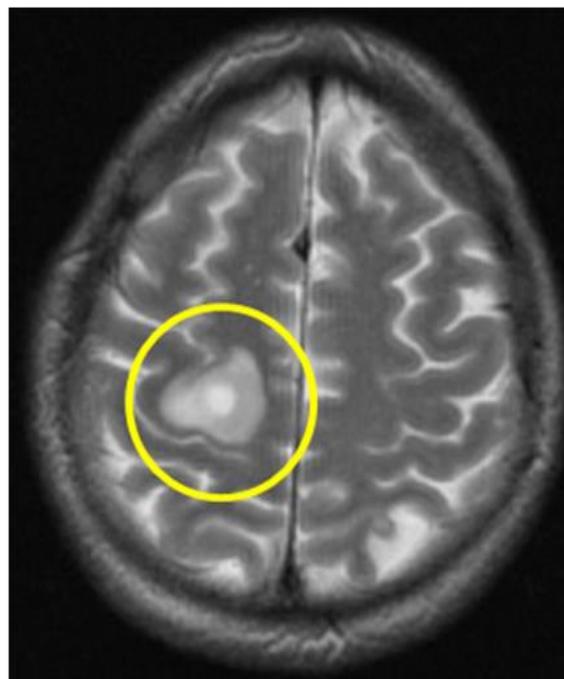
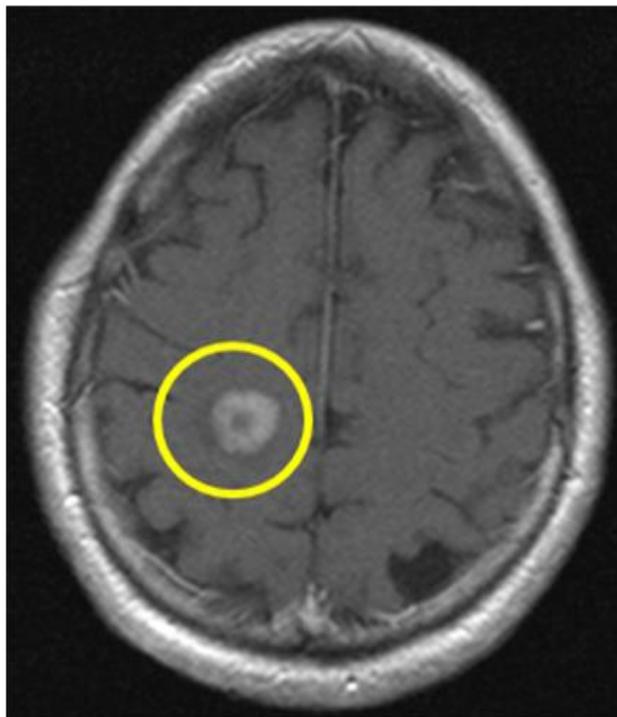


**Figure 2:** 55 year-old male with Baló's concentric sclerosis. This magnified view of the axial T1-weighted MR image shows a non-uniform, round mass (circled) with irregular alternating bands of hypointense and isointense signal, representing demyelinated and myelinated areas, respectively.

There is surrounding edema and no mass effect. The lesion measures 1.9 cm x 1.6 cm x 1.1 cm in AP, transverse, and craniocaudal directions. This mass is located in the right precentral gyrus, corresponding to the region of the motor strip and hand area. (Protocol: 1.5 tesla magnet strength, 5 mm slice thickness, TR: 468, TE: 12)

enhance after contrast administration. This enhancement is demonstrated by the hyperintense ring circled in this image. Areas of myelination remain isointense as they were in the non-contrast T1-weighted image. The lesion has surrounding edema but no mass effect.

