

Cervicomedullary Injury After Pneumococcal Meningitis With Brain Edema

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Objectives: To demonstrate a rare but potential mechanism of quadriplegia in a patient with fulminant pneumococcal meningitis complicated by severe intracranial hypertension.

Design: Case report.

Setting: Intensive care unit.

Patient: A 21-year-old man who presented with 3 days of headache, combativeness, and fever.

Intervention: Antibiotics and steroids were initiated after lumbar puncture yielded purulent cerebrospinal fluid and streptococcus pneumoniae.

Results: The patient's course was complicated by severe cerebral edema necessitating intracranial pres-

sure monitoring and intracranial pressure-targeted therapy. Within 5 days he developed quadriplegia and areflexia. Brain and cervical spine magnetic resonance imaging revealed patchy areas of T2 signal hyperintensity with associated gadolinium enhancement in the superior cervical spinal cord, cerebellar tonsils, and medulla.

Conclusions: Quadriplegia secondary to tonsillar herniation in fulminant meningitis is rare but should be considered in patients with acute quadriparesis after treatment of increased intracranial pressure. Magnetic resonance imaging signal changes and gadolinium enhancement may be demonstrated. Significant improvement of cord symptoms can be expected.

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REPORT OF A CASE

A 21-year old man was brought into the hospital with 3 days of headache followed by acute stupor, combativeness, and fever. A noncontrast computed tomographic scan showed mild generalized cerebral edema, and his white blood cell count was 22 000. He was intubated for airway protection and then started receiving sedation with propofol and fentanyl. His temperature was 39.4°C; blood pressure, 105/57; and heart rate, 123 beats per minute. Findings of neurologic examination were normal except for positive Brudzinski sign. Lumbar puncture yielded markedly purulent cerebrospinal fluid, a protein level of 0.773 g/dL (to convert to grams per liter, multiply by 10.0), and a glucose level of less than 20 mg/dL (to convert to millimoles per liter, multiply by 0.0555). A Gram stain showed gram-positive cocci that was later proved to be streptococcus pneumoniae. He started receiving 10 mg of ceftriaxone, vancomycin, and dexamethasone intravenously every 6 hours. He was overbreathing the

ventilator and was placed on spontaneous mode of ventilation overnight. The next morning, he did not awaken despite discontinued sedation and was found to be deeply comatose. Minimally reactive pupils without corneal, gag, or cough reflexes were observed. Oculocephalic reflexes were only present on one side. He had no motor response to pain and did not breathe during the set ventilatory rate. There were subtle rhythmic facial movements that were suspicious for seizure. A repeated computed tomographic scan showed global effacement of cisterns, marked increase in cerebral edema, low cortical density areas consistent with encephalitis, and early cerebellar tonsillar herniation (**Figure 1A**).

The patient started receiving high-dose dexamethasone (100 mg followed by 10 mg every 4 hours) and 20% mannitol (2 g/kg followed by 0.5 g/kg every 4 hours) in an effort to reduce cerebral edema. Levetiracetam was given after an electroencephalogram showed epileptogenic abnormalities with severe generalized slowing and little background variability. Severe intracranial hypertension was

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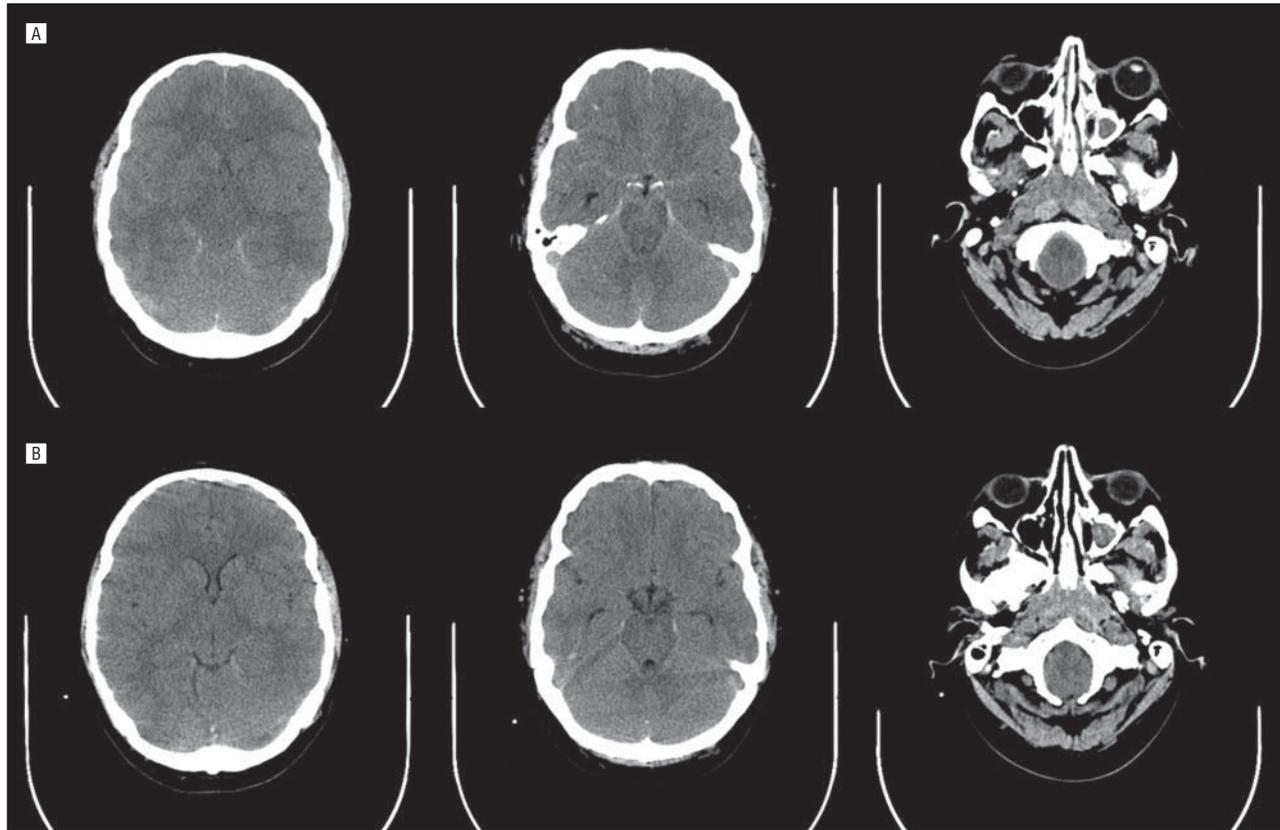


