CASE REPORT

ONION PEEL APPEARANCE IN BALOS CONCENTRIC SCLEROSIS-A VARIANT OF MULTIPLE SCLEROSIS

Saeed Arif, Muhammad Waseem Wali*, Atiq ur Rehman Slehria, Hina Khalid***, Hamza Malik**

Department of Medicine, *Department of Neurology, **Department of Radiology, ***Department of Biochemistry, Combined Military Hospital Medical College, Lahore-Pakistan

Balo's concentric sclerosis (BCS) is a variant of multiple sclerosis (MS). It may present as a lesion clinically and radiologically indistinguishable from brain tumour particularly on computerized tomography (CT) scans. Diagnosis only gets clear when magnetic resonance imaging and spectroscopy (MRI & MRS) and brain biopsy is done. We report a case of 30 year old male with progressive headache and left hemi paresis for 3 weeks. There was upper motor neuron (UMN) facial palsy on the left with bilateral papilledema. CT scan of brain showed large hypo-dense area in right frontoparietal lobe consistent with brain tumour. On MRI the diagnosis of BCS was made on basis of concentric lesions of myelinated and demyelinated rings. Demyelination was confirmed on brain biopsy.

Keywords: Balo's concentric sclerosis, Multiple sclerosis, Tumefactive demyelinating lesion

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INTRODUCTION

Balo's concentric sclerosis (BCS) is a rare variant of multiple sclerosis(MS).[1] Any solitary demyelinating lesion greater than 2 cm in size with radiological features suggestive of tumour is called tumefactive demyelinating lesion (TDL).[2] Differentiation between demyelinating lesions like TDL and CNS tumours, in presence of single lesion is difficult, so it is mandatory to consider a demyelinating process in the differential diagnosis of tumour-like brain lesions.[3] Tumours resemble demylinating lesion on CT as they appear hypointense. Contrast magnetic resonance imaging and spectroscopy (MRI &MRS) is the modality of choice to differentiate between two diseases. Presence of concentric rings of myelinated and demyelinated lesions in BCS on MRI with specific peaks on MRS differentiates it from central nervous system tumours. The BCS disease is not well documented in the developing countries. This is the first case being reported from Pakistan.

CASE REPORT

A 30 years old previously healthy male presented with progressive headache and left sided weakness for 3 weeks. There were no visual problems, seizures, fever, cough or weight loss. He was married and denied any extra-marital relationship and drug abuse. Patient was afebrile with regular pulse of 85 beats/min, blood pressure of 110/70 mmHg in sitting position, respirations 20/min and 143 lbs weight. Cognition and other higher mental function were normal. Muscle power in left upper and lower limb was 1/5 and 2/5 respectively on Medical research council (MRC) scale with hypereflexia. There was upper motor neuron (UMN) facial palsy on the left with bilateral papilledema. Rest of the systemic examination remained unremarkable.

Autoimmune screening, angiotensin converting enzyme level and syphilis serology was normal. A plain chest radiograph and echocardiography revealed no abnormality. CT scan of brain revealed large hypo-dense area consistent with space occupying lesion in right frontal and parietal lobe. CSF analysis was normal with no oligoclonal bands or malignant cells. MRI brain showed well-defined, lobulated area in right frontal and parietal region which was hypo-intense on T1WI (Fig-1A). This lesion was hyper-intense on T2WI and FLAIR sequences (Fig-1B &1C). The lesion showed facilitated diffusion on DWI/ADC mapping (Fig-1E). No significant contrast enhancement was appreciated on T1W1CE (Fig-1D) sequences. GRE was unremarkable. The lesions showed concentric rings pattern with no significant peri-lesional oedema or mass effect was seen. On MR Spectroscopy, there was raised choline peak and relatively reduced N-acetyl-aspartate (NAA) peak along with double inverted peaks of lipid and lactate. NAA/ Choline ratio was 0.41 and choline/creatine ratio was 2.52. These findings were consistent with a diagnosis of Balos concentric sclerosis. Open brain biopsy of lesion was performed, and microscopy showed gliosis, foamy histiocytes and perivascular cuffing of lymphocytes consistent with acute demyelinating disease.

1g methylprednisolone infusion was given daily for 5 days followed by five sessions of plasmapheresis. He was started on mitoxantrone infusion 12mg/m^2 every three months for the next six months. Patient's muscle power improved to 4/5 in upper limb and 5/5 in lower limbs but the lesions did not regress on repeat MRI after six month. Same treatment was repeated for 6 more months and repeat MRI showed reduction in lesion size. At present patient is asymptomatic except for mild residual weakness in left upper limb.