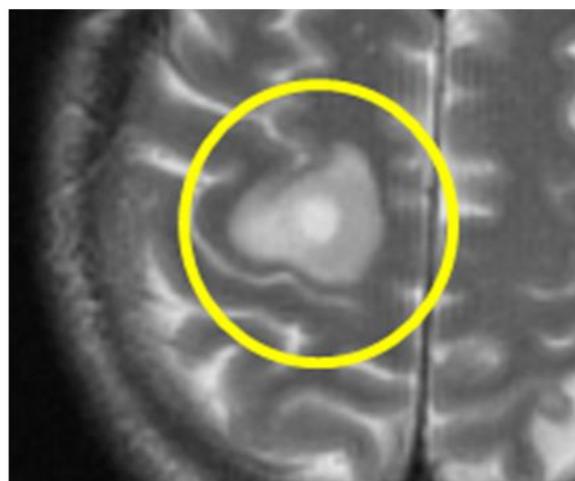
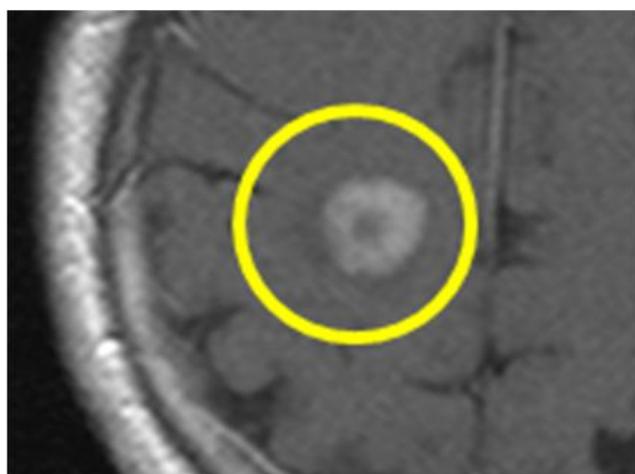
(Protocol: 1.5 tesla magnet strength, 5 mm slice thickness, TR: 657, TE: 12, 18.5 mL of gadolinium contrast given)



**Figure 3:** 55 year-old male with Baló's concentric sclerosis. This axial contrast enhanced T1-weighted MR image shows that the areas of demyelination enhance after contrast administration. This enhancement is demonstrated by the hyperintense ring circled in this image. Areas of myelination remain isointense as they were in the non-contrast T1-weighted image. The lesion has surrounding edema but no mass effect. (Protocol: 1.5 tesla magnet strength, 5 mm slice thickness, TR: 657, TE: 12, 18.5 mL of gadolinium contrast given)

**Figure 5:** 55 year-old male with Baló's concentric sclerosis. This axial T2-weighted MR image shows the non-uniform, round mass (circled) with alternating areas of isointense and hyperintense signals, corresponding to myelinated and demyelinated tissue, respectively. There is surrounding edema without mass effect.

(Protocol: 1.5 tesla magnet strength, 5 mm slice thickness, TR: 3300, TE: 97)



**Figure 4:** 55 year-old male with Baló's concentric sclerosis. This magnified view of the axial contrast enhanced T1-weighted MR image shows that the areas of demyelination

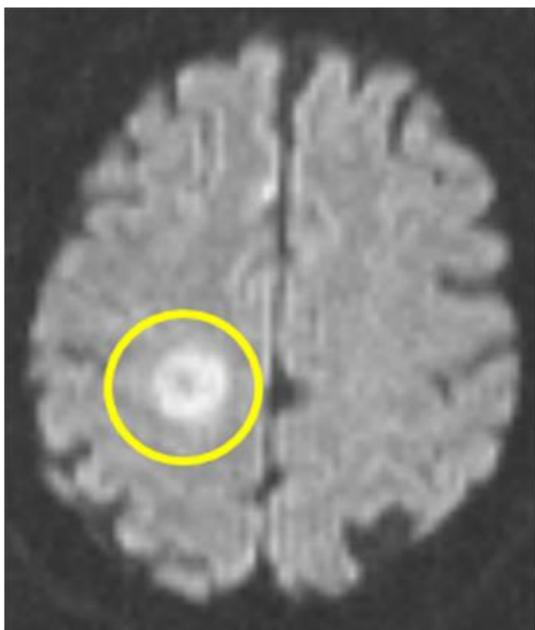
**Figure 6:** 55 year-old male with Baló's concentric sclerosis. This magnified view of the axial T2-weighted MR image shows the non-uniform, round mass (circled) with alternating

areas of isointense and hyperintense signals, corresponding to myelinated and demyelinated tissue, respectively. There is surrounding edema without mass effect.