Figure 1. A, A computed tomographic scan of the brain without contrast before the start of intracranial pressure–targeted therapy shows severe generalized edema, effacement of cisterns within the brainstem, and tonsillar herniation; notice the pseudosubarachnoid hemorrhage appearance due to the severity of brain swelling and purulent cerebrospinal fluid. B, A repeated scan 5 days later shows dramatic improvement in cerebral edema, reappearance of subarachnoid spaces, and retraction of cerebellar tonsils.

detected after placement of ICP monitor (80 mm Hg on insertion, stabilizing between 50–58 mm Hg) around the time the patient was receiving the bolus doses of dexamethasone and mannitol. Systemic cooling to 33°C was instituted. Dopamine intravenous infusion was implemented to maintain cerebral perfusion pressures between 60 and 70 mm Hg. Within a few hours, his ICP decreased to 18 to 20 mm Hg and he recovered cough and right corneal reflexes. By the fifth hospital day, he had regained all brainstem reflexes but had generalized areflexia with no response to noxious stimulation of his extremities. Repeated computed tomography of the head showed marked improvement with reappearance of subarachnoid spaces and global reduction in cerebral edema (Figure 1B). Mannitol and intracranial pressure (ICP) monitoring were discontinued and he was extubated on day 10. He mouthed responses to questions with normal facial strength but had brisk reflexes in the lower extremities with clonus and extensor plantar response. Diminished touch, temperature, and pinprick were present on his cheeks, mandible, and forehead. His joint position was recognized by the patient only at the level of his left shoulder and knee and reduced diffusely on his right side. Vibration was reduced in all extremities, though in the left more than the right. Patchy pinprick loss was detected in both extremities to his mid–upper arm. There was no definitive sensory level. Lesions were localized to the corticospinal tracts, spinal tract of trigeminal nerve,

dorsal columns, and anterior columns. The “onion skin” pattern of facial sensory involvement was localized to spinal trigeminal tract located in the upper cervical cord and suggested pathology affecting the upper cervical cord and lower medulla. Magnetic resonance imaging of the brain with contrast showed scattered abnormal parenchymal signal and enhancement compatible with inflammatory changes of meningitis, without residual cerebral edema. Magnetic resonance imaging of the cervical spine showed abnormal patchy areas of T2 signal with associated enhancement in the superior cervical spinal cord, cerebellar tonsils, and medulla (Figure 2). There was no abnormal cord signal or enhancement below the level of C2. He was discharged to our inpatient rehabilitation service and, after 2 months, has regained most strength in his upper extremities and is walking with 2-person assistance.

COMMENT

This is the first reported case of quadriplegia due to tonsillar herniation after pneumococcal meningitis with successfully treated brain edema since Ropper and Kanis’ seminal case 10 years ago.¹ Our case is also remarkable because of neurologic improvement after immediate intervention with high-dose steroids, ICP monitoring with aggressive ICP management, and antibiotics.

