Delayed posthypoxic leukoencephalopathy: a new case of a rare reversible white matter disease

G. Peppoloni, A. Cerase, C. Battisti, F. Simeone, L. Marchetti and A. Federico
Department of Medicine, Surgery and Neurosciences. University of Siena, Siena,Italy

Background

Delayed posthypoxic leukoencephalopathy (DPHL) is a rare condition characterized by a neuropsichiatric relapse after a full recovery from an acute and prolonged cerebral hypo-oxigenation (usually ranging from 7 to 21 days).

It can be caused by any cerebral hypoxic insult (mainly associated with carbon monoxide poisoning and overdoses of opiates or benzodiazepines).

It has also been suggested that pseudodeficiency of arylsulfatase-A predisposes to DPHL.

MRI findings consist in diffuse and confluent T2 hyperintensity, predominately within the centrum semiovale and subcortical white matter with associated prolonged restricted diffusion.

Partial or full recovery can be obtained with supportive treatment.

Case report

A 58-year-old woman, with an history of cocaine abuse, was found unconscious with shallow breathing and pulseless (GCS 5).
After intubation, she was admitted to an intensive care unit; an echocardiogram revealed a severe heart failure nonresponsive to inotropic treatment, so that ECMO V-A positioning was necessary.

MRI (Fig. 1a): negative
EEG: diffuse slowing.
Toxicological exam: negative for common drugs
Lumbar puncture: clear CSF with normal parameters.

Significant clinical improvement was obtained, but **on the 16th day** the patient developed a sudden and severe state of unresponsiveness with spastic tetraparesis, left lateral decubitus and diffuse hyperreflexia.

MRI (Fig. 1b): FLAIR images showed signal alteration in the subcortical white matter of anterior frontal lobes and corpus callosum, mainly in the right side, as well in the left globus pallidus.
Arylsulfatase enzyme activity was normal.
Clinical improvement occurred with supportive care only. The patient returned to her neurological baseline after 6 months.

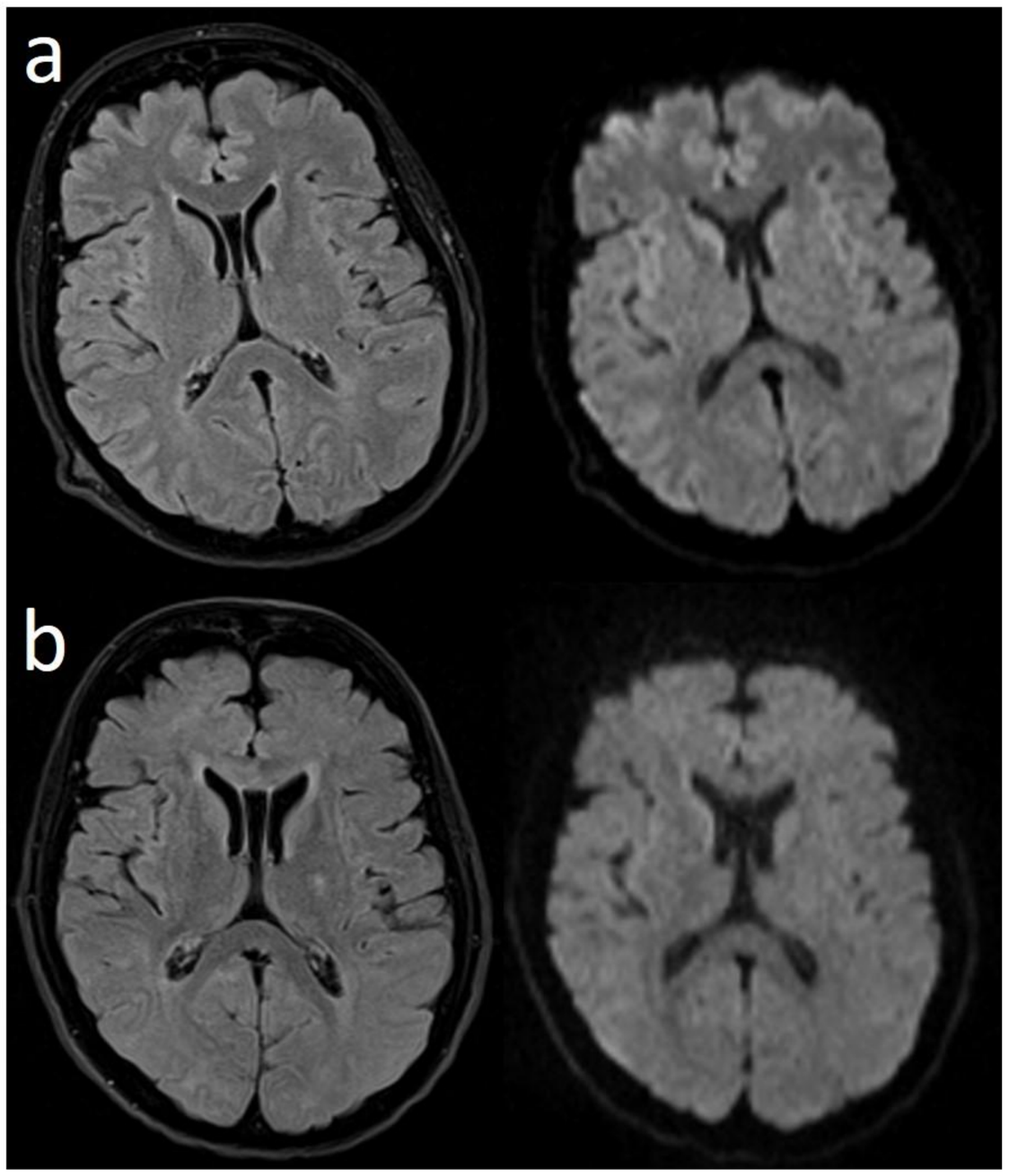

Fig.1 Magnetic resonance of the brain.
At admission (a), fat-suppressed FLAIR (left image) and DWI (right image) axial images were negative. Three weeks later (b), only fat-suppressed FLAIR images showed signal alteration in the subcortical white matter of anterior frontal lobes and corpus callosum, mainly in the right side, as well in the left globus pallidus.

Conclusion

The etiology responsible for DPHL in our patient remains unknown but clinical course and MRI findings allowed us to obtain a certain diagnosis.

DPHL is a rare event and patophisiology is still poorly understood. In reporting this case we aim to focus on an often misdiagnosed condition, generally characterized by a **favourable prognosis, obtaining only with supportive care**.

According to clinical course and neuroradiologic features, an early diagnosis can be achieved, avoiding unnecessary and invasive diagnostic procedures.

References

1.Zamora CA, Nauen D, Hynecek R, Ilica AT, Izbudak I, Sair HI, Gujar SK, Pillai JJ. *Delayed posthypoxic leukoencephalopathy: a case series and review of the literature*, Brain Behav. 2015 Aug;5(8):e00364. doi: 10.1002/brb3.364. Epub 2015 Jul 3. Review.
2.Shprecher D, Mehta LM *The Syndrome of Delayed Post-Hypoxic Leukoencephalopathy,* NeuroRehabilitation. 2010;26(1):65-72.
3.Molloy S, Soh C, Williams TL., *Reversible Delayed Posthypoxic Leukoencephalopathy*, AJNR Am J Neuroradiol. 2006 Sep;27(8):1763-5.