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Speech Disorders Revealing Balo's Concentric Sclerosis: A Case Report

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Abstract **Case Report**

Introduction: Balo's concentric sclerosis (BCS) is a particular and rare demyelinating diseaseof the white matter of the brain that was first described by J. Balo in 1928 and considered as a rare variant of multiple sclerosis. The clinical manifestations usually consist of aphasia, behavioral disturbances, and seizures. This term results from a pattern of concentric aspect of the lesions with alternating areas of demyelination and areas of normal myelin in various parts of the brain and spinal cord. BCS was described as monophasic, progressive, and treated as a disease with a fulminant course. ***Case report***: We report a case of 38 years old woman who developed slurring of speech with mild swallowing disorders for solid foods accompanied by vertigo. Her MRI showed "onion peel-like" demyelinating lesions over the subcortical areas ofthe left parietal lobe. Treatment given to the Patient was IV methylprednisolone followed by oral corticosteroids, with a favorable outcome without clinical recurrence after two years of follow-up. ***Conclusion:*** BCS is a rare disease with an unpredictable course, the pathognomonic brain MRI features help to secure the diagnosis. The clinical case describedis interesting because of the rarity of this entity.

Keywords: Balo's concentric sclerosis, Multiple sclerosis, Speech disorders, Brain MRI.

INTRODUCTION

Balo's concentric sclerosis (BCS), is a rare demyelinating central nervous disease system, considered as one of the forms of multiple sclerosis (MS), characterized by a formation of concentric rings corresponding to bands of preserved and destroyed myelin areas in the white matter of the brain often described as "onion bulbs" due to the appearance [1-3]. For the first time, concentric elements in the areas of demyelination were mentioned by O. Marburg in 1906. Initially, this disease was called "Concentric periaxial encephalitis ", according to the definition author, who showed that the brain's white matter is destroyed like concentric layers. Based on this, the Hungarian pathologist J. Balo in 1927 first suggested that the described pathology is a variant of the acute course of MS [4]. Clinical manifestations of BCS are usually paresis, aphasia, cognitive disorders in combination with headaches or seizures. Most of the cases described in the literature usually presented like acute demyelination syndrome with a monophasic rapidly progressive course and death within a few weeks or months. However, in recent years, the publication of clinical observations has shown a good prognosis in the course of SBC with longer life expectancy or spontaneous remission [5]. We report this case of a Moroccan woman diagnosed with BCS whose history was marked by resolution of symptoms without relapse to confirm that BCS is not always fatal, with a good response to early treatment by steroids.

CASE REPORT

A 38 years old woman, right-handed, mother of 2 children, with no history of particular personal or family health problems, was admitted to the neurology department of Mohamed V Military Teaching Hospital whose history of illness goes back to 8 days of her admission with the acute installation of slurring of speech, slight disorders of swallowing for solids accompanied by vertigo and headache which was gradually progressive, without history suggestive of sensory or motor disturbance, sphincter disorders, seizures or clinical infectious syndrome.

The evolution was marked by the persistence of the symptomatology. The clinical examination on admission found isolated spasmodic dysarthria and slight disorders of swallowing for solids, the examination of Cranial nerves was without abnormality.

Laboratory results including hepatitis B, human immunodeficiency virus, syphilis, autoimmune

disease workup in addition to the thyroid-stimulating hormone, renal and liver function test, vitamin B12, and folate levels were normal. CSF analysis showed no cells, protein 0.2g/l and glucose 0.65 g/l, with negative oligoclonal bands. The Brain MRI revealed a tumefactive demyelinating lesion in the left parietal lobe with alternating hypointense and hyperintense concentric pattern "onion-like" bands on T2-weighted (Figure-1b) and FLAIR (Figure1a), with peripheral enhancement in post-contrast images in parietal white matter with mild edema and no hemorrhage, and containing areas of necrosis (Figure 1c, 1d) suggesting a BCS diagnosis. Spinal cord MRI was without abnormality.

The diagnosis of BCS was retained and the Treatment given to the Patient was IV methylprednisolone 500 mg/day for 8 days followed by oral corticosteroids 1 mg/day for 2 months followed by a decrease in dose and as symptomatic treatment of headaches paracetamol 1g/12 hours has been prescribed, with a favorable outcome, the patient showed a good response to the treatment we used, with the disappearance of swallowing disorders for solid foods, vertigo, and headaches, for speech disorders a marked resolution was obtained. After two years of follow-up, at a frequency of every six months, the MRI performed showed a residual left parietal lesion without having a new lesion, clinically the patient still has no clinical recurrence, and her neurological examination remains normal.

