Balo's Concentric Sclerosis – A Case Report

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ABSTRACT

Introduction: Balo's concentric sclerosis is a rare demyelinating disease that was first described by Joszef Balo in 1928. BCS and multiple sclerosis like lesions are being increasingly reported to occur simultaneously, indicating that they usually represent a continuation process of the same disease. It is a disease of the white matter of brain, in which white matter appears to be damaged in concentric layers. Balo sclerosis comes under the MS lesion type III pattern (distal oligodendrogliopathy). Our case report summarizes the pathology, clinical manifestations, imaging features and management strategies of BCS.

Case report: We report the case of a 45 year old male who developed unsteadiness of gait, weakness of both lower limbs, retention of urine and slurring of speech. His MRI showed multiple onion peel like demyelinating lesions over the subcortical areas of parietal, frontal and occipital lobes. Treatment given to the Patient was IV methyl prednisolone, and other supportive measures.

Conclusion: BCS may have varied clinical manifestations however; the pathognomonic MRI features help to clinch the diagnosis.

Keywords: Balo's concentric sclerosis, Multiple sclerosis, Atypical idiopathic inflammatory demyelinating lesions

INTRODUCTION

Balo's disease is also known as Balo disease, encephalitis periaxialis concentrica, leukoencephalitis periaxialis concentrica and concentric sclerosis. The term "concentric sclerosis" comes from a pattern of concentric (circular) areas of damaged myelin alternating with areas of relatively undamaged myelin in various parts of the brain and spinal cord. This pattern can be seen on magnetic resonance imaging (MRI). Balo's concentric sclerosis is a demyelinating disease similar to multiple sclerosis but with the particularity that the demyelinating tissues form concentric layers.1 Usually the clinical course of the disease is progressive, but a relapsing remitting course has been usually reported.²⁻⁶ Reported clinical manifestations are headache, aphasia, cognitive or behavioral dysfunction and or seizure. CSF studies often reveal a mononuclear inflammatory reaction and occasionally oligoclonal bands.7 It seems that the course gets better with prednisolone therapy8, although evidence of this is unreliable.

CASE REPORT

A 45 year old male was admitted in Tirunelveli Medical College Hospital with complaints of unsteadiness of gait for 4 months, weakness of both lower limbs for 15 days and retention of urine for 15 days. 4 months before, he had developed unsteadiness of gait and swaying to both sides while walking which does not worsen in dark and improved by treatment with steroids. Then he developed slurring of speech for 1 month with weakness of both lower limbs and painful acute retention of urine for 15 days which was gradually progressive. Patient had no history suggestive of sensory disturbance, higher mental function and no history of fever. On examination his pulse was normal and regular, his blood pressure was normal. His higher mental examination was normal. Examination of Cranial nerves was normal, and his fundus did not reveal any optic atrophy. His tones in both lower limbs were decreased, power was 2/5 for both hips and knee joints and was 0/5 for both ankle joints. On examination of reflexes, all deep tendon reflexes were present except ankle and plantar reflex, which was absent bilaterally. Further Investigations on complete hemogram, Renal function test, Liver function test and Serum electrolytes found normal. His ECG, Chest x-ray, USG abdomen and CSF analysis were normal. Patient was non reactive for HIV and VDRL was negative. CSF analysis showed no cells, protein 60 mg and glucose 65 mg. His MRI brain revealed multiple onion peel like demyelinating lesions over the subcortical areas of parietal, frontal and occipital lobes (figure:1-6).

DISCUSSION

Multiple sclerosis is disease of the central nervous system which is autoimmune. Concentric sclerosis or Balo's disease is an unusual variant which usually progress very rapidly. The Symptoms are usually headache, seizure, gradual paralysis, involuntary muscle spasm and loss of cognitive function.^{3-6,9-11} Other names for this condition include encephalitis periaxialis concentrica and leukoencephalitis periaxialis concentrica.12 MRI T1 weighted image shows low density lesions.T2 weighted image shows high signal intensity lesions. FLAIR image reveals edema surrounding the lesions. Postcontrast T1 weighted images show enhancement in a concentric ring¹¹ like fashion and hold the key to the diagnosis.MRI features will show hyperintense bands on T2 weighted images correspond the concentric bands of demyelination with gliosis and perivascular lymphocytic infiltration. The isointense bands will represent the white matter which is usually spared or it had undergone remyelination. T1 weighted post contrast images will reveal, all the lesions shows concentric rings of enhancement, indicating rings of active inflamed and spared white matter. The concentric pattern is observed only if the MRI is performed early in the course of the disease.13

BCS- Histologically it is characterized by alternating rings of myelin preservation or remyelination involving the cerebral

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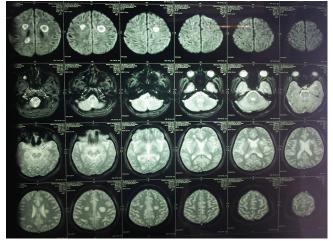


Figure-1: T2W sagittal shows concentric rings

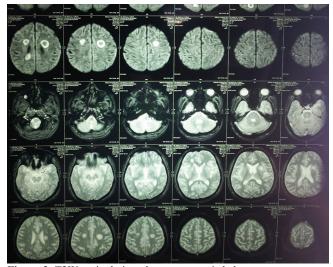


Figure-2: T2W sagittal view shows concentric halos



Figure-3: T2 coronal view shows concentric halos

hemisphere, brainstem, spinal cord and optic chiasma.^{10,14,15} Clinical DDS include acute demyelinating encephalomyelitis, stroke, neoplasm, infection and abscess.¹⁶ Clinical course is primary progressive but a relapsing – remitting course has been reported. This patient presented with clinical features and classical MRI findings.

