

Favorable Outcome in a Case of Von Balo's Sclerosis Mimicking a Juvenile Stroke at the Onset A New Possible Role of Dynamic Susceptibility Contrast (Dsc) Perfusion-Weighted Imaging (PWI).

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Abstract

We describe a juvenile stroke-like onset of Von Balò's sclerosis, with a favorable outcome after 4 years of follow up, even if treatment's protocols could not have been completed, because her low compliance.

Following the patient with annual MRI imaging we surprisingly discovered associations between which was reported at a Perfusion-weighted Imaging (PWI) Dynamic susceptibility contrast (DSC)-MRI executed after 9 days from the exordium and patient's clinical residues.

By describing the case we focus on a new way to use PWI-DSC in order not only to determine areas of Blood-Brain-Barrier active lesion but also to have information on patients' prognosis and to guide neurologist in his therapeutical choices.

PWI can't substitute other MRI sequences, which describe, in that moment of execution, how many cerebral areas are involved in the process of demyelization, but PWI, surely, is an excellent sequence to integrate diagnosis and improve patients' clinical, diagnostic and therapeutic follow up.

Case Report

A 39-year-old, heavy smoker, left-handed woman, affected by obesity, dyslipidemia and hypertension presented in an emergency hospital, due to the subacute onset of left brachio-crural hemiparesis, and motor aphasia lasting for two days. The patient underwent brain CT scan that showed some periventricular nuanced hypodensities in the right frontoparietal white matter, reported as probably ischemic (Fig. 1.1).

Despite the antiplatelet therapy, hemiplegia and cognitive/behavioral dysfunctions emerged in a few days and the patient was transferred in our Neurological Department. She underwent brain MRI that showed multiple and not ischemic grossly rounded lesions, bilaterally located in the deep frontoparietal white matter, some of which surrounded by vasogenic edema. They appeared hypo-isointense on T1-weighted and hyperintense with peripheral hypo-intensity on T2-weighted images (Fig. 1.2). Some regions of signal restriction associated to intense pseudo nodular enhancement were respectively observable on Apparent Diffusion Coefficient (ADC) map of Diffusion Weighted Imaging (DWI) and after gadolinium injection (Fig. 1.3).

Because of the rapid worsening of symptoms another MRI was executed after nine days, which was also integrated by Dynamic susceptibility contrast (DSC) Perfusion-weighted Imaging (PWI).

New T2 hyperintense lesions had appeared, while others had enlarged (Fig. 1.4). Signal restriction on DWI had disappeared, and perilesional vasogenic edema had increased (Fig. 1.5). Different "open ring" and pseudostratified patterns of enhancement were now observable (Fig. 1.6), although, cerebral blood volume map (CBV) acquired with dynamic susceptibility contrast perfusion MRI showed no increased perfusion, but only a mild hypoperfusion in the right frontal lobe. (Fig. 1.7).

These new imaging findings allowed the diagnosis of a Von Balò's demyelinating disease (supported also by an increase of cerebrospinal fluid proteins after lumbar puncture and normal cervical spinal MRI).

Corticosteroid therapy with adjunctive plasmapheresis were started with minimal benefit, but the patient didn't end the whole cycle because of her low compliance. So, she decided to come back home, against doctors' opinion, with an important disability characterized by left-body hyposthenia, left arm intentional tremor, and alternating moments of strong depression and euphoria.

Nevertheless, we obtained by mutual agreement to follow the patient clinically and with annual MRI follow-up.

MRI, performed 4 years after the onset, showed vasogenic edema and contrast medium uptake disappearance (Fig. 1.8). Fewer and smaller T2-hyperintensities were present in the white matter of the frontal lobes, especially on the right side (Fig. 1.9). Slight bilateral frontal atrophic changes had appeared in the meantime, although clinical improvement was still noted. Milder symptoms such as mood instability, left hyposthenia, intentional left-tremor at upper limb, and some frontal behavioral defects were residual.