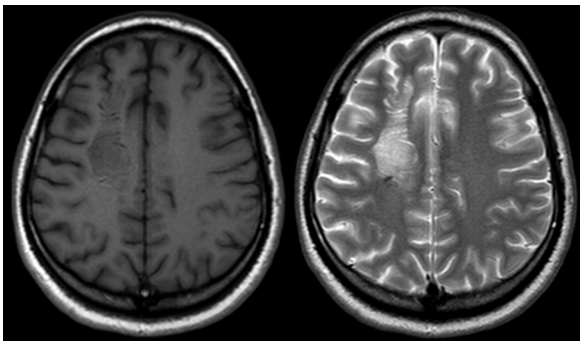

Figure-1A **Figure-1B:**

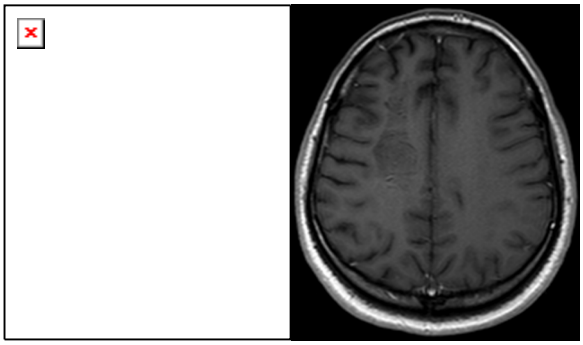

Figure-1C **Figure-1D**

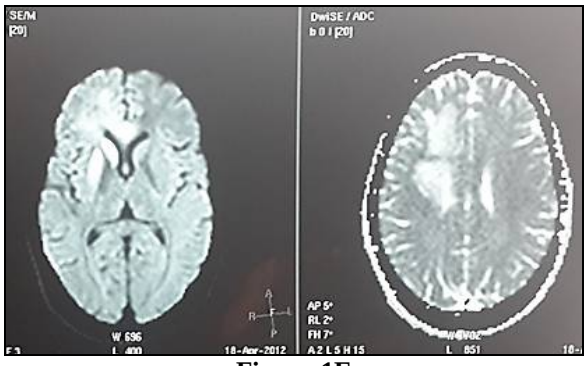

Figure-1E

Figure-1: Axial images of brain including T1WI, T2WI, FLAIR, DWI /ADC & T1WICE. Frontal & parietal lobes show hypointense lesion on T1WI (1A) which appears hyperintense on T2WI, FLAIR (1B&1C respectively) with typical concentric ring pattern (onion peel appearance), predominantly facilitated diffusion is visible on DWI/ADC (1E) and no significant contrast enhancement on T1WI CE (1D) sequence.

DISCUSSION

MS is a common disease in Pakistan with male to female ratio of 1.45:1. Relapsing remitting MS occurs in 81% of patients. In patients with severe disability mean disease duration is 5.2 year which is shorter than developed countries.[4] Diagnosis of MS and its variants is delayed due to MRI facility only available at certain hospitals in Pakistan.

BCS is a rare variant of MS. "Leuko-encephalitis periaxialis concentric" was the initial term coined by Joszef Balos in 1928 for BCS. This name was based on initial observation that "white matter is destroyed in brain in concentric layers in manner that leaves the axis cylinders intact".[5] It is characterized by alternating rings of demyelinated and myelinated lesions in concentric fashion in white matter.[6]

A self-limited, monophasic illness; relapsing-remitting and primary rapidly progressive disease are three known clinical subtypes of BCS.[7] CSF is either normal or shows mild mononuclear inflammatory reaction with oligoclonal band present only in few cases.[6,8]

Pathognomonic MRI features in BCS include alternating bands of demyelinated and myelinated white matter seen on T2 weighted images and the presence of concentric ring enhancement on T1 weighted images with gadolinium.[9] Chen reported a decrease of the N-acetyl-aspartate (NAA)/creatine (Cr) ratio, an increase of the choline (Cho)/Cr ratio, and emergence of a lactate peak in Balo´s concentric lesions.[10] These features are not present in CNS tumour.

Pathophysiology of BCS is not properly known, initially vascular pathology was considered culprit but according to Chen *et al*.[11] Balo's rings develop in stepwise fashion in a centrifugal direction.

According to preconditioning hypothesis by Christine *et al*.[12] demylinated layers in concentric lesion resembles lesion due to hypoxic injury with release of neuroprotective factors such as hypoxia-inducible factor 1a and heat-shock protein 70 at the edge of expanding peripheral lesion, making these lesions resistant to damage hence appear as mylinated layers in concentric lesion.

According to kira J aquaporin-4 (AQP4) are lost in hypertrophic astrocytes. This occurs in demyelinated and myelinated layers of lesion, with MRI-confirmed Balo's disease, but none of them were seropositive for anti-AQP4 antibody. So he proposed AQP4 astrocytopathy, in the absence of anti-AQP4 antibody, which is considered characteristic of Balo's disease.[13]

CONCLUSION

In developing countries MS is under diagnosed due to restricted MRI facility. Differentiation of demyelinating disease like MS and BCS require MRI facility. The diagnosis of brain tumour should not be made only on CT scan. Characteristic MRI feature of BCS lesions differentiate it from CNS tumour. If there is any ambiguity about diagnosis, brain biopsy should be done before starting treatment.

AUTHOR CONTRIBUTIONS

SA: Study concept, design, data acquisition, data analysis and interpretation. MWW: Critical revision of the manuscript for important intellectual content. ARS: Analysis and critical revision of the manuscript for important intellectual content. HK and HM: Literature review, formatting and data analysis.

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Address for Correspondence:

Dr. Saeed Arif, Resident in Medicine, Department of Medicine, CMH Lahore Medical College, Lahore-Pakistan
Cell: +92 333 455 5721
Email: drsaeedarif@gmail.com