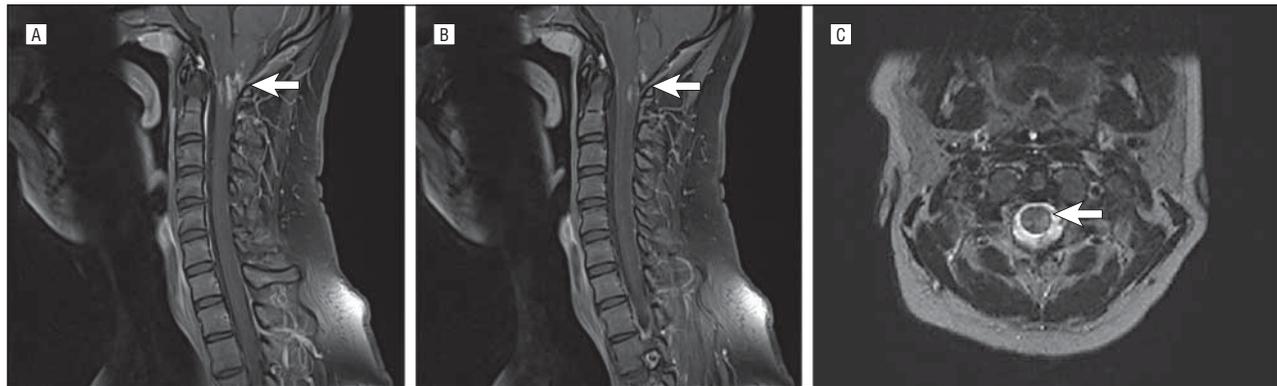


Figure 2. T1-weighted postgadolinium sagittal (A and B) and T2-weighted axial (C) images 1.5 weeks after the onset of illness show cerebellar tonsillar herniation below the foramen magnum with subsequent cervicomedullary compression. Patchy gadolinium enhancement and T2 signal changes are restricted to the cervicomedullary junction and cerebellar tonsils, as indicated by the arrows.

Pneumococcal meningitis is well known for its associated intracranial complications including cerebral edema reported in as many as 5.7% to 29% of patients with cerebrovascular complications.^{2,4} The benefit of steroid administration in pneumococcal meningitis is unproven⁵ but improved neurological outcome has been reported, presumably by mediating toxic inflammatory mediators released during bacterial lysis that can hasten cerebral edema and accelerate herniation.^{2,6-8} Edema tends to be an early feature of meningitis in adults, often presenting with stupor or coma on examination,^{2,8} and findings of CT may be normal.⁹⁻¹¹ It was noted in a study by Lindvall et al¹¹ that mean ICP was higher and cerebral perfusion pressure was lower in nonsurvivors of bacterial meningitis, with streptococcus pneumoniae being the most common pathogen identified. Results from The Dutch Cohort Study showed that death in younger patients (<60 years) was primarily secondary to neurological complications such as cerebral edema and herniation, while patients older than 60 years died of systemic complications.³

Treatment according to ICP-targeted therapy may help direct management in comatose or stuporous patients with meningitis, potentially leading to improved outcomes in this subset of patients, who already face higher mortality rates.^{2,8,10-13} Although there is no proof of benefit of ICP monitoring in meningitis, ICP monitor placement and supportive therapies geared at lowering ICP may be beneficial in younger adults with symptoms of intracranial hypertension. There are also scattered case reports that describe successful use of ICP monitoring, Transcranial Doppler, craniectomy, and medical therapy based on the “Lund concept,” but none have been validated.^{12,14-16}

Quadriplegia after meningitis has been reported to occur in 2% of patients with meningitis and can be associated with tonsillar herniation, myelitis, vasculitis, cord infarction, arachnoiditis, systemic hypotension, and epidural abscess.^{2,10,17,18} Cervicomedullary injury due to the unwanted consequence of tonsillar compression within the foramen magnum following severe brain edema with resultant elevated intracranial pressures is the most likely mechanism in our patient. We suspect the prevalence of this injury is rare because few comatose patients with early brainstem involvement recover after treatment of se-

vere brain edema. The high-dose steroids given to these patients might indicate a diagnosis of steroid myopathy. Spinal shock resulting in flaccid paraparesis immediately following tonsillar herniation may also be mistaken for a critical illness polyneuropathy. Myelitis associated with the primary infection has also been reported to account for 2.3% of quadriplegia in these patients.^{4,19} However, careful inspection of the magnetic resonance images of the spine in patients with myelitis reveals that the T2 signal hyperintensity in the spinal cord and leptomeningeal gadolinium enhancement are usually most pronounced in the gray matter and are longitudinally extensive, spanning from the cervical to thoracolumbar cord,⁴ whereas in patients with cervicomedullary injury secondary to tonsillar herniation, the intramedullary and meningeal changes are patchy and limited to the upper cervical cord and cerebellar tonsils. Though herniation may also lead to vascular compromise with resultant cord infarction,¹⁸ the pattern of T2 signal changes in our patient did not conform to a spinal arterial distribution.

In conclusion, quadriplegia secondary to cervicomedullary injury arising from tonsillar herniation due to severe intracranial hypertension is a rare complication of pneumococcal meningitis. Magnetic resonance imaging signal changes and gadolinium enhancement are restricted to the upper cervical cord and cerebellar tonsils. Prognosis is generally poor but favorable recovery with aggressive corticosteroid treatment and ICP-targeted therapy can be achieved, as demonstrated in our patient.

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