

Superficial Siderosis Following Posterior Fossa Exploration

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Abstract

A 70 year old lady presented with symptoms and signs suggesting lesions in the vestibulocochlear nerve, the cerebellum and brainstem. Features on magnetic resonance imaging of the brain led to the diagnosis of superficial siderosis. This condition is a very rare neurodegenerative disorder characterised by deposition of haemosiderin in several areas of the nervous system. This occurs as a result of recurrent bleeding in the subarachnoid space. Various sources of chronic bleeding have been implicated, including dural defects, neoplasms or arterio-venous malformations. Whilst presenting symptoms depend on the site of haemosiderin deposition, there is a predilection for the eighth cranial nerve, the cerebellum and brainstem. The investigation of choice is magnetic resonance imaging of the brain, and linear hypointensity is pathognomonic. Early identification of this condition will obviate the need for further, extensive investigation of the patient's symptoms. It will also enable the physician to search for an underlying, potentially treatable cause. Our patient gave a history of posterior fossa exploration half a century prior to her current presentation, suggesting the presence of a dural defect as the cause of this disorder.

Keywords: Superficial siderosis; Ataxia; Sensorineural deafness; Neurodegenerative; Haemosiderin

Introduction

Superficial siderosis is a very rare neurodegenerative disorder characterised by deposition of haemosiderin in several areas of the nervous system. Early identification of this condition will obviate the need for further, extensive investigation of a patient's symptoms. Moreover, the focus must be on the search for an underlying aetiological agent, as this may be amenable to correction. We present one such case of this disorder.

Case Presentation

A 70 year old lady presented with recurrent falls. She described a change in gait which had been steadily progressing over two years. She also complained of deafness from her right ear. She had a past history of posterior fossa exploration in 1962, at 18 years of age, when she had presented with mild ataxia. The surgery had revealed herniation of the cerebellar tonsils but no other suspicious lesions. Her symptoms had remained stable for several decades until she presented with her current, disabling complaints. She also suffered from hypertension and hypothyroidism, both controlled on treatment. A neurological



Figure 1: Superficial siderosis of the midbrain. The rim of hypointensity around the midbrain (arrow) is a pathognomonic feature of superficial siderosis.



Figure 2: Superficial siderosis of the cerebellum. There is a predilection for the deposition of haemosiderin along the cerebellar folia (arrow) due to the large quantity of Bergmann glia, where ferritin synthesis is prevalent.

examination revealed bilateral cerebellar signs, with nystagmus and dystaxia. Reflexes were brisk, and there was increased tone in all limbs. Her gait was spastic-ataxic in nature. Examination of the cranial nerves revealed decreased hearing from the right ear; this was sensorineural.

Magnetic resonance (MR) imaging of the brain was organised to investigate these signs. Axial T2 weighted images showed linear hypointensity lining the midbrain (Figure 1), pons and cerebellar folia (Figure 2). Gradient echo imaging revealed blooming artefact along the affected areas in keeping with the presence of haemosiderin. These findings suggested a diagnosis of superficial siderosis. An audiogram

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was organised, which revealed the presence of bilateral sensorineural hearing loss, worse on the right side. Other confirmatory tests, such as a lumbar puncture, were deemed unnecessary in view of the pathognomonic features seen on MR imaging. While the history of craniotomy suggests the presence of a dural defect as a cause for the siderosis, this can only be postulated since MR imaging of the whole spine did not reveal a causative lesion for this condition. The patient opted against further investigation for the cause of the lesion. Our patient was offered physiotherapy and occupational therapy, as well as assessment by an audiologist.

Discussion

Superficial siderosis is a very rare neurodegenerative disorder which follows deposition of haemosiderin along the meninges, leading to neuronal injury [1,2]. This occurs as a consequence of recurrent, occult, bleeding into the subarachnoid space.

