Anomalous Intracranial Venous Drainage Associated with Basal Ganglia Calcification

**SUMMARY:** We describe the neuroradiologic findings in a 7-year-old boy with anomalous intracranial venous drainage and cerebral calcification. CT scans demonstrated that his scalp mass was a plexus of scalp veins filled through the emissary foramen, and there were cerebral calcifications. Angiography revealed bilateral sigmoid sinus atresia with most of the intracranial venous drainage via the prominent mastoid emissary veins into dilated scalp vein. The possible relationship between cerebral calcification and anomalous intracranial venous drainage is discussed.

CASE REPORT

Z. Chen
H. Feng
G. Zhu
N. Wu
J. Lin

Normally, most of the cerebral venous drainage collects ultimately into the transverse and sigmoid sinuses of the skull base, and the pathway from the dural sinuses of the posterior fossa into the internal jugular veins is anatomically obvious through the jugular foramen and numerous connections with the vertebral plexus.1,2

In this article, we present a very rare case of bilateral sigmoid sinus atresia with most of the cerebral venous drainage through the prominent mastoid emissary vein to a plexus of dilated scalp veins, presenting as a posterior auricular mass lesion and, more unusually, with a combination of basal ganglia and cerebral calcification.

Case Report

The patient was a 7-year-old boy who presented with a tender mass lesion in the left posterior auricular region and with left tinnitus. His parents first noticed the mass 2 years ago by accident, and the patient noticed left tinnitus 1 year ago. The mass had grown slowly during the past 2 years. The patient did not report nausea, vomiting, headache, or pain from the lesion. On his admission, physical examination revealed a fluctuant mass approximately 3 cm in diameter. Very soft thrill and bruit were noted on the surface of the mass. The mass disappeared when manually compressing the skull depression under the center of the mass. Physical examination showed no specific neurologic deficits. Neuropsychological testing, including Chinese translation of the Wechsler Intelligence Scale for Children (Revised) and Chinese translation of the Wechsler Memory Scale for Children, suggested no impairment. Blood analyses for copper, ceruloplasmin, ferritin, calcium, and phosphorus levels, as well as thyroid and parathyroid profiles, were normal. Neurophysiologic studies, including an electroencephalogram and a determination of brain stem auditory-evoked potentials, were generally normal.

A CT scan suggested that the mass was a plexus of dilated scalp veins filled through the prominent mastoid emissary foramen (Fig 1). There were also diffuse calcifications on the bilateral basal ganglia and subcortical white matter in CT scan (Fig 2). Carotid artery angiography revealed grossly anomalous intracranial venous drainage (Fig 3). There was remarkable stenosis of the left transverse sinus and atresia of both sigmoid sinuses with florid collateral scalp vein drainage via the prominent mastoid emissary vein. Additionally, after manual compression of the mastoid emissary foramen, there was no other obvious collateral drainage passageway. On the basis of these findings, together with the normal growth and life of the patient, he was discharged with close follow-up.

Discussion

Oclusion of the bilateral transverse-sigmoid sinus is rare, which may be idiopathic or caused by various disorders such as thrombosis and syndromic craniosynostoses.3-5 Anatomic and radiologic studies6 have shown extensive extracranial collateral drainage networks in the posterior fossa dural sinus via condylar/mastoid/occipital emissary veins, diploic veins, and marginal/occipital sinuses, and these studies have shown that the morphologic changes in the posterior fossa dural sinuses, emissary veins, and jugular bulb are closely related to development of the brain, shifts to postnatal types of circulation, and to postural hemodynamic changes. A MR venography study7 also showed multiple variants of intracranial venous drainage, including a patient absent of both transverse sinuses whose occipital sinuses were present on both sides, serving as alternative drainage pathways from torcular herophili to internal jugular veins. The etiology of the bilateral sigmoid sinus occlusion in this patient was unknown. He had no history of mastoiditis, meningitis, or head trauma. Also no sign of craniosynostoses was seen on CT scans. Thus, despite the lack of initial angiographic findings, we presumed that our patient’s mass was from developmental morphologic anomalies.

When there is atresia at the bilateral sigmoid sinus or transverse sinus, venous drainage may occur via 2 routes: 1) by collateral emissary veins or, if absent, by the marginal/occipital sinus; or 2) by anterior drainage into the Galenic system or into the superior sagittal sinus and cavernous sinus.6,9 Cases with anomalous intracranial venous drainage present various manifestations depending on the different drainage routes. Tech et al9 reported a case of bilateral sigmoid sinus hypoplasia/aplasia with most of the cerebral venous drainage occurring through the right cavernous sinus and diploic emissary veins. The patient presented with swelling of the right eye and bluish discoloration over the periorbital region, mimicking a cavernous arteriovenous fistula. In our patient, anomalous intracranial venous drainage collected into dilated plexus of scalp veins via unilateral prominent mastoid emissary veins, mimicking a posterior auricular mass lesion.

Our patient was even more unusual in that he had bilateral (almost symmetrical) calcification involving basal ganglia and cerebral white matter. Brain calcification could be autosomal-dominant, family based, or secondary to a variety of developmental, metabolic, infectious, and other conditions.10 In the
In summary, our case was unusual because the anomalous intracranial venous drainage in our patient was mainly through the unilateral prominent emissary vein and dilated scalp veins filled through the mastoid emissary foramen.
scalp veins, and he presented with a scalp mass lesion. We think that anomalous intracranial venous drainage associated with intracranial calcification as found in our patient has not been reported previously. Dilated collateral scalp veins should be included in the differential diagnosis of scalp lesions, especially before surgical procedures.

References