Moreover, such residual symptoms anatomically correlated with the right frontal lobe which had been the most hypo-perfused region on PWI during the acute/subacute phase of the disease.

So, we asked ourselves if PWI could be used to improve diagnosis and predict prognosis.

Discussion

Balò's sclerosis is a rare demyelinating disorder characterized by concentric rings of alternating demyelinated and partially myelinated fibers ¹ progressively extending to the periphery. It is still considered as an aggressive variant of multiple sclerosis ², but to date there are still no officially approved pathogenetic theories. Although historically fatal, nowadays around half of patients recover fully after receiving immunosuppressive treatment. ^{3,4}

The diagnosis of Balò's sclerosis is still not easy due to its rarity and to the lack of specificity of the symptoms. Officially, brain MRI with T2- and T1-weighted sequences is enough for diagnosis.

Some studies have shown that DSC-MRI can be used to differentiate metastasis or necrosis regions after radiotherapy from active tumefactive demyelinating lesions based on results of analysis of rCBV. This property depends from the fact that demyelinating lesions, are characterized by intrinsically normal or inflamed vessels with only mild inflammatory angiogenesis producing a normal or only mildly increased rCBV. ⁵.

To our knowledge, no study has reported the usefulness of PWI in differential diagnosis of Von Balò's sclerosis lesions, nor its usefulness for prognosis.

We found positive correlation between cerebral hypoperfusion, clinical course and outcome in a patient with Von Balò's sclerosis, through the use of DSC PWI.

A lot of studies reported the diagnostic role of PWI also in patients with cerebral atrophy, cognitive or psychiatric diseases, as in frontotemporal Dementia (FTD), where regions of cortical atrophy and concordant hypoperfusion have been found, and also associations between regional perfusion and psycho-emotional scores specific to depression or anxiety. ⁶ .

Mild cognitive impairment and mood alteration are the same symptoms which the patient still presents, and they have been anatomically identified four years before without knowing it.

To date any neurodegenerative diseases produce significant brain-function alterations detectable with brain Positron Emission Tomography (PET) or 99mTc-hexamethylpropyleneamine oxime single-photon emission-computed tomography (HMPAO-SPECT) even when structural CT or MRI images ⁷, because of the significant voxel wise correlations between changes in grey matter atrophy and FDG up- take ⁸, although MRI PWI can represent a diagnostic tool for detecting pathologic perfusion alterations ⁹ and therefore also a useful clinical support.

DSC PWI gave us precious prognostic information which have been precisely interpreted and correlated with the patient's residual symptoms only after 4 years. It could be interesting to discover if this case represents an isolated event or it really resumes PWI usefulness in management of patients with Von Balò's disease, although many other cases will need to be analyzed with the same MR technique in this regard.

Conclusions

This observation highlights a unique case of Balò's sclerosis, which despite the initial wrong diagnosis of juvenile stroke and the patient's decline to undergo proper treatment, had a good and predictable outcome.

Our aim is to invite to add PWI in MRI protocols in cases of cerebral neuroinflammation, which could be useful to neuroradiologists in diagnosis, differentials and prognosis and it could guide neurologist through therapy's choice and through monitoring timing decisions, especially in cases of Von Balò's sclerosis which, due to the clinical and prognosis variability, represents an intriguing differential diagnosis and a therapeutical challenge still today without an explication.

Declarations

No funding was received.

We have no conflict of interest to disclose.

The submitted work has not been published previously and is not being considered for publication elsewhere.

Material has not been reproduced from prior publications, whether by the same or different authors. Data and materials are original. MRI sequences were performed in our Hospital, Policlinico di Napoli, AOU "Federico II".

Any previously published material is explicitly quoted and referenced.

Prior publications or other submissions on the same data set or problem have been disclosed.

Additional Contributions:

We thank the patient for granting permission to publish this information. Not Ethical Approval was needed but only patient's consent.

