

Is It All In His Head?

J Zimmer, J Bhatt, J Conway, M Edwards-Brown, D Sokol

Citation

J Zimmer, J Bhatt, J Conway, M Edwards-Brown, D Sokol. *Is It All In His Head?*. The Internet Journal of Pediatrics and Neonatology. 2005 Volume 6 Number 1.

Abstract

In this case report, of a teenage boy presents to your emergency room with headache and cranial nerve palsies. Differential diagnosis discussed herein include carotid artery dissection, complicated migraine headache, pseudotumor cerebri, subarachnoid hemorrhage and cavernous sinus thrombosis associated with sinusitis.

INTRODUCTION

You are half-way through your shift at the emergency room of a tertiary care pediatric hospital. The nurse hands you the chart of a 15-year-old boy transferred from his local hospital. The reason for transfer is progressively worsening headaches. The experienced nurse appears concerned, saying she has a feeling there's something wrong with this boy. You listen to her hunch and see the child immediately.

The patient is a previously healthy 15-year-old male. He appropriately acknowledges you when you enter the room, but he appears to be diaphoretic with significant pain. You immediately notice his disconjugate gaze. He's holding an emesis basin on his lap, and asks you to dim the lights. The mother relays the history since it hurts his head when he talks.

The boy's headaches began three weeks prior, when he was injured in gym class. While playing kickball, he was hit on the left side of his head by the ball causing the right side of his head to hit the bleachers. He did not lose consciousness and had no facial swelling, but he began having headaches afterwards. Three days after the accident a head CT was performed (read as normal), and he was started on prednisone for a presumed mild concussion. The headaches, however, continued.

Five days after the injury he woke up vomiting. He went to the local ER and head CT performed that day showed sinusitis. The oral steroids were stopped, and he was started on oral cephalexin, completing a ten day course. The mother was relieved, thinking there was finally an explanation for his persistent headache.

There was little time for celebration, however. The next

morning he began to develop swelling around his left eye. The following day, his eye was completely swollen shut. Another day passed, and he was able to open his eye partially. When his eye was open, his friends commented about his eye being turned inward.

Brain Magnetic Resonance Imaging (MRI) performed two weeks after the injury clearly showed ethmoid and sphenoid sinusitis with inflammation around the cavernous sinus. There was narrowing of the left carotid artery, presumed to be a carotid artery dissection which had occurred when he was hit by the kickball. He was started on daily aspirin for the dissection.

As the next two weeks progressed, his headaches were worse and no longer relieved by narcotics. He could not localize his headache, saying it hurt everywhere. He was vomiting daily and complained of double vision, a stiff neck and yellow rhinorrhea. He denies any fever during the past three weeks.

There was no family history of migraine headaches. Social history revealed no ill contacts and no recent travel. Although he has been caught smoking a few times, he denied illicit drug or alcohol use.

On exam tonight, his temperature is 37.6C, HR 80, RR 20, BP 150/84, WT 49 kg. The patient appears toxic, but rates his headache as "5 of 10". He has meningismus and a left-sided ptosis, but his pupils react. He cannot abduct his left eye. The remainder of his physical/neurologic exam is unremarkable.

Despite the numerous prior imaging studies, you send the patient for another MRI with Magnetic Resonance

Angiogram (MRA) and Venogram (MRV). While your patient is in the radiology department, you try to tie his symptoms and physical findings together: severe headache, vomiting, oculomotor (CN III) and abducens nerve (CN VI) palsies, meningismus.

Could carotid artery dissection explain these findings? His type of injury involving a sudden twist of the neck could certainly cause carotid artery dissection. Patients with dissection often have a complete or partial Horner syndrome (ptosis, miosis, and anhydrosis). You remember from your anatomy course years ago that Horner's syndrome is due to disruption of the sympathetic nerve fibers that run along the carotid wall. The hypoglossal nerve (CN XII) may also be affected due to proximity to the carotid artery, causing the tongue to deviate from midline. CNVI is rarely involved. Your patient has only ptosis, not the complete Horner's triad. Unilateral neck pain and headache are also seen with dissection. Your patient's neck pain seems more diffuse. You have a strong feeling that carotid artery dissection is not the answer.