(Protocol: 1.5 tesla magnet strength, 5 mm slice thickness, TR: 3300, TE: 97)

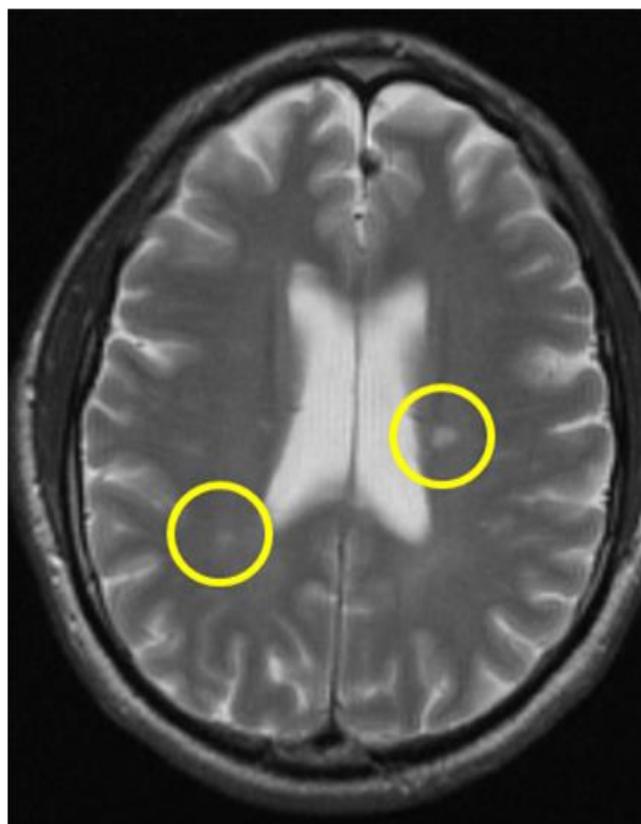
restricted diffusion, which is demonstrated as low signal intensity, in the intermediate ring of the mass (circled). Relatively unrestricted diffusion is shown as high signal intensity.

(Protocol: 1.5 tesla magnet strength, 5 mm slice thickness, TR: 3600, TE: 89)



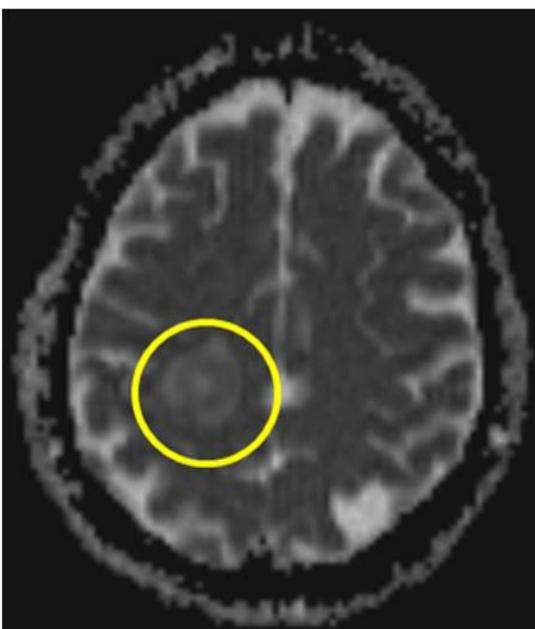
**Figure 7:** 55 year-old male with Baló's concentric sclerosis. This diffusion-weighted MR image shows the mass (circled) as a central core of low signal with high signal intensity in intermediate and outer rings. Restricted diffusion, which is shown as high signal intensity, is in areas of active demyelination while areas of facilitated diffusion are represented by low signal intensity.

(Protocol: 1.5 tesla magnet strength, 5 mm slice thickness, TR: 3600, TE: 89)

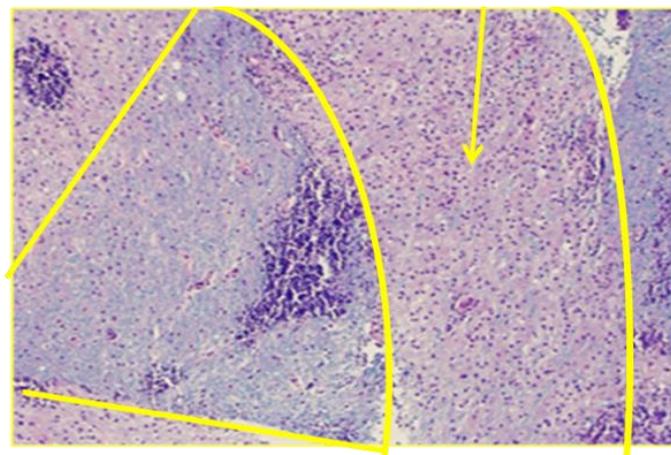


**Figure 9:** 55 year-old male with Baló's concentric sclerosis. This axial T2-weighted MR image highlights tiny, nonsymmetrical hyperintense foci (circled) that are seen in the periventricular and subcortical white matter and the septocallosal interface. These lesions represent areas of demyelination and support the diagnosis of multiple sclerosis.

