Cerebral venous drainage occurs primarily through the IJVs. However, external jugular pathways in the vertebral venous system can also contribute to cerebral venous drainage. We describe the case of a patient with hydrocephalus and Dandy–Walker variant who presented with spastic lower-extremity paraparesis that was associated with VP shunt failure. Further evaluation revealed the presence of bilateral occlusion of the IJVs and a distended epidural venous plexus within the cervical canal that had led to greater than 50% stenosis of the C2–5 spinal canal. This case provides an opportunity to further explore and characterize the physiological role of external jugular venous pathways in cerebral venous drainage in a patient with congenital hydrocephalus.

Case Report

History and Examination. This 36-year-old woman had a medical history significant for Dandy–Walker variant, hydrocephalus, and resection of a dorsal epidural spinal arachnoid cyst. She presented after suffering progressively worsening spastic lower-extremity paraparesis for 1 month. Her physical examination was notable for decreased lower-extremity strength that was more pronounced on the right side, limited range of motion in the lower extremities, 3+ deep tendon reflexes bilaterally, sustained clonus in the left ankle, and mild dysmetria of the right hand. The patient denied a headache and displayed no cranial nerve deficits. Magnetic resonance imaging of the brain and cervical spine revealed a Dandy–Walker variant with mild increase in ventricular size along with an enhancing area ventral to the spinal cord beginning at the level of the foramen magnum and causing significant compression of the cervical spinal cord (Fig. 1). No recurrence of the dorsal arachnoid cyst was observed. A 3D reconstructed CT angiogram of the head and cervical spine was obtained and revealed jugular veins that terminated bilaterally at the angle of the base of the skull and formed again around the angle of the mandible. Engorgement of the cervical venous plexus and subsequent compression of the thecal sac from C-2 to C-5 was also evident (Fig. 2). Thus the compression of the cervical

Abbreviations used in this paper: CT = computed tomography; IJV = internal jugular vein; VP = ventriculoperitoneal.
Cord was attributed to the engorged cervical venous plexus and not a neoplastic lesion.

Operation and Postoperative Course. Due to the change in ventricular size, the ventricular shunt system was revised. In addition, a cyst in the occipital horn of the right ventricle was fenestrated endoscopically. The patient did not suffer any complications during the postoperative period, and her myelopathy improved. Follow-up CT angiography of the head and cervical spine revealed a decrease in epidural venous engorgement compared with the CT study obtained before the shunt revision (Fig. 3). The patient did well for approximately 5 months but again began to develop myelopathic symptoms. She was reevaluated, and CT angiography of the head and cervical spine again revealed an engorged epidural venous plexus and 50% stenosis of the upper cervical canal (Fig. 4). The VP shunt was revised and the patient’s symptoms improved.

Discussion

To our knowledge, this is the first report of myelopathy due to an engorged venous plexus associated with VP shunt failure. Cerebral venous drainage appears to exit the cranial vault through the transverse sinuses into the cervical and intraspinal plexuses, which then drain into the vertebral veins bilaterally. The increased intracranial pressure caused by shunt failure in this case was sufficient to increase the venous outflow through the vertebral system via the emissary veins that drain the intracranial compartment. The increased venous flow through the cervical epidural plexus led to venous engorgement, spinal cord compression, and subsequent myelopathy. The findings on the CT angiogram support this theory by demonstrating effacement of the ventral subarachnoid space from C-2 to C-5 secondary to prominent epidural venous structures.

In normal circumstances, cerebral venous drainage occurs primarily via the IJVs. However, clinical evidence suggests that the vertebral venous system significantly contributes to cerebral venous drainage in circumstances in which the internal jugular venous flow is impeded.\textsuperscript{1,3,5,6–13} The vertebral venous system can be divided into an anterior internal segment consisting of an intraspinal epidural venous plexus and a posterior external paravertebral segment.\textsuperscript{2,8,9,11} These segments make up a valveless system that courses the entire length of the spinal cord.\textsuperscript{2,8,9,11} The posterior vertebral segment communicates superiorly with the dural venous sinus of the posterior cranial fossa via the emissary veins.\textsuperscript{2,8,9,11} These vessels include the condylar veins as well as the occipital and mastoid emissary veins, and all have multiple connections with the anterior condylar confluence, which connects the intracranial venous circulation and the vertebral venous system.\textsuperscript{11} The posterior vertebral segment also communicates with the anterior vertebral venous system via the intervertebral and basivertebral veins.\textsuperscript{11,12} Thus there are multiple anastomoses that...
connect the posterior cranial dural sinuses, the internal segment of the vertebral venous system, and the external segments of the vertebral venous system, thereby providing alternate pathways through which cephalic drainage can occur.

Previous investigators have attempted to characterize the external jugular pathways that contribute to cerebral venous drainage, particularly in instances in which the IJV flow is impeded. Doepp et al. showed that the contribution of the IJVs to cerebral drainage varies among individuals such that the percentage of total cerebral venous flow contributed by the IJVs can range from 6 to 72%. Moreover, patients undergoing radical neck resections with bilateral IJV removal have been shown to tolerate this procedure, and the cranial vault is still effectively drained. Baston estimated that the vertebral venous system at the level of the skull base has a total cross-sectional area larger than that of the IJVs, and Eckenhoff estimated that the vertebral venous system could tolerate a volume capacity of up to 1000 ml, thus illustrating the ability of the vertebral venous system to accommodate the entire cerebral venous drainage if needed. Additionally, bilateral compression of the IJVs was shown by Schreiber et al. to cause vertebral venous flow to increase by a mean of 82 ml/minute. Previous investigators have also shown that cerebral venous drainage through the IJV predominates in the supine position, but a postural change to the upright position causes a drastic decrease in IJV flow and a concomitant increase in vertebral venous flow. Moreover, Valdueza et al. proposed that the spinal architecture serves as a structural support to prevent collapse of the spinal epidural veins, resulting in a negative-pressure environment that accommodates increased flow through this region in the presence of decreased IJV flow. These data illustrate that less characterized external jugular pathways significantly contribute to cerebral venous drainage and predominate in instances during which drainage through the internal jugular venous system is diminished.

Authors of previous studies have suggested that the vertebral venous system should be adept at tolerating increased venous drainage when internal jugular venous flow is compromised, thereby raising the question as to why our patient became symptomatic. Gius and Grier examined the effects of bilateral radical neck dissection with removal of the jugular veins. Postoperative angiography performed in select patients revealed the development of collateral drainage pathways that flowed primarily through the vertebral venous plexus. Each of their eight patients was able to tolerate bilateral jugular vein excision; in addition, the authors were able to drain the cranial vault. This study established that the absence of IJVs should not preclude cerebral venous drainage. However,
there were physiological consequences associated with bilateral IJV resection including facial cyanosis and edema. These complications illustrate that although the vertebral venous system can drain the cranial vault, it may not be as adept as the jugular venous system at tolerating large increases in flow. In addition, events that alter or impede distal outflow through the vertebral venous system can further compromise the ability of this system to tolerate increases in venous flow. Of interest, our patient underwent resection of a thoracic arachnoid cyst 3 years prior to presentation. Removal of this cyst may have disrupted the distal outflow of the vertebral venous network in the thoracic spine, thereby decreasing its ability to tolerate substantial increases in flow. Therefore the pathophysiology of this patient’s presentation can be attributed to a primary anatomical predisposition and a secondary acquired insult that ultimately led to spinal cord myelopathy in association with VP shunt failure.

This case illustrates a fascinating clinical presentation of VP shunt failure that highlights the physiological importance of the external jugular pathways of cerebral venous drainage.

References


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