A 3-Month-Old Infant with Upper Extremity Weakness

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A 3-month-old, full-term, previously healthy girl was transferred to an outside pediatric intensive care unit (PICU) after being evaluated by her primary medical doctor for decreased upper extremity movement. She was born at 38 weeks’ gestation to a G2P2 mother, who received complete prenatal care. The mother had gestational diabetes, controlled by diet alone. She was group B streptococcus positive during her first pregnancy, was negative during this pregnancy, and did not receive any antibiotics before delivery. There were no postnatal complications, and she was able to go home after 2 days.

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Two weeks before admission, she developed upper respiratory infection symptoms with rhinorrhea and cough. She was seen by her primary medical doctor (PMD), who provided reassurance. These symptoms resolved without intervention or treatment. One week before admission, she had increased irritability, worse while in the upright position. She continued to have appropriate oral intake and urine output, without vomiting or diarrhea. Two days before admission, she developed a temperature of 100.2°F.

The following day, she had increased irritability that did not seem to improve with typical consoling maneuvers. She had decreased movement of her bilateral upper extremities; her PMD noted absent reflexes. Her lower extremities were not affected. She continued to lift her head while prone, but would no longer reach for objects or place her hands to midline. There was no associated rash, bruising, or continued fevers.

Given the unusual nature of the presentation, a skeletal survey was obtained to evaluate for nonaccidental trauma. No fractures were seen. Because of her PMD’s concern for infantile botulism, she was immediately transferred to the...
PICU. Her family and social history were both noncontributory.

While in the PICU, neurology evaluated her further. She was able to track and had grossly intact cranial nerves. She had moderately decreased tone in her bilateral upper extremities, with normal leg strength. They confirmed the absent upper extremity reflexes first reported by her PMD. There were brisk patellar and ankle reflexes, with no associated head lag, facial droop, or other neurological deficits.

Magnetic resonance imaging (MRI) of the cervical, lumbar, and thoracic spine revealed a retropharyngeal abscess, measuring 4 mm x 2 mm x 9 mm, with extension into the superior-posterior mediastinum (Figure 1). An epidural abscess extended from C2 to T2 with associated cord compression; the largest area of narrowing was at the level of C3. A horizontal-oriented linear area of T2 prolongation and enhancement was also seen extending through the superior aspect of the C3 vertebral body, communicating with the epidural space and likely the hypopharynx (Figure 2). An MRI of the brain was unremarkable.

She underwent an incision and drainage of the retropharyngeal abscess, with postop reports noting very minimal pus. Cultures were sent, which ultimately grew *Escherichia coli* and *Enterobacter cloacae*. Susceptibility patterns revealed no evidence of extended-spectrum beta-lactamase resistance.

She was empirically started on broad-spectrum antibiotics, as well as systemic corticosteroids. The epidural abscess was located anteriorly to the thecal sac; therefore, neurosurgical intervention was not recommended. With ongoing physical therapy and long-term antibiotic treatment, she began to regain movement in both arms. She passed a brainstem auditory evoked response (BAER) test, and had no evidence of immediate sequelae or complications.
Diagnosis:
Cervical Neurenteric Fistula and Epidural Abscess

DISCUSSION

Between 1928 and 2002, there were 25 reported cases of neurenteric cysts (fistulas) in children younger than 12 months of age. Seven of these cases occurred in the cervical spinal region, with only four between C1 and C4. None of these patients presented with an epidural abscess as a complication. Epidural abscesses are very rare in the pediatric population. Most occur in the mid-thoracic or lower lumbar region;2 and there are previous reports of cases in the presacral,3 lumbar, and intradural extramedullary thoracic areas.5 It appears that an epidural abscess complicating a congenital neurenteric fistula at the level of the cervical spinal cord has not been previously reported.

