Different etiologies of superficial siderosis

Diferentes etiologias de siderose superficial

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ABSTRACT

Superficial Siderosis (SS) is an uncommon condition caused by hemosiderin deposition into the subarachnoid space. SS is characterized by cerebellar ataxia, progressive sensorineural hearing loss and pyramidal signs, but is often an unrecognized disorder. Magnetic Resonance Imaging (MRI) is the diagnostic procedure of choice due its high sensitivity to hemosiderin deposits in addition to being a non-invasive exam. This paper aims to describe a case of SS and to perform a literature review about SS etiologies, neuroimaging features and clinical characteristics. A 65-year-old man came to a neurology outpatient clinic with seizures and cerebellar ataxia with a history of car accident and severe traumatic brain injury 45 years ago. MRI SWAN showed a hyposignal in the cisterns of the base and on the cerebellar surface and T1-weighted images left hippocampal sclerosis.

Keywords: siderosis, magnetic resonance imaging, epilepsy.

RESUMO

A Siderose Superficial (SS) é uma condição rara causada por depósitos de hemossiderina no espaço subaracnóideo. SS é caracterizada por ataxia cerebelar, perda neurosensorial auditiva progressiva e sinais piramidais, mas é frequentemente uma desordem de difícil diagnóstico. A Ressonância Magnética (RM) é o exame de escolha para o diagnóstico devido a sua alta sensibilidade aos depósitos de hemossiderina, além de ser um exame não invasivo. Este artigo tem como objetivo descrever um caso de SS e realizar uma revisão da literatura sobre as etiologias da SS, suas características na neuroimagem e suas características clínicas. Um homem de 65 anos de idade procurou o ambulatório de neurologia com convulsões e ataxia cerebelar. Ele informou histórico de acidente automobilístico e lesão cerebral traumática grave há 45 anos. A RNM SWAN mostrou hipossinal nas cisternas da base e na superfície cerebelar e as imagens em T1 evidenciaram a presença de esclerose hipocampal esquerda.

Palavras-chave: siderose, imagem por ressonância magnética, epilepsia.

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INTRODUCTION

Superficial Siderosis (SS) is a rare condition caused by the deposition of hemosiderin along the pial and subpial structures of the central nervous system due to low-grade bleeding into the cerebrospinal fluid (CSF). SS is mainly caused by subarachnoid hemorrhage due to idiopathic bleeding, which represents around 35% of the cases[1-4]. Hemosiderin deposition mainly affect the upper vermis, the cerebellar leaves, the frontal and temporal lobes, the ventricular epithelium, choroid plexus epithelium, brain stem, spinal cord, nerve roots and cranial nerves I and VIII[5].

The clinical syndrome is characterized by sensorineural hearing loss, cerebellar ataxia, signs of pyramidal tract dysfunction and dementia[6]. For a long time the diagnosis of superficial siderosis could only be made by post-mortem,[1,7] but with the modernization of radiological imaging methods, including blood-sensitive sequences, it is possible to diagnose SS using Magnetic Resonance Imaging (MRI)[7,8].

The aim of this study is to describe a case of superficial siderosis with mesial temporal sclerosis and to evaluate SS etiologies, imaging characteristics and features of diagnosis.

CASE REPORT

A 65-year-old man came to the neurology outpatient clinic reporting episodes of seizures characterized by behavioral arrest and oral automatism (chewing and sucking) accompanied by aphasia, lasting 3 to 5 minutes. He maintained consciousness during the episodes and had no post-ictal clinical manifestation. Episodes occurred on average once a day for 3 months. He had a history of car accident with severe traumatic brain injury 45 years ago without a neurosurgical approach at the time. He reported being a 40 pack-year smoker but denied any comorbidities. On physical examination he presented a broad-based gait, dysmetria and bilateral dysdiadokinesia, more prominent on the right.

Sleep electroencephalogram did not show epileptiform paroxysms. Cranial MRI revealed hypersignal in T2/FLAIR in periventricular and cerebellar white matter which suggest areas of gliosis, associated with marked cerebellar atrophy (fig. 1). In the images weighted in magnetic susceptibility, a hyposignal was highlighted in the basal cisterns and in the cerebellar surface (fig. 1: F, G, H). The left hippocampus had distorted morphology and a slight hypersignal in T2 / FLAIR images. Based on these findings the patient was diagnosed with superficial siderosis associated with mesial temporal lobe epilepsy due to left hippocampal sclerosis. Cervical, thoracic and lumbar spine MRI did not characterize spinal cord injuries or superficial siderosis in this topography. The patient was treated with lamotrigine 75mg 12/12h with a 70% reduction in the frequency of epileptic seizures.

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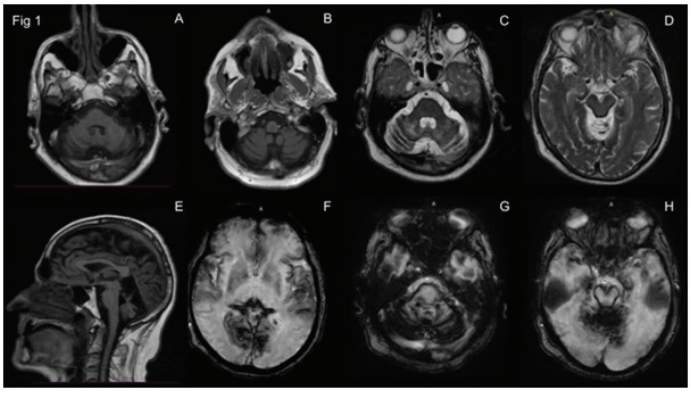

Figure 1. Prominent cerebellar sulci consist with diffuse cerebellar atrophy (A, B, E: T1WI). Hypointensity at the pial surface of midbrain, surface of cerebellum and cerebellar atrophy (C, D: T2WI; F-H: SWAN).

