BACKGROUND

Superficial siderosis (SS) of the Central Nervous System (CNS) is a rare entity characterized by the deposition of hemosiderin in the leptomeninges initiated by chronic and recurrent bleeding into the subarachnoid space, in most cases, remaining sub-clinical and for long periods of time. The cases described in the literature are associated with tumors, aneurysms, arterial-venous malformations, post-surgical changes, traumatic cervical and brachial plexus injuries. However, the cause of bleeding remains unclear in 40-50% of cases.

CASE REPORT

We present a case of a 61 year old woman with a diagnosis of SS. No family history was known for CNS tumors or degenerative ataxias. It all started in 2008 with the insidious development of gait difficulties and non preference falls. To elucidate the reason behind these complaints, the first investigative approach was to perform CNS imagiology study that at the time shows what was described as an apparent cystic formation in the left lateral ventricle (LV) and probable signs of hemosiderin deposition. She was submitted to a surgical intervention in April 2009 by interhemispheric transcallosal approach that reveals old hemorrhagic coloration at the cingulum, xanthochromatic liquor, and a voluminous expansive lesion of the left LV with corpus callosum infiltration. Facing these findings it was only performed extemporaneous biopsy that shows what appears to be normal cerebral and subcortical tissue; complete absence of postural reflexes, osteotendinous hyperreflexia, vertigo, nausea, emesis, frontalis contracture, anosmia and limitation of upward gaze. It was performed an MRI scan that clearly shows signs of hemosiderin deposition compatible with the SS.

DISCUSSION

In most patients with adult-onset progressive ataxia, the condition manifests without an obvious familial background. The correct diagnosis of such patients remain a challenge, because almost the entire spectrum of non-genetic and genetic causes of ataxia has to be considered. A wide range of potential causes of acquired ataxia exist, including chronic alcohol use, various other toxic agents, immune-mediated inflammation, vitamin deficiency, chronic CNS infection, and chronic leptomeningeal deposition of iron leading to SS. It is a well known that hemosiderin arising from the bleed causes oxidative cellular damage leading to symptoms. Even in the absence of an inciting event, years of chronic bleeding can precede clinical manifestations. Patients often present after the fourth decade. The most common clinical presentation is slowly progressive cerebellar ataxia, which is often associated with hearing impairment. Sensorineural hearing loss may be the first symptom. A myelopathic presentation is also described. Pyramidal and sensory signs, and bladder dysfunction are common and may relate to brainstem or spinal cord involvement. Anosmia or hyposmia are common but underreported, mainly because olfaction is rarely tested.

In recent years SS has become an increasingly common diagnosis due to advances in imaging. Prior to MRI technology, the diagnosis of siderosis was made at autopsy. MRI is the investigation modality of choice, though (particularly in the early stages) the findings are subtle and a high index of suspicion is necessary to permit an early diagnosis. A dark lining of the meningeval surfaces on T2 images is the signature of the disease and is due to the paramagnetic nature of hemosiderin. T2 GRE/T2* confirm the diagnosis. Imaging of the entire neuraxis is indicated to localize a potential bleeding source.

One last word to CT, though head CT results are generally unremarkable, cerebellar atrophy or a clue to the potential cause of superficial hemorrhagic signs at several time stages. After surgery, there was a progressive exacerbation of the cerebellar ataxia, that leads in September 2009 to her admission in the H.U.C. neurology ward. At that time, she presented with incapacity for autonomous gait, severe cerebellar ataxia, complete absence of postural reflexes, osteotendinous hyperreflexia, vertigo, nausea, emesis, frontalis contracture, anosmia and limitation of upward gaze.

CONCLUSION

Although a rare entity, we should be aware of it, mainly in the imaging study in patients with deafness or ataxia. An extensive radiological examination sometimes is needed in order to localize the source of bleeding, that even the could remain unknown.

TECHNICAL SPECIFICATIONS

CT: Images - with a General Electric LightSpeed VCT

MRI: images obtained in a Siemens, Magnetom Symphony Maestro Class, 1,5 T T1WI, DP/T2 WI, T2WI GRE/T2*, FLAIR.

BIBLIOGRAPHY