Estimates in 2006 indicated the presence of less than 270 cases worldwide [3]. Increasing use of MR imaging over the years has enabled the detection of presymptomatic cases, hence the true incidence of this condition cannot be determined with certainty [4]. There appears to be a male predilection, with a male-to-female ratio of 3:1 [1]. Many cases have been associated with vascular neoplasms, such as a spinal ependymoma [5] or teratoma [6]. Cerebral arteriovenous malformations [7] or cavernous malformations have also been implicated. In fact, any lesion which results in prolonged bleeding can potentially result in superficial siderosis. Dural defects, whether congenital or following trauma or surgery, have been described in 47% of cases according to a 1995 survey [1]. These have the potential to lead to an intra-spinal fluid filled collection, a possible source of recurrent bleeding. Cervical nerve root avulsion similarly has been implicated as an aetiological agent [8]. A history of trauma or intradural surgery should thus be considered in all cases of superficial siderosis [9,10]. Cerebral amyloid angiopathy has also been linked with haemosiderin deposition associated with this neurological condition [4]. 46% of cases are idiopathic [1]. The most likely source of bleeding in our index patient is a dural defect following the posterior fossa exploration carried out several decades previously. The presence of a fluid-filled collection in the spinal canal on MR imaging has often been described in such cases [4]. This lesion is not apparent in our patient; we thus conclude that the likely dural defect is small, albeit significant.

Once blood appears in the cerebrospinal fluid, it is broken down into various components including haem, which will eventually result in storage as ferritin [11]. The presence of excess intrathecal iron overloads this pathway, and is deposited in neuronal tissues. This prompts the formation of reactive oxygen species, with subsequent neuronal injury and myelopathy [12].

The predilection of superficial siderosis for the superior vermis and crests of the cerebellar folia can be explained by the high incidence of Bergmann glia, in which ferritin synthesis is extensive [11]. While the eighth cranial nerve also has an extensive microglial segment, its long course inside the subarachnoid space furthermore contributes to its susceptibility [12]. Other preferred regions include the brainstem, the basal frontal lobe and temporal cortex, spinal cord and nerve roots, as well as the first cranial nerve [12].

Presentation varies according to the sites affected by the deposition of haemosiderin. 95% of patients develop gradual sensorineural hearing loss, which is often bilateral [1]. 88% of patients cerebellar dysfunction, while 76% have pyramidal signs [1]. Less common symptoms include

dementia, bladder dysfunction, sensory deficits and other cranial nerve involvement [1]. While MR imaging has now enabled the detection of asymptomatic cases, presentation usually occurs after 40 years of age [1,2]. MR imaging is the investigation of choice, and has been shown to be superior to computed tomography (CT) imaging in the determination of the underlying cause [10]. T2-weighted MR imaging, in particular, reveals linear hypointensity outlining the brainstem, cerebellar folia and the pial surface of the cord [2,12]. Similar changes can sometimes be seen in the Sylvian fissures, the cerebral sulci and the cauda equina roots.

Findings can be very subtle, however, once identified, are pathognomonic [12]. Gradient-echo T2-weighted images have a superior rate of detection [4]. The degree of imaging abnormality does not always correlate with the degree of clinical impairment. Imaging of the whole neuraxis should be performed in all cases to search for a bleeding source [12]. MR imaging and CT myelography have often revealed an intraspinal fluid-filled collection, such as a meningocele or pseudomeningocele, a neoplasm or vascular abnormality [4]. Nerve root avulsions, usually in association with brachial plexus injury, have also been identified. The use of cerebral and spinal angiography in determining the aetiology of superficial siderosis has been described in the literature. These techniques may, however, not always prove useful, due to the indolent nature of the bleeding [4].

Despite extensive investigation, a cause is often not identified [12]. No abnormalities were identified on MR imaging in our patient. A lumbar puncture may reveal xanthochromia and elevated red cells, as well as high iron and ferritin levels in the cerebrospinal fluid (CSF) [1]. An audiogram can be organised if there is suspicion of vestibulocochlear nerve involvement; this will often demonstrate sensorineural hearing loss [8]. Unfortunately there is no proven direct treatment for established siderosis [13]. The use of iron chelating agents such as desferrioxamine and deferiprone is being studied. A recent study showed reduced hemosiderin deposition on MR imaging with deferiprone, however further studies are required to prove the efficacy and safety of this drug [13,14].

Early identification of a causative lesion is key to management of this condition, as this may be amenable to intervention [4]. Longitudinal intra-spinal fluid filled collections following a dural defect may be amenable to repair [5]. Exploration and repair of a pseudomeningocele due to cervical nerve root avulsion can also be undertaken [10]. A variety of treatment options exist for vascular tumours and arteriovenous malformations. These include excision or cauterization of abnormal vasculature [2]. Several recent case reports have described a positive outcome in patients treated with such techniques [2,8,10]. It has been postulated that they may actually halt progression of this disorder, with a significant decrease in the presence of red blood cells on CSF examination seen in patients who have had such surgery. While these results are very encouraging, it is recognised that the natural history of this condition necessitates a longer follow-up for these cases, in order to establish the actual outcome of such promising interventions [12].

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