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Superficial siderosis of the central nervous system

An unusual cause of sensorineural hearing loss

Ana Claudia Rodrigues de Cerqueira[1,2], Antônio Egídio Nardi[2], José Marcelo Ferreira Bezerra[1]

Superficial siderosis (SS) of the central nervous system (CNS) is a rare disorder characterized by a classic triad of symptoms consisting of sensorineural hearing loss, cerebellar ataxia, and myelopathy. Less frequently, it is also associated with bladder disturbance, anosmia, extraocular palsies, anisocoria, dementia, and Parkinsonism[1,2]. SS is caused by chronic or recurrent haemorrhage into the subaracnoid space with haemossiderin deposition in the subpial layers of brain and spinal cord, which leads to progressive and irreversible neurological dysfunction. There are many causes for it, including aneurysms, vascular malformations, and CNS tumours. However, in most cases despite extensive investigations, the cause of bleeding is often not apparent. Magnetic resonance imaging (MRI) of the brain is the main laboratory screening exam to confirm the diagnosis, which shows a characteristic rim of hypointensity seen on T2- weighted around the brain, brainstem and spinal cord[3].

Sensorineural hearing loss may be the first symptom of the disease, and early diagnosis is extremely important, because this progressive disorder may lead to deafness, dementia and confinement[4].

We present a case study related to the possibility of superficial siderosis of the CNS in patients presenting sensorineural hearing loss, will be discussed.

CASE

A 53-year old woman was admitted for treatment with a 5-year history of progressive bilateral sensorineural hearing loss and constant tinnitus. She also complained of headaches, dizziness and nausea. A year before admission, the patient was suffering from progressive unsteadiness of gait. There was no history of CNS trauma or surgery. The neurological evaluation revealed bilateral hearing loss, lower limb spasticity, while coordination testing demonstrated severe bilateral limb ataxia. Her tendon reflexes were symmetrically hyperactive, with extensor plantar responses. Her gait was mixed spastic-ataxic. The remainder of the examination did not reveal anything noteworthy.

Audiometry demonstrated bilateral profound sensorineural hearing loss. MRI of the brain showed on T2-weighted images a rim of hypointensity surrounding the surface of the sylvian fissures, brainstem and the cerebellum consistent with SS (Figure). Recurrent subarachnoid haemorrhage was confirmed by serial lumbar punctures showing a predominance of red blood cells and signs of xanthochromia. The diagnosis of SS was made based on the classical symptomatology supported by typical neuroradiological features. The cerebral angiogram and MRI of the spinal cord were normal. At a follow-up visit one year later, she became more ataxic, with several falls at home. Despite detailed investigation, the origin of the bleeding remained undetermined.

The patient authorized the publication of this case study by signing a consent form.

Correspondence
Ana Cláudia Rodrigues de Cerqueira
Rua Mariz e Barros 501 / 607
24220-120 Niterói RJ - Brasil
E-mail : anacerqueira@globo.com

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SIDEROSE SUPERFICIAL DO SISTEMA NERVOSO CENTRAL: UMA CAUSA INCOMUM DE PERDA AUDITIVA NEUROSSENSORIAL

[1]Neurology Service, State University of Rio de Janeiro (UERJ), Rio de Janeiro RJ, Brazil; [2]Laboratory of Panic & Respiration, Institute of Psychiatry, Federal University of Rio de Janeiro (UFRJ), INCT Translational Medicine, Rio de Janeiro RJ, Brazil.

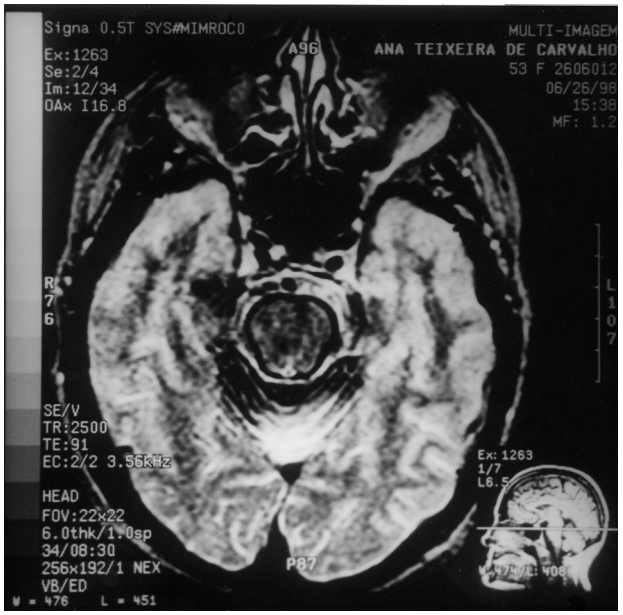

Figure. MRI axial section in T2 weighted shows low signal intensity, consistent with haemossiderin deposition, around the midbrain and cerebellum.