(Protocol: 1.5 tesla magnet strength, 5 mm slice thickness, TR: 3300, TE: 97)



**Figure 8:** 55 year-old male with Baló's concentric sclerosis. This diffusion coefficient map further confirms the presence of



**Figure 10:** 55 year-old male with Baló's concentric sclerosis. This sagittal STIR MR image of the cervical spine shows two hyperintense foci that measure 6 mm and 5 mm in diameter (circled) and are located at the level of C2 and C5, respectively. These lesions also represent areas of demyelination and further support the diagnosis of multiple sclerosis.

(Protocol: 1.5 tesla magnet strength, 3 mm slice thickness, TR: 4230, TE: 42)

**Figure 11:** 55 year-old male with Baló's concentric sclerosis. The Luxol Fast Blue/H&E combination stain of the biopsy specimen shows the demyelinated lesion as a hypercellular focus (pink, arrow) bounded by less cellular normal myelinated white matter (stained blue, outlined by yellow lines). There is also Luxol Fast Blue positive debris (myelin fragments) in many of the histiocytes. There is gliosis both within the demyelinated areas and in the surrounding zones with preserved myelin.

<b>Etiology</b>	Demyelination of axons of the central nervous system
<b>Incidence</b>	Rare
<b>Gender ratio</b>	Present in males slightly more than females
<b>Age predilection</b>	Mostly in patients 4 to 56 years old
<b>Risk factors</b>	Male, 4-56 years old, others unknown
<b>Treatment</b>	Corticosteroids
<b>Prognosis</b>	Wide range from death within a few weeks to asymptomatic cases
<b>Findings on imaging</b>	Irregular, concentric rings

**Table 1:** Summary table for Baló's Concentric Sclerosis

	CT	MRI – T1	MRI – T1 post-contrast	MRI – T2	MRI – DWI
<b>Baló's concentric sclerosis</b>	Normal or may show low attenuation lesions	Alternating rings of demyelinated (hypointense) and myelinated (isointense) axons	Avid Concentric ring enhancement in demyelinated areas	Alternating rings of demyelinated (hyperintense) and myelinated (isointense) axons	Alternating rings of demyelinated (high signal intensity or restriction diffusion) and myelinated (low signal intensity) axons
<b>Acute disseminated encephalomyelitis</b>	Usually normal at onset and abnormal 5-14 days later; low attenuation, multifocal lesions in the subcortical white matter	More extensive hypointense lesions	Lesions may enhance	More extensive hyperintense lesions	Lesions may show restricted diffusion (high signal intensity)
<b>Multiple sclerosis</b>	Normal or may show low attenuation plaques	Smaller, more sharply demarcated hypointense lesions	Mixture of enhancing and non-enhancing lesions	Smaller, more sharply demarcated hyperintense lesions	Active plaques may show restricted diffusion (high signal intensity)
<b>Primary neoplasm</b>	Variable, mass effect often seen, single lesion common	Variable, but most show low or intermediate signal intensity	Variable	Variable, but most appear bright	Usually no restricted diffusion

**Table 2:** Differential diagnosis table for Baló's Concentric Sclerosis

**ABBREVIATIONS**

ADEM = Acute disseminated encephalomyelitis  
 BCS = Baló's concentric sclerosis  
 CSF = cerebral spinal fluid  
 DWI = diffusion-weighted imaging  
 MRI = magnetic resonance imaging  
 MS = multiple sclerosis

**KEYWORDS**

Baló's concentric sclerosis; demyelinating disease; MRI; Pathology

**Online access**

This publication is online available at:  
[www.radiologycases.com/index.php/radiologycases/article/view/1251](http://www.radiologycases.com/index.php/radiologycases/article/view/1251)

**Peer discussion**

Discuss this manuscript in our protected discussion forum at:  
[www.radiolopolis.com/forums/JRCR](http://www.radiolopolis.com/forums/JRCR)

**Interactivity**

This publication is available as an interactive article with scroll, window/level, magnify and more features.  
 Available online at [www.RadiologyCases.com](http://www.RadiologyCases.com)

Published by EduRad



[www.EduRad.org](http://www.EduRad.org)