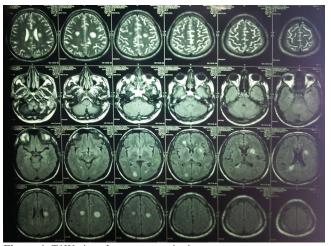


Figure-4: T1W view shows concentric rings

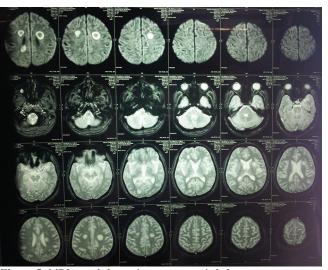


Figure-5: MRI reveals hypertintese concentric halos

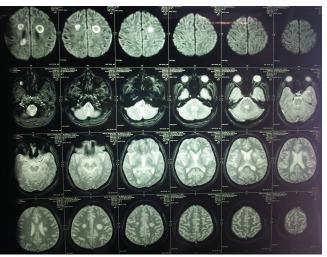


Figure-6: MRI reveals characteristics concentric halos

CONCLUSION

Patient's clinical features and MRI findings - concentric rings of enhancement, indicating rings of active inflamed and spared white matter are highly suggestive of Balo's concentric sclerosis. The clinical course of this disease is primarily progressive but relapsing and remitting course has also been reported. Our patient was diagnosed in early course of the disease, so he had the characteristic MRI findings and Patient was treated with IV methyl prednisolone and other supportive measures.

REFERENCES

- Balo J. Encephalitis periaxialis concentrica. Arch Neurol Psychiatry. 1928;19:242–244.
- Osborn AG: Demyelinating and inflammatory diseases; in: Osborn's Brain. Imaging, Pathology, and Anatomy. Mannitoba, Amirsys. 2013;405-442.
- Wallner-Blazek M, Rovira A, Fillipp M, et al: Atypical idiopathic inflammatory demyelinating lesions: prognostic implications and relation to multiple sclerosis. J Neurol. 2013;260:2016-2022.
- Koelblinger C, Fruehwald-Pallamar J, Kubin K, et al: Atypical idiopathic inflammatory demyelinating leions (IIDL): conventional and diffusion-weighted MR imaging (DWI) findings in 42 cases. Eur J Radiol. 2013;82:1996-2004.
- Wang C, Zhang KN, Wu XM, et al: Balo's disease showing benign clinical course and co-existence with multiple sclerosis-like lesions in Chinese. Mult Scler. 2008;14:418-424.
- Li Y, Xie P, Fan X, Tang H: Balo's concentric sclerosis presenting with benign clinical course and multiple sclerosis-like lesions on magnetic resonance images. Neurol India. 2009;57:66-68.
- Weinshenker BG, Miller D. Multiple sclerosis: one disease or many? In: Siva A, Kesselring J, Thompson AJ, eds. Frontiers in Multiple Sclerosis. vol 2. London: Martin Dunitz Ltd. 1999;37–46.
- Garbern J, Spence AM, Alvord EC. Balo's concentric demyelination diagnosed premortem. Neurology. 1986;36:1610–4.
- Chitnis T, Hollmann TJ: CADASIL mutation and Balo concentric sclerosis: a link between demyelination and ischemia? Neurology. 2012;78:221-223.
- Chen CJ, Chu NS, Lu CS, Sung CY. Serial magnetic resonance imaging in patients with Balo's concentric sclerosis: natural history of lesion development. Ann Neurol. 1999;46:651-656.
- Karaarslan E, Altintas A, Senol U, et al: Balo's concentric sclerosis: clinical and radiologic features of five cases. Am J Neuradiol. 2001;22:1362-1367.
- Purohit; et al. Balo's Concentric Sclerosis with Acute Presentation and Co-Existing Multiple Sclerosis-Typical Lesions on MRI. Case Rep Neurol. 2015;7:44–50.
- Chen CJ, Ro LS, Chang CN, Ho YS, Lu CS. Serial MRI studies in pathologically verified Balo's concentric sclerosis. J Comput Assist Tomogr. 1996;20:732-735.
- Gray F, Leger JM, Duyckaerts C, Bor Y. Balo's concentric sclerosis: lesions restricted to the pons. Rev Neurol (Paris). 1985;141:43-45.
- Moore GR, Neumann PE, Suzuki K, Lijtmaer HN, Traugott U, Raine CS. Balo's concentric sclerosis: new observations on lesion development. Ann Neurol. 1985;17:604-611.
- Karaarslan E, Altintas A, Senol U, et al: Balo's concentric sclerosis: clinical and radiologic features of five cases. Am J Neuradiol. 2001;22:1362-1367.

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