Could this be the initial presentation of cluster headaches? Your patient would be the right age: cluster headaches typically present in males between the ages of 10-20 years. Cluster headaches are severe, unilateral, and retro-orbital. Patients may develop a watering eye, rhinorrhea, and a Horner syndrome. Individual headaches are usually less than three hours in duration. Cranial nerve findings and meningismus would not be expected.

Could this be a complicated migraine? Patients experiencing complicated migraines may have neurologic symptoms that persist in the absence of headache. These neurologic symptoms may even become permanent. Ophthalmoplegic migraine, or cranial neuralgia, is a complicated migraine with oculomotor nerve dysfunction. CN III is most frequently involved, followed by CN VI. Patients frequently experience blurred vision, which is not the case with your patient. Meningismus would not be expected. Regardless, patients with new-onset complicated migraines deserve a thorough evaluation to rule out other causes of headaches.

Could this be pseudotumor cerebri? Your patient is not the stereotypical overweight female. Yet you remember recently learning that there is no gender difference in adolescents experiencing pseudotumor cerebri, and only 30% are obese. You were also surprised to learn that papilledema is not a universal finding in children and adolescents with this diagnosis. Patients experience pulsatile headaches, nausea,

vomiting, and neck pain. Patients may have CN VI palsy due to increased intracranial pressure, and occasionally CN III palsy. You will remember to measure opening pressure when you perform the patient's lumbar puncture.

Could subarachnoid hemorrhage explain these findings? Subarachnoid hemorrhage (SAH) is often due to rupture of an intracranial aneurysm, but can also be caused by trauma or bleeding from an intracranial carotid dissection. Patients who are conscious present with severe headache, nausea, and vomiting. Patients may develop meningismus due to blood in the CSF. A non-contrast head CT will often demonstrate SAH within the first 24 hours of the bleed. Your patient's head CT at the outside hospital, by report, never showed SAH. Since the initial head CT was days after the trauma, and your patient was being treated for a presumed carotid dissection, you still consider the diagnosis. A lumbar puncture will be mandatory to rule out SAH in your patient. You will have the laboratory technician immediately centrifuge the CSF to evaluate for blood and xanthochromia.

Could orbital cellulitis explain these findings? Your patient was diagnosed with sinusitis on a head CT two weeks prior to presentation. Orbital cellulitis is one of the most common complications of sinusitis. Patients typically present with periorbital swelling and impaired extra-ocular movements, as seen in your patient. You remember reading a recent review article about ocular infections. It stressed that if bilateral periorbital edema develops, you should think bilateral cavernous sinus thrombosis. You are relieved that your patient's symptoms are unilateral since bilateral cavernous sinus thrombosis carries a very dismal prognosis.

Could meningitis explain these findings? Meningitis is a known intracranial complication of sinusitis. Your patient's symptoms and physical findings can all be seen in meningitis. Ptosis, CN VI palsy, and hypertension (with bradycardia) suggest increased intracranial pressure and impending brain herniation. Could your patient have other intracranial complications of sinusitis as well?

A picture is worth a thousand words. Before your patient has even returned from the radiology department, the radiology resident calls you with the results of the imaging. The explanation unfolds in front of you. It is all in his head! The study shows paranasal sinusitis, involving the left ethmoidal and sphenoid sinuses most notably. The left cavernous sinus and left supraorbital vein are thrombosed. There is severe stenosis of the left internal carotid artery. This stenosis is most prominent in the petrous and cavernous sinus portion

of the internal carotid artery. There is also a tentorial subdural empyema.

His lumbar puncture showed WBC: 845 with differential: P89%; L9%, M2% RBC: 275; glucose 23mg/dl; protein 196mg/dl. The CSF was negative for xanthochromia. Opening pressure was normal.

Indeed, your patient has several intracranial complications of sinusitis: cavernous sinus thrombosis, subdural empysema, and meningitis. With your answer in hand, you start the patient on IV vancomycin, cefotaxime, and metronidazole and admit him for treatment.

Figure 1

Figure 1: Magnetic resonance angiogram (on day of admission) demonstrating stenosis of the distal left internal carotid artery in the region of the cavernous sinus.

