Clinical picture

Superficial siderosis of central nervous system of unknown cause

Clinical images

A 44-year-old man with a past medical history unremarkable presented with a 4-month evolution of progressive cognitive deterioration, deafness and unsteady gait. Initial neurological examination revealed mild cognitive impairment, dysarthria, cerebellar ataxia with wide-based unsteady gait and bilateral sensorineural hypoacusia. Laboratory serum analysis was unremarkable, including vitamin B12 and thiamine levels, thyroid function, autoimmunity panel; negative serologies for human immunodeficiency virus, Borrelia burgdorferi, Trophera whippe-lii, Treponema pallidum, besides other bacterial and fungal infections. Brain-computed tomography (CT) was normal. A non-traumatic lumbar puncture was performed and a hematic cerebrospinal fluid (CSF) was obtained, which characteristics persisted during its collection (Figure 1a). CSF analysis revealed 110 cells/μl, predominantly lymphocytes, uncountable red blood cells, and 0.7 mg/dl of protein.

Figure 1. (a) Hematic macroscopic aspect of CSF, obtained on the first lumbar puncture. (b) Axial T2-weighted images showing a hypointense rim surrounding cerebellum, due to haemosiderin deposits.

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cells, 453 mg/dl of proteins and normal glucose level (44 mg/dl). Brain magnetic resonance imaging (MRI) disclosed extensive, regular rim T2-weighted leptomeningeal hypointensities images, involving supratentorial and posterior fossa (Figure 1b) and no evidence of artery–venous malformations. Neurological improvement was noticed, with ataxic gait partial recovery after 24-month follow up. The reminding imaging study, including a myelography, a myelo-CT and scintigraphy with labelled erythrocytes were normal and a haemorrhage origin was not possible to identify.

Comment
Superficial siderosis of the nervous system results from chronic or intermittent haemorrhage into the subarachnoid space, causing haemosiderin deposition in subpial layers with gliosis and neuronal death. Although previously defined as a rare pathological condition with less than 300 cases described until 2006, recent data suggest that it seems to be more frequent. Cerebellum, basal frontal lobes, mesial temporal lobes, optic, vestibulocochlear nerves and spinal cord are particularly prone to haemosiderin deposition. Progressive sensorineural hypoacusis, cerebellar ataxia and pyramidal signal constitute the hallmark; however less than half of the patients present with this triad. T2-weighted magnetic resonance imaging is characterized by hypointense marginal rim around the neuroaxis. The CSF is altered in up to 75% of cases with abundant red cells and/or xanthochromia, elevated protein and ferritin levels. Despite the advances in imaging and functional techniques, the origin of the haemorrhage fails to be detected in up to 50% of the reported cases, impairing a chirurgical intervention. No effective pharmacologic therapeutics is available, although the use of iron chelates and corticoid drugs has been reported.

Superficial siderosis of central nervous system should be considered in differential diagnosis of unexplained progressive cerebellar ataxia. In our patient, the clinical improvement under no specific treatment could suggest a transitory bleeding source.

Conflicts of Interest: None declared.

References