Selective loss of Purkinje cells in transverse and sigmoid dural arteriovenous fistulas

Report of two cases

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INTERRACRANIAL dural AVFs comprise 10 to 15% of intracranial vascular malformations. Transverse–sigmoid sinus dural AVFs are the most common and present with a variety of symptoms resulting from venous hypertension, ischemia, and hemorrhage. In several studies histological changes have been reported in the lesions of fistulas, including changes affecting the venous sinus and adjacent vessels. Nevertheless, studies of brain tissue have rarely been documented. We studied two patients with transverse–sigmoid dural AVFs and report the histological findings in tissue from the affected cerebellar cortices obtained during biopsy procedures.

Case Report

Case 1

This 71-year-old man presented with sudden-onset nausea and vomiting. Although the initial CT scan revealed no significant abnormalities, the patient suddenly lost consciousness 2 weeks later. The CT scan obtained at that time demonstrated subarachnoid hemorrhage (Fig. 1A). The patient showed confusion, although no other deficits, including cerebellar dysfunction, were detected on neurological examination. Angiographic studies revealed a transverse–sigmoid sinus dural AVF fed by the left middle meningeal and ascending pharyngeal arteries, and branches of the occipital artery (Fig. 1B). The AVF drained to the left transverse sinus, straight sinus, right occipital sinus, and the bilateral bridging veins of the occipital lobe. Venous hypertension caused an ischemic condition severe enough to cause selective neuronal damage in the cerebellum. Neuroimaging scans demonstrated a decrease in cerebral blood flow, including flow through the cerebellum. Venous hypertension caused an ischemic condition severe enough to cause selective neuronal damage in the cerebellum.

Case 2

This 73-year-old man received a diagnosis of Parkinson disease after presenting with dementia and resting tremor of...
both upper extremities. His worsening dementia resulted in an apallic state and the patient was admitted to our hospital. Neurological examination showed no cerebellar dysfunction. Admission T2-weighted MR images showed hyperintense lesions bilaterally in the thalamus (Fig. 2A). Cerebral angiographic studies revealed a dural AVF in the transverse–sigmoid sinus fed by the branches of the left ECA and the left posterior meningeal artery, with draining to the partially thrombosed sigmoid sinus (Fig. 2B). Retrograde venous drainage was observed into the straight sinus, right basal vein, vein of Labbé, and left occipital cortical vein. An IMP–SPECT scan demonstrated significant flow reduction in the left occipital lobe, cerebellar cortex, basal ganglia, and thalamus (Fig. 2C). The patient underwent surgical occlusion of the draining venous channels and resection of the left transverse sinus. The left cerebellar cortex and occipital lobe appeared black with petechial hemorrhages, similar to findings in Case 1 (Fig. 3A). We obtained biopsy samples of cerebellar cortical tissue for histological examination. Follow-up MR images obtained 10 days after surgery demonstrated improvement of the bilateral thalamic abnormalities. Six months later, no significant signal abnormality was evident in the thalamus. The left cerebellum and occipital lobe demonstrated increased CBF on postoperative IMP–SPECT scans. The resting tremor disappeared completely, and dementia had greatly improved 6 months postsurgery.

**Histological Findings in the Cerebellar Cortices**

Histological findings for both patients were essentially similar (Fig. 3B–D). The cortices of the cerebellar hemisphere showed marked dilation and congestion of the parenchymal veins. There were multiple small foci of breakdown hemorrhage in the parenchyma surrounding congestive veins (Fig. 3B–D). The cortical structures, including the granular and molecular layers, were preserved. The Purkinje cells, however, showed hypoxic changes, such as shrinkage of the cell body and cytoplasmic eosinophilia with nuclear pyknosis, and there was a marked decrease in the number of cells (Fig. 3C and D). Bergmann glia in the Purkinje cell layer were also slightly increased.

**Discussion**

Clinical manifestations of transverse–sigmoid sinus dural AVFs include pulsatile tinnitus, headache, visual disturbance, seizure, hemorrhage, cerebellar ataxia, dementia,
and so on.\textsuperscript{5,10–12} Recently, transverse–sigmoid sinus dural AVFs have received attention as a potential cause of reversible dementia.\textsuperscript{13} Case 2 in this report provides an example of the neurological and neuroimaging course of that vascular dementia. Bilateral hyperintense thalamic hyperintense lesions on T\textsubscript{2}-weighted MR images were apparent before treatment and disappeared after surgery, concurrently with improvement of the dementia.\textsuperscript{5} This sort of dementia or change of mental status may be due to encephalopathy caused by venous hypertension resulting from transverse–sigmoid sinus dural AVFs.\textsuperscript{7} In contrast to the neuroimaging reports, histological evidence of encephalopathy has rarely been documented.\textsuperscript{2,7,8} Hurst, et al.,\textsuperscript{7} reported autopsy findings in a patient with a transverse–sigmoid sinus dural AVF. They documented marked dilation of the superficial and intraparenchymal veins, gliosis of the central semiovale, petechial hemorrhage, and so on. There are no reports, however, that describe the histological changes of the infratentorial structures.

In this report we have described the pyknotic change and selective loss of Purkinje neurons in the cerebellar cortex affected by a dural AVF. The granular and molecular layers were well preserved. Although the Purkinje cells are subject to age-related depletion in the human brain, they do not show a total loss without the presence of a pathological condition. They are known to be highly vulnerable to ischemic insult in the cerebellum as well as the hippocampus in the cerebrum. Other histological findings, such as the marked dilation of parenchymal vessels and petechial hemorrhage, indicated that loss of the Purkinje neurons was associated with moderate ischemia caused by venous hypertension. This pathological hypothesis is thought to be the same mechanism by which the early loss of anterior horn cells in the spinal cord is caused by the venous congestion in Foix–Alajouanine syndrome (spinal dural AVFs).\textsuperscript{1,3,4} Recent studies in which positron emission tomography was used demonstrated decreased CBF in the territory of retrograde venous drainage in dural AVFs.\textsuperscript{9} In our cases, IMP–SPECT also revealed reduction of cerebellar CBF. These observations indicate that venous hypertension due to transverse–sigmoid sinus dural AVFs may provoke an ischemic condition severe enough to cause selective neuronal damage in the cerebellum.

References


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