

Neurological image

Cerebellar atrophy revealing superficial siderosis

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A 57-year-old man without medical history presented with progressive gait unsteadiness and hearing loss since the age of 54. Clinical examination showed ataxia and dysarthria together with bilateral profound sensorineural hearing impairment. MRI revealed extensive cerebellar atrophy (predominant in the superior vermis) on T1-weighted images (Fig. A), together with a hypointense rim along cerebral convexities, sylvian and interhemispheric fissures, brain stem, cerebellar folia, and the eighth cranial nerve on T2- and gradient-echo T2-weighted images (Fig. B, C, and D), typical of superficial siderosis of the CNS. Cerebral MRA, spinal cord MRI, and lumbar puncture showed no abnormalities.

Superficial siderosis of the CNS results from haemosiderin deposition in the subpial layers of the brain and the spinal cord causing progressive parenchymal damage. The commonest neurological manifestations are progressive cerebellar ataxia and sensorineural hearing impairment. Cerebellar dysarthria and gait ataxia are frequently seen. Other possible clinical findings include sensorimotor symptoms, urinary symptoms, olfactory dysfunction (due to haemosiderin deposition along cranial nerve I), and cognitive impairment. A history of severe headache, indicative of subarachnoid haemorrhage, is often absent.

Prior trauma or intradural surgery is often identified as a risk factor. In search for a source of chronic or recurrent subarachnoid bleeding, several intracranial and spinal abnormalities have been reported: CSF cavities (e.g. meningocele, pseudomeningocele, cavity remaining after neurosurgery), plexus and root pathology (e.g. avulsions), tumours, vascular abnormalities, and amyloid angiopathy. Ferritin synthesis – the CNS tissue response to CSF haemoglobin and haemoglobin breakdown products – is thought to be neuroprotective by binding iron released by red cells. When this reserve is exhausted, tissue damage occurs. Accelerated ferritin synthesis in the cerebellar Bergmann glial cells may account for the cerebellar predilection. The vulnerability of cranial nerve VIII

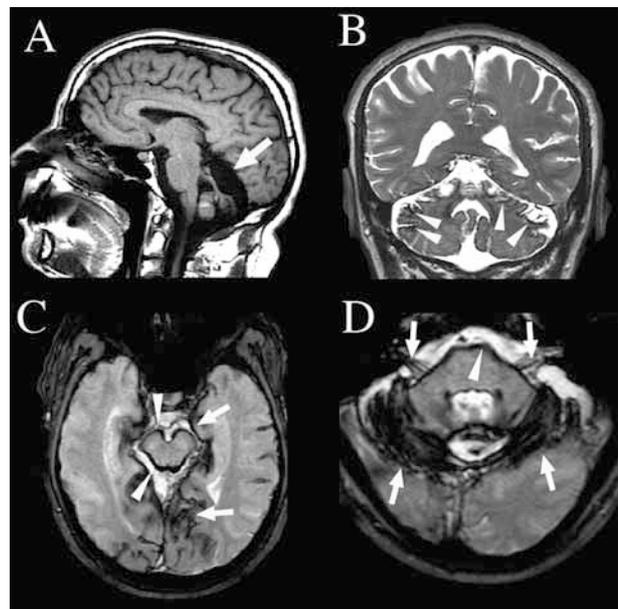


FIG. 1. — (A) T1-weighted MRI showing extensive vermis atrophy, especially involving its superior part (arrow). (B) A hypointense rim around the cerebellum (arrowheads) is seen on T2-weighted sequences. (C, D) Echo-gradient T2-weighted imaging shows hypointense haemosiderin deposition around the surface of the temporal and occipital cortex (C, arrows), the brainstem (C and D, arrowheads), the cerebellum (D, lower arrows), and along both eighth cranial nerves (D, upper arrows).

may be explained by its long glial segment that predisposes it to iron deposition.

CT rarely demonstrates haemosiderin deposition, seen as a hyperdense rim around the brain. Haemosiderin deposition can be seen on T2- and echo-gradient (with higher sensitivity for haemoglobin breakdown products) T2-weighted MRI as a hypointense rim on the surface of cerebral convexities, cortical fissures, brainstem, cerebellum, spinal cord, and nerve roots. Associated brain and cerebellar atrophy (preferentially involving the superior part of the vermis, as observed in our patient) is frequently seen. Xanthochromia, red blood cells, and elevated protein content can be found on CSF

examination. Entire CNS imaging is indicated to localize the potential bleeding source. Early diagnosis and intervention directed at removal of the bleeding source may prevent progression. However, despite extensive investigations, the bleeding cause remains often undetermined.

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