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The spinal cord begins to differentiate during the third week of gestation. Ectoderm overlying the notochord thickens and forms the neural plate. In a matter of days, the neural plate consists of two neural folds, which ultimately form the neural tube. This tube first occurs in the cervical region and fusion of the neural folds proceeds both cephalically and caudally.5 Abnormal adhesions between the endoderm and ectoderm during this phase of development lead to the formation of neurenteric cysts. They are rare congenital anomalies, located primarily in the lower cervical and upper thoracic regions associated with a ventral predominance.4

Incomplete separation of the notochord (ie, ectoderm) and primitive gut (ie, endoderm) leads to persistence of these cysts. Possible communications between the spinal cord and gastrointestinal tract can include a fibrous tract, fistula or, in extreme situations, an open cleft, which allows visceral herniation to the surface of the back. They are benign congenital tumors, representing 0.3% to 5% of all spinal tumors.5,6 They are most commonly seen in the first 6 months of life, with symptoms primarily resulting from mass effect on the spinal cord, brainstem or peripheral nerves. The review by Kadhim and colleagues7 reported an 8-week-old infant who presented with paraparesis; the cyst was located in the extra/intradural space from T6 to T9. Sarkar and colleagues8 reported three cases; one was in a 2-month-old child who presented with dyspnea and lower limb weakness, with the cyst located in the intradural space from T6 to T7.

Of all the pediatric cases reviewed by Santos De Oliveira and colleagues,9 only five presented with an infectious etiology. Most had symptoms related to cord compression, such as paraparesis, hemiparesis, or isolated muscle involvement.

Epidural abscesses are another rare phenomenon in the pediatric population. Most are related to previous lumbar punctures, epidural anesthesia, or recent spinal surgery.2 Spontaneous abscesses can also occur in patients with an underlying immunodeficiency or those receiving hemodialysis. In the absence of any spinal instrumentation, or direct communication with the spinal cord, spontaneous epidural abscesses are uncommon.

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A case report by Darwish and colleagues10 highlighted a newborn presenting with an epidural abscess with presacral neurenteric fistula. These abscesses are usually located in the midthoracic or lower lumbar spine because of the narrowing of the spinal cord at those levels. There is a rich venous plexus located within a large dorsal extradural space, with the primary blood supply originating from the inferior vena cava. This plexus is thought to be the primary route of infection, with Staphylococcus aureus accounting for approximately 80% of pediatric cases.2

There can be many associated anomalies in patients with neurenteric cysts or fistulas. They are a form of spina bifida occulta and consist of an intradural cyst lined by mucin-producing epithelium. It can include a mixture of gastrointestinal, pancreatic, or squamous epithelium.1

Anterior spina bifida with hemi-vertebrae or butterfly vertebrae is also common associations.4 If anterior spina bifida is the primary concern, possible connections between the spinal cord and some part of the alimentary tracts should be evaluated.10 It is, therefore, critically important to evaluate the entire spinal canal in patients with suspected or confirmed neurenteric cysts. Other reported associations include anal atresia, cardiac anomalies, tracheoesophageal fistulas, and meningomyelocles.4

SUMMARY

A detailed history should be obtained for all pediatric patients presenting with acute neurological changes. A thorough physical exam can help narrow the possible etiologies. Prompt imaging can lead to a more precise understanding of the level of neuronal involvement.
Neurenteric cysts and fistulas are extremely rare in the pediatric population, with most occurring in the lower cervical and upper thoracic region. Most patients will present with symptoms related to cord or brainstem compression; however, associated infections have been reported. Epidural abscesses are also rare, occurring in the mid-thoracic or lower lumbar region.

When there is no communication between the gastrointestinal tract and spinal canal, *S. aureus* accounts for most of these infections. When gram-negative pathogens, such as *E. coli* and *E. cloacae*, are isolated, it is imperative to evaluate for the presence of neurenteric fistulas. Because of numerous anomalies associated with neurenteric cysts, the entire spinal canal should be evaluated completely.

**REFERENCES**