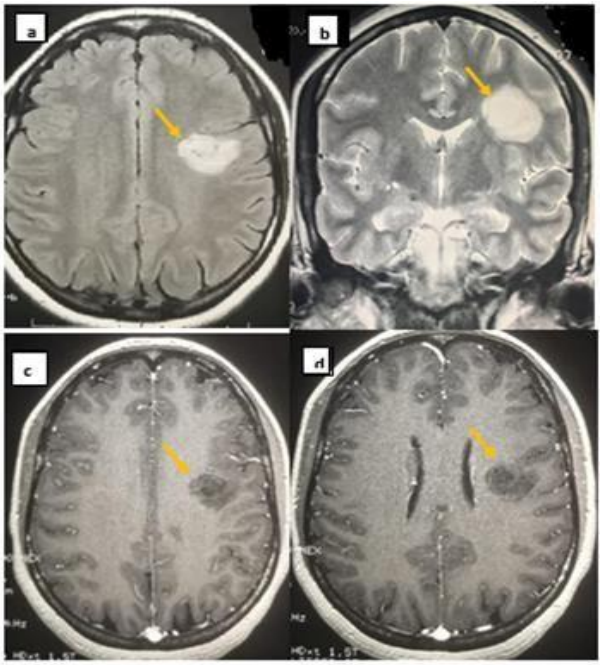

Fig-1: Frontal T2-weighted (b) and axial FLAIR (a) images demonstrate a left alternating iso/hyperintense rings and heterogeneous parietal lesion which show a concentric lamellar appearance.. Axial T1-weighted image (c, d) after administration of gadolinium shows hypo-intense rings enhancement of the lesion measuring 18x22x19 mm and containing areas of necrosis, consistent with Balo's concentric sclerosis

DISCUSSION

The diagnosis of BCS is now based on clinical manifestations and pathognomonic pattern on brain MRI with alternating hypo-isointense and hyperintense concentric rings, the frontal and parietal regions of the white matter are the usual sites of lesions, however, involvement of the cerebellum and brainstem, as well as the spinal cord, is possible with bad prognosis (absence in our case with a normal spinal cord MRI). To visualize this concentric pattern, brain MRI should be performed early in the course of the disease [6]. BCS can present with different clinical pictures, in our case speech disorders dominated the clinical presentation.

The histology of Balo's concentric sclerosis is marked by alternating bands of myelin preservation or demyelination in the cortex, brainstem, optic chiasm and spinal cord [7].

The first cases reports were considered for a long time BCS as an acute demyelinating lethal disease and the definitive diagnosis was made by autopsy [8]. The typical clinical course is progressive but a pattern of relapses and remissions has been reported [9]. Our patient had classic clinical features and MRI findings. The association between lesions of BCS and multiple sclerosis is not completely understood. BCS may be a real variant of multiple sclerosis or a different but related entity. It is evident that the classical multiple sclerosis and BCS do not only have an overlap in pathology. BCS lesions can appear during the clinical evolution of relapsing- remitting multiple sclerosis [10]. About 55% of patients who present with a BCS or BCS-like lesion have typical multiple sclerosis lesions somewhere on the MRI scan [11]. In a Chinese series involving 7 patients with BCS diagnosis, two cases relapsed with demyelinating lesions typical of multiple sclerosis [11]. In addition, some patients with BCS lesions and positive oligoclonal bands in the CSF go on to develop multiple sclerosis[7], raising the question of having positive oligoclonal bands predict the possibility of transformation to MS. Not every patient with BCS lesions has oligoclonal bands; in a study of 11 patients with BCS disease, CSF synthesis of oligoclonal bands was found in only one patient [12], which is the case for our patient with a normal CSF study.

The prognosis of patients with symptoms of a BCS disease is variable. Cases of total physical and radiological resolution have been described; whereas, patients with aggressive BCS can die or remain with considerable morbidity. Outcomes intermediate between these two averages seem to be the most frequent. In a study of 10 patients with typical BCS MRI lesions, 83% had marked or total clinical recovery and no deaths were recorded [13]. A comparably favorable prognosis was also reported in a study of five cases who responded to therapy after the first attack, over a mean follow-up of 30 months, all patients remained relapse-free [14]. Our case o with a total

recovery, and without relapse after 2 years of follow-up provides further evidence of a possible good prognosis for patient with BCS, and is consistent with the results of these studies.

For the therapeutic strategy in this entity, it is difficult to choose the optimal protocol in the absence of clinical trials. Because of the pathological similarities to multiple sclerosis (MS), corticosteroids are the recommended first-line treatment for an acute episode of neurological dysfunction. Most clinicians, based on case reports and experiences, use high doses of intravenous or oral steroids, or both. Doses and duration of treatment vary from case to case [15, 16]. The results of steroid treatment also vary considerably, many patients have shown improvements in the range of days to months, followed by full recovery, relapse, or delayed recovery [17]. Plasma exchange seems a reasonable second-line option for some authors [18, 19]. There are insufficient data for cyclophosphamide, azathioprine, mitoxantrone, and intravenous immunoglobulin [20]. The failure of monoclonal antibodies has been demonstrated with alemtuzumab [21].

Despite the fact that MRI lesions have provided a better understanding of BCS, still many areas of doubt exist. The rarity of the disease makes it impossible to conduct large studies. An international collaborative project or a registry of the disease is required to get enough patients to make conclusions about the various aspects of the disease.

CONCLUSION

The described case represents a rare neurological disease. The diagnosis in our observation was based on clinic manifestations, especially as an acute Speech disorders, and brain MRI with a total resolution of symptoms. Our observation confirm that BCS is not always lethal, with a good response to early treatment by steroids. BCS disease still subject of studies and research, the multiplication of publications on BCS will allow us to better understand its pathophysiology and therefore to find a better therapeutic arsenal for patients.

Ethical Approval

We confirm that no ethical approval is required to publish case reports from our institutions and informed consent was obtained from the patient to report this case.

Conflicts of Interest: The authors declare no conflicts of interest regarding the publication of this paper.

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