Authors Contributions:

Each one of the authors has contributed with professionalism and experience at this work.

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Figures

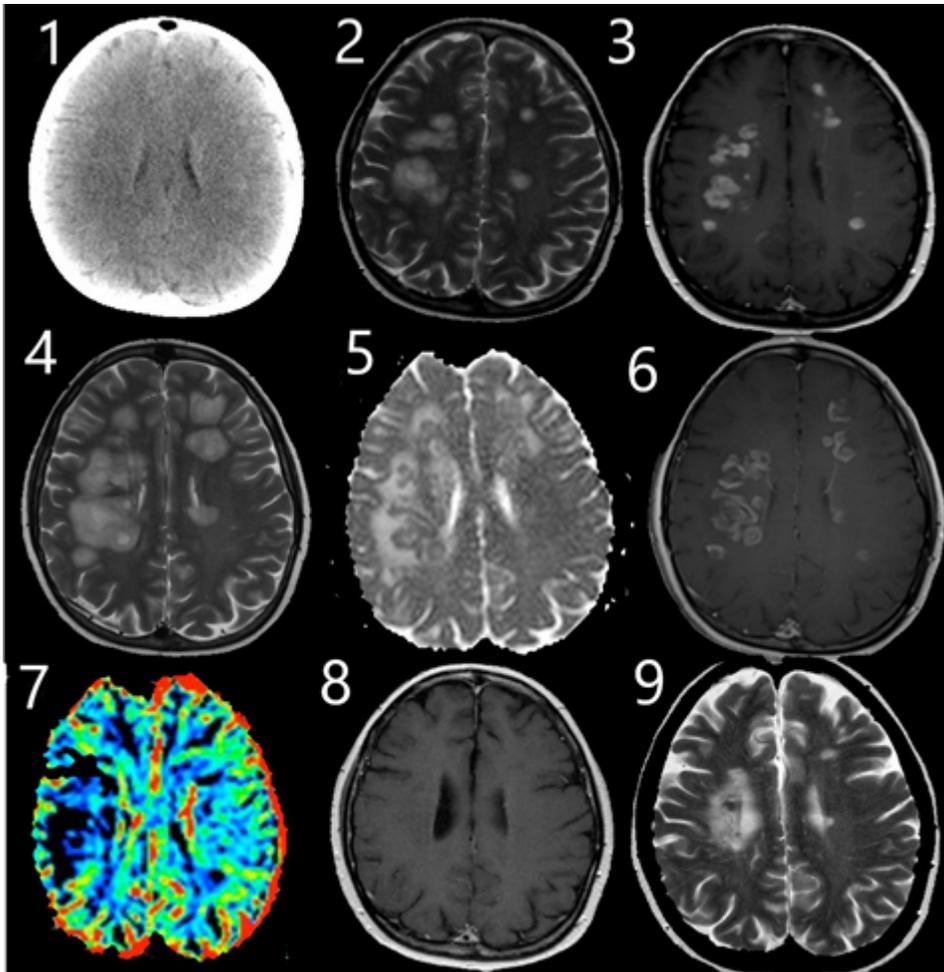


Figure 1

1.1 CT scan at the onset of symptoms. 1.2 T2-weighted MRI performed at the recovery. 1.3 T1-weighted MRI after gadolinium injection performed at the recovery. 1.4 T2-weighted MRI performed 9 days after the recovery. 1.5 Diffusion Weighted Imaging (DWI) MRI performed 9 days after the recovery. 1.6 T1-weighted MRI after gadolinium injection performed 9 days after the recovery. 1.7 Dynamic susceptibility contrast (DSC) Perfusion-weighted Imaging (PWI) MRI performed 9 days after the recovery. 1.8 T1-weighted MRI after gadolinium injection performed 4 years after the recovery. 1.9 T2-weighted MRI performed 4 years after the recovery. Note. Brain Imaging acquisitions.