CHRONIC POSTDURAL PUNCTURE HEADACHES
IN A CHILD WITH CROUZON SYNDROME

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Abstract

Children with Crouzon Syndrome (CS) may experience chronic headaches. Evaluation sometimes reveals an association of the headaches with elevations of intracranial pressure. Low CSF pressure as an etiology is generally not included in the differential diagnosis. We present our successful management with an epidural blood patch (EDBP) of a teenager with CS who presented to us with a three year history of recurrent headaches following a lumbar drain placement.

Introduction

Crouzon Syndrome (CS), also known as craniofacial dysostosis, is characterized by premature closure of the cranial sutures, midfacial hypoplasia, and exophthalmos. Thirty percent of affected individuals complain of frequent and severe headaches, which may or may not be associated with elevations of intracranial pressure (ICP)¹². A persistent cerebrospinal fluid (CSF) leak as a source of headaches in this population has not been previously described and is not generally considered in the differential diagnosis. A CSF leak may be a complication of accidental or intentional dural puncture during spinal anesthesia, epidural anesthesia, or surgical intervention, and can result in symptoms of intracranial hypotension including postdural puncture headache (PDPH), diplopia, and nausea, which may resolve with placement of an epidural blood patch (EDBP)³. This case report details the first documented use of an EDBP for treatment of chronic PDPHs in a child.

Case history

The patient is a 14-year-old female with CS, bipolar disease, and autism. Three years prior to presentation she underwent a lumbar drain placement for CSF diversion to manage CSF rhinorrhea. Subsequently, she had intermittent daily headaches with an orthostatic component, with exacerbations about twice weekly. She denied nausea, but experienced dizziness with the headaches. Ineffective pharmacologic remedies for the headache pain included olanzapine, topamax, topiramate, acetaminophen, and ibuprofen. Physical examination revealed no neurological deficits.

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A head computed tomography (CT) scan and brain MRI showed no etiology for the headaches. Given her history of prior durotomy and symptoms suggestive of PDPH, we performed an EDBP in advance of possible surgical dural repair.

Under conscious sedation, in the right lateral position, we inserted a 10 cm, 19 gauge Touhy needle at the L1-L2 interspace with loss of resistance to normal saline at 5 cm. Sterile autologous blood was injected to a total volume of 15 mL, with discontinuation once she complained of pressure and pain in her legs. She remained in a supine position for two hours following the procedure, with resolution of her headache upon leaving the hospital. To date, she has had no further complaints of headache similar to that pre-procedure.

**Discussion**

Patients with CS often receive evaluation and treatment of headaches, which may be associated with elevations of ICP. In this teenager, multiple normal MRIs ruled out any association of the headaches with hydrocephalus or other etiologies of increased ICP. Multiple practitioners treated her headaches with a variety of pharmacologic medications, as the headaches were not recognized as resulting from low CSF pressure and her prior dural puncture. Her combination of previous lumbar drain placement and subsequent orthostatic headaches were suggestive to us of PDPH, which we confirmed by successfully treating her headaches with placement of an EDBP.

Low CSF pressure headaches are typically orthostatic, causing pain due to traction on the intracranial and meningeal structures. Headaches and associated symptoms are usually self-limiting and resolve within 7 days or less. Most patients recover with conservative measures such as bed rest, hydration, and caffeine administration. Analgesic medications may be utilized, but are often ineffective. Following a recent dural puncture, placement of an EDBP has been a mainstay of treatment for PDPH if conservative measures fail, although randomized trials in support of this therapy are generally lacking.

It is now becoming more common to consider EDBP as a front line treatment in low CSF pressure headaches of any etiology, as patients may suffer for months, or even years, before effective treatment is initiated. In fact, a report in 1986 describes the successful treatment of an adult with chronic PDPHs using EDBP placed years after the initial dural puncture. When there is a history of a dural tear or puncture, no matter how remote, placement of an EDBP by an anesthesiologist should be considered for treatment of these headaches for both diagnostic and therapeutic purposes prior to proceeding to neurosurgical repair.

**Conclusion**

To the authors’ knowledge, this is the first reported case of successful treatment of chronic low CSF pressure headaches in a child using an EDBP. It demonstrates that even years after dural injury, anesthesiologists should consider placement of an EDBP in a patient when symptoms are consistent with those of a PDPH.
References
