Spontaneous Cerebrospinal Fluid Leak Precipitating Catastrophic and Complicated Venous Sinus Thrombosis

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Background

Spontaneous intracranial hypotension (SIH) is an uncommon headache etiology, typically attributable to an unprovoked cerebrospinal fluid (CSF) leak. Although frequently benign, serious complications have been documented, including cerebral venous thrombosis (CVT). We report a highly complicated case of CVT attributable to SIH, and review the literature on this rare and dangerous condition.

Case History

A 43-year-old man presented with one week of non-postural headaches, dizziness, and nausea. While undergoing outpatient evaluation, he developed acute right-sided weakness, prompting presentation to our emergency department. Head CT identified CVT involving the superior sagittal sinus and adjacent cortical veins; IV heparin was initiated, and the patient was admitted to the NICU. Three hours later, he had a generalized tonic-clonic seizure, and repeat head CT demonstrated 3.1x3.8x3.1cm left parietal intracerebral hemorrhage, prompting initiation of lacosamide and hyperosmolar therapy. The hemorrhage subsequently expanded, which was managed via cessation of anticoagulation and escalation of hyperosmolar therapy. Digital subtraction angiography demonstrated near-normal filling of the venous system, with a “spot sign” suggesting likely contrast extravasation. In spite of maximal medical therapy, the patient developed herniation syndrome, and was brought emergently to the OR for exploratory craniotomy, clot evacuation, and EVD placement.

Upon elevating the bone flap, the dura was noted to be remarkably deflated despite clinical signs indicating mass effect and herniation, suggesting underlying CSF hypotension. Postoperatively, the patient was placed in 5° Trendelenburg with the EVD open at 15mmHg; positioning and the EVD were progressively weaned until he was stable for cerebral MRI, which identified a large T2-hyperintense ventral epidural fluid collection. CT-guided myelogram confirmed high-flow CSF leak at T1 adjacent to a large osteophyte, and the leak was treated with two separate injections of autologous blood and synthetic fibrin glue. The patient discharged to inpatient rehabilitation with moderate right hemiplegia, hemisensory loss, and partial left VI palsy, after which he mounted an extraordinary neurologic recovery, returning for three-month follow-up with mild sensory symptoms only.