DISCUSSION

In the past few decades, SS could only be diagnosed by biopsy, surgical inspection of the brain or at a post-mortem examination. The wide availability of MRI in recent years has enabled physicians to make the non-invasive diagnosis of SS, and discover incidental presymptomatic cases[3,4].

This rare disorder is caused by chronic slow or repeated haemorrhage into the subarachnoid space with resultant haemosiderin deposition in the subpial layers of CNS. There are many causes including CNS tumours (21%), head or back trauma (13%), arteriovenous malformations / aneurysms (9%), post-surgical changes related neurosurgeries (7%), brachial plexus injury (6%), amyloid angiopathy (3%) and chronic subdural hematomas. However, the source of the bleeding remains obscure in approximately 36% of the cases, as informed in a recent study by Levy et al.[1] in a literature review of 270 cases of SS worldwide.

In 1908, Hamill[5] described the first case of SS in a post-mortem examination, and the pathological finding demonstrated haemosiderin deposition on the surface of the CNS. For many years, the etiology of SS remained controversial, two hypotheses were proposed: the first was a consequence of chronic subarachnoid haemorrhage, and the second was a metabolic disease, similar to systemic hemochromatosis[6]. The first hypothesis was confirmed by the classical experiment undertaken by Iwanowski and Olzewski[7] in 1960. The authors reproduced SS experimentally in dogs after repeated subarachnoid injections of blood or iron dextran.

According to Koppen et al.[8] the following mechanisms have been implicated in the pathogenesis of SS: [1] chronic or intermittent extravasations of blood into the subarachnoid space and dissemination of heme by circulating cerebrospinal fluid (CSF), [2] hemolysis, [3] entry of heme into exposed tissue, [4] conversion of heme to free iron, ferritin, and hemossiderin, leading to [5] damage of neural tissue. Microscopically, there is reactive gliosis, demyelination, neuronal loss, and intracellular ovoid bodies.

The susceptibility of the cortex cerebellar and eighth cranial nerve may be explained by the ability of glia cells in these areas to accelerate the biosynthesis of ferritin in response to prolonged contact with hemoglobin iron. Furthermore, the vestibulocochlear nerve is vulnerable because, it runs through the pontine cistern and has a long glial segment that predisposes it to iron deposition[8]. As a result, sensorineural hearing loss may be the first symptom. It is usually bilateral and is associated with high-tone frequencies, probably related to the tonotopic organization of axons transmitting the high-frequency sound at the surface of eighth cranial nerve[9].

In their review, Levy et al.[1] found that the classic manifestations of hearing loss, cerebellar ataxia and myelophathy are present in 39% of patients with SS. Other relatively frequent clinical findings are bladder dysfunction (14%), headaches (14%), anosmia (14%), diplopia (4%), bowel problems (3%), and ageusia (2%), and cranial nerve palsies (2%).

Brain MRI is the main screening exam to confirm the diagnosis of SS. The magnetic susceptibility effects of blood degradation products, such as ferritin and haemossiderin, produce a characteristic rim of hypointensity surrounding the surface of the brainstem, cerebellum, and cortical fissures on gradient-echo T2-weighted images. Furthermore, MRI may also show cerebellar atrophy, especially in the superior vermis and the anterior cerebellar hemispheres, or atrophy of the spinal cord. The presence of a fluid-filled collection in the spinal canal is a common finding of MRI and may be the likely source of bleeding[3,10].

There is no specific treatment available for idiopathic SS. Therefore, identification of the underlying cause when possible and ablation of the bleeding source may prevent the progression of this condition. The administration of iron chelating agents, such as desferrioxamine or trientine, has been tried in several patients[1,4], but their efficacy remains limited.

In this study, the patient showed clinical manifestations and MRI findings consistent with SS of the CNS. However, in this case the etiology of the condition remained unknown. In conclusion, it is believed that an early diagnosis for prompt intervention is extremely important whenever possible, because the symptoms are usually progressive and irreversible.

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