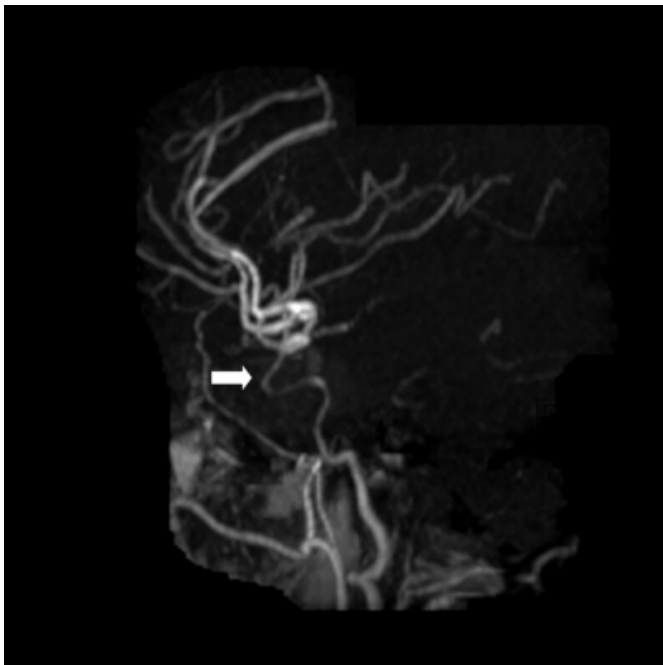


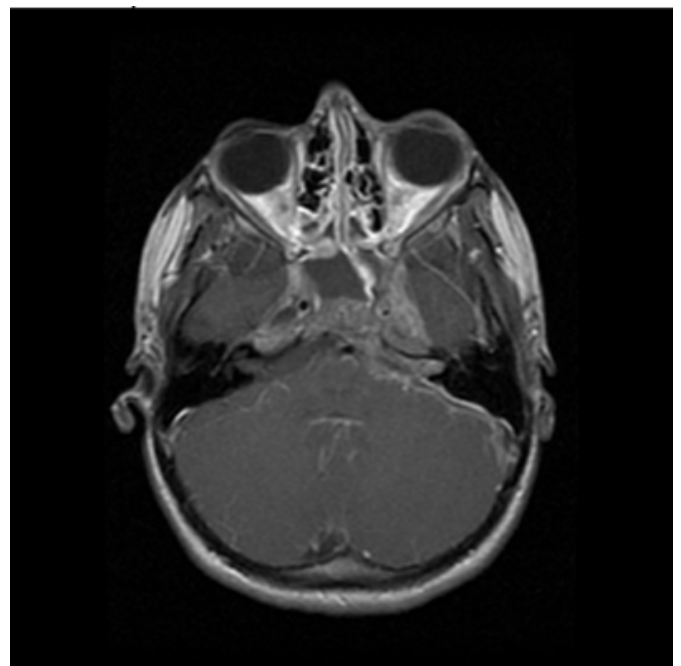
Figure 2

Figure 2: Coronal T1 MRI with contrast (on day of admission) showing left cavernous sinus enlargement and thickening.



Figure 3

Figure 3: Axial MRI on day of admission. The MRI demonstrates left cavernous sinus enlargement, decreased caliber of the left internal carotid artery, sphenoid and ethmoid sinusitis, posterior fossa empyema, and suppuration of the left internal auditory canal.



DISCUSSION

Septic cavernous sinus thrombosis (CST) is almost always caused by infections of the middle third of the face and paranasal sinuses. In the pre-antibiotic era, CST was almost universally fatal. The current estimate of mortality is approximately 30%, in part due to delayed diagnosis.¹ Patients who do survive often have residual cranial nerve palsies.

Sphenoid and ethmoid sinusitis are almost always implicated in sinus-related CST. Valveless diploic veins draining the sinuses communicate with the cavernous sinus, allowing spread of infection. The veins are most abundant in adolescents, increasing their risk of intracranial complications of sinusitis. For reasons unknown, males are more likely to develop CST.² Infection may also spread through defects in sinus walls. Our patient's imaging was negative for bony fractures.

The internal carotid artery and CN VI pass directly through the cavernous sinus. This, in retrospect, explains the internal carotid artery narrowing mistaken as carotid artery dissection and the inward deviation of his left eye. Thrombosis of the supraorbital vein, which drains directly into the cavernous sinus, caused the periorbital swelling. The third cranial nerve passes through the cavernous sinus on the lateral wall. As the CST progressed, CN III palsy developed, leading to pupil-sparing ptosis. Almost all patients with CST have pronounced fever¹, which was absent in our patient, but may have been masked by some of the medications being administered for pain control. Headache is very common, and may be unilateral or retro-orbital. Patients with subsequent bacterial meningitis or meningeal irritation will have nuchal rigidity.