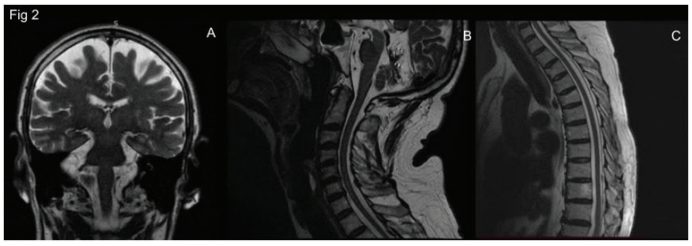

Figure 2. Discrete hypersignal and morphological distortion in the left hippocampus consist with mesial temporal sclerosis (A: T2WI). Absence of injuries or superficial siderosis (B, C: T2WI).

DISCUSSION

SS is an uncommon and often unrecognized disorder[9]. The only established causes of SS are chronic or intermittent extravasations of blood into the subarachnoid space [2,6,10]. Various etiologies can cause SS and they can be classified into brain and spinal disorders (Table 1). The main brain disorders that cause SS are CNS tumors, such as craniopharyngioma, paraganglioma and ependymoma, amyloid angiopathy, head trauma and history of brain surgery. Spinal cord disorders can also cause SS and the main causes are arteriovenous fistula, neural tube defect, spinal meningeal diverticulum and dural ectasia. Despite that, idiopathic bleeding is still the main cause of subarachnoid bleeding [4,10,11].

Idiopathic bleeding is the main cause , but it also can be caused by a central nervous system (CNS) tumor, arteriovenous malformations, meningocele, history of trauma or CNS surgery and is also common in patients with advanced cerebral amyloid angiopathy [4,10,11]. Spontaneous intracranial hypotension has also been associated with superficial siderosis[11].

Studies report that SS pathogenesis occurs from the breakdown of erythrocytes followed by the release and deposit of hemosiderin throughout the arachnoid and pia mater. Accumulation of blood products causes the symptoms through an inflammatory process, including reactive gliosis, neuronal loss and demyelination,[10] especially in the posterior region of the cerebellum and cranial nerves [2,10]. The most affected nerve is the vestibulocochlear, probably due its long glial segment that predisposes it to iron deposition[9].

Table 1. Causes of Superficial Siderosis.

Etiologies of superficial siderosis
Brain
CNS tumor (craniopharyngioma, paraganglioma e ependymoma)
Amyloid angiopathy
Hemorrhagic transformation of cortical infarction
Reversible cerebral vasoconstriction syndrome
Cerebral venous thrombosis
Head trauma
History of brain surgery
Spinal/medullary
Arteriovenous fistula
Neural tube defect
Spinal meningeal diverticulum
Dural ectasia
Intradural herniated disc
Brachial plexus / root avulsion injuries
Spinal meningeal diverticulum
Spinal cavernous malformation in the vertebral body
Back trauma
Others
Drugs (anticoagulants and antiplatelet agents)
Idiopathic

Although clinical condition can vary depending on hemosiderin distribution, almost all reported cases of SS were diagnosed in patients with clinical symptoms that are now considered typical presentation for the disease: cerebellar ataxia, progressive sensorineural hearing loss, cognitive impairment and pyramidal signs. Usually, the symptoms develop slowly and progressively [12,13].

MRI is largely more sensitive to hemosiderin deposits than computerized tomography (CT), therefore MRI is the diagnostic procedure for the diagnosis of SS [9]. Brain or spine intra-arterial digital subtraction angiography and surgical exploration can be performed as well[7]. SS appearance on MRI is characterized by signal loss on T2 gradient recalled echo (GRE) and susceptibility-weighted imaging (SWI) sequences following the gyral cortical surface in a curvilinear pattern[1]. Typically T2 hypointensity can be consistently seen outlining the cerebellum, brainstem, and pial surface of the cord. These imaging findings can also be seen in the sylvian, interhemispheric fissures and hemispheric convexities [9]. Although T2 GRE has a high sensitivity, SWI sequence is superior to identify microbleeds [1].

Even with MRI the diagnosis of SS can still be missed because the findings in images can be overlooked, once the abnormalities follow the contour of the brain. Frequently an experienced neuroradiologist is required. In some cases cerebrospinal fluid (CSF) evaluation can be useful and may reveal xanthochromia or red blood cells. Because it is an invasive procedure, CSF puncture is only performed when SS is suspected but imaging diagnosis is not conclusive [9].

Iron chelators, such as deferiprone can be used in some cases[14]. However, patients with SS are usually treated with symptomatic therapies, since there is not an evidence based treatment for this condition. When the source of bleeding is identified it is possible to perform surgical removal. Cochlear implants can be indicated for patients with progressive sensorineural hearing loss. Antiepileptic drugs may also be used in patients with SS and comorbid epilepsy [14].

CONCLUSION

SS is an uncommon and often unrecognized disorder, characterized by cerebellar ataxia, progressive sensorineural hearing loss and pyramidal signs. MRI is the diagnostic procedure of choice because of its high sensitivity to hemosiderin depositions. SS is a challenging condition for clinical practitioners due to its defying diagnosis and nonspecific symptomatology.

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