While CST is a clinical diagnosis, radiologic imaging often helps confirm the diagnosis. MRI with MR venogram is preferred over CT in detecting CST.³ MRV will show absence of venous flow in the affected cavernous sinus.⁴ MRI may demonstrate compromise of the internal carotid artery in the region of the cavernous sinus. CT with contrast frequently demonstrates sinusitis, enlargement of the superior ophthalmic vein, and filling defects of the cavernous sinus.⁵ However, CT imaging may be negative in early CST.⁶

CST requires emergent treatment with IV antibiotics. Retrospective review of imaging at the outside hospital showed findings of CST one week prior to presentation at our facility. Prompt treatment may have prevented our

patient's subdural empyemas and meningitis. Surgical sinus drainage is recommended in the literature⁷, but patients have been known to survive without this procedure. Cultures typically yield streptococci or *Staphylococcus aureus*, but anaerobes or gram-negative rods may also be cultured. CSF cultures are frequently negative, probably due to the fastidious nature of the organisms involved. Because of this, this boy was treated with broad spectrum antibiotics to cover all potential organisms associated with sinusitis. The typical duration of treatment with IV antibiotics is four to six weeks, or two weeks past clinical resolution. There is no consensus as to whether anticoagulant therapy may improve clinical outcome, but may be considered in deteriorating patients. Maintaining hydration appears to be important.

Two days after admission, your patient develops acute hydrocephalus and subsequent respiratory arrest. He is taken emergently to the operating room by the neurosurgeons for evacuation of his subdural empyemas and for externalized ventricular shunt placement. A culture from the evacuated fluid grew *Streptococcus mitis*. He required re-evacuation two days later and placement of a permanent ventriculoperitoneal shunt. He was discharged home after completing six weeks of antibiotics. At six-month follow up, he was back to his healthy self, with no residual deficits.

CORRESPONDENCE TO

Deborah K. Sokol, PhD, MD Pediatric Neurology-RI 1757
Riley Hospital for Children Indiana University School of
Medicine 702 Barnhill Drive Indianapolis, IN 46202
317-278-2219 FAX: 317-274-3622 dksokol@iupui.edu

References

1. Southwick FS, Richardson Jr. EP, Swartz MN. Septic thrombosis of the dural venous sinuses. *Medicine*. 1986; 65: 82-106.
2. Clayman GL, Adams GL, Paugh DR, Koopmann Jr. CF. Intracranial complications of paranasal sinusitis: A combined institutional review. *Laryngoscope*. 1991; 101:234-239.
3. Igarashi H, Igarashi S, Fujio N, et al. Magnetic resonance imaging in the early diagnosis of cavernous sinus thrombosis. *Ophthalmologica*. 1995; 209: 292-296.
4. Saah D, Schwartz A. Diagnosis of cavernous sinus thrombosis by magnetic resonance imaging using flow parameters. *Ann Otol Rhinol Laryngol*. 1994; 103: 487-489.
5. Ahmadi J, Keane JR, Segall HD, et al. CT observations pertinent to septic cavernous sinus thrombosis. *American Journal of Neuroradiology*. 1985; 6: 755-758.
6. Hulcelle PJ, Dooks EC, Mathurin P, Conelis G. MRI assessment of unsuspected dural sinus thrombosis. *Neuroradiology*. 1989; 6: 217-2.
7. Lew D, Southwick FS, Montgomery WW, Weber AL, Baker AS. Sphenoid sinusitis: A review of 30 cases. *New England Journal of Medicine*. 1983; 309: 1149-1154.

Author Information

Jennifer Zimmer, M.D.

Department of Pediatrics, Riley Hospital for Children, Indiana University School of Medicine

Jay Bhatt, M.D.

Department of Neurology, Riley Hospital for Children, Indiana University School of Medicine

James H. Conway, M.D.

Infectious Disease, Department of Pediatrics, Riley Hospital for Children, Indiana University School of Medicine

Mary Edwards-Brown, M.D.

Neuroradiology, Department of Radiology, Riley Hospital for Children, Indiana University School of Medicine

Deborah K. Sokol, Ph.D., M.D.

Pediatric Neurology, Department of Neurology, Riley Hospital for Children, Indiana University School of Medicine