Presacral Neuroenteric Fistula in a Newborn Presenting With an Epidural Abscess: Case Report and Review of the Literature

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ABSTRACT. We describe a newborn infant (<24 hours of age) who presented with mild swelling on the back and buttocks attributable to a neuroenteric fistula complicated by an epidural parasacral abscess infected with mixed coliforms. Epidural abscesses in infancy are extremely rare, and one has not been observed previously in the newborn period. The infant was surprisingly mildly affected. Prompt intervention led to an excellent outcome. Coliforms may colonize the infant gut in <24 hours, even in the developed world. Unexplained swellings on the backs of infants should lead to a search for underlying malformations and an early surgical review, which is best conducted with a combined pediatric surgical and neurosurgical approach. Pediatrics 2004;114:527–531. URL: www.pediatrics.org/cgi/doi/10.1542/peds.2003-0496-F

newborn infant, neuroenteric fistula, epidural abscess, coliform, meningitis.

ABBREVIATIONS. MRI, magnetic resonance imaging; SEA, spinal epidural abscess.

Neuroenteric cysts (fistulas) are rare, with <30 cases being described in the pediatric literature. Epidural abscesses presenting in infancy are even rarer, with the youngest patient reported being 20 days of age.1 An epidural abscess complicating a congenital neuroenteric cyst appears not to have been reported previously.

CASE REPORT

A 1-day-old boy was admitted to the neonatal unit at a peripheral hospital with irritability, lethargy, and poor feeding. He had been delivered at 42 weeks of gestation through normal vaginal delivery and weighed 3250 g. His mother had an older healthy child and had remained in good health during this pregnancy. She admitted to smoking cigarettes and occasionally drinking alcoholic drinks. Results of ultrasound scanning performed in the first trimester had been reported as normal and consistent with dates. The infant’s condition at birth was excellent, and he was allowed home, breast-feeding successfully, at 12 hours of age. In the examination, the infant was mildly jaundiced and lethargic. He had cool extremities, with a capillary return of 4 seconds, a heart rate of 150 beats per minute, a respiratory rate of 68 breaths per minute, and a skin temperature of 36.6°C. There was no sign of respiratory difficulty. The patient’s anterior fontanelle was not distended, and his posture was normal. His abdomen was soft, without organomegaly. Movements were normal in all limbs, with normal tone and reflexes. The patient’s anus was in a normal position, and he passed stools without difficulty. His back was normal, with no sign of sinuses or fistulas. Blood and urine cultures were obtained. Blood tests showed a C-reactive protein level of 25 µg/mL (normal: <10 µg/mL) and a white blood cell count of 10 300 cells per mm³ (neutrophils: 8000 cells per mm³). Results of a chest radiograph were normal. Lumbar punctures were performed as part of the septic screening and on 2 occasions revealed frank yellow-green pus, which showed large numbers of pus cells and mixed flora with Gram-staining and subsequently showed heavy growth of Escherichia coli and enterococci. A provisional diagnosis of mixed-organism meningitis was made, and the infant began broad-spectrum antibiotic treatment and was transferred to the neonatal intensive care unit of Wellington Hospital for additional investigations and treatment.

At the time of arrival, the infant’s posture was noted to be abnormal; he was lying somewhat hyperextended and was irritable during handling. There was an indistinct but definite swelling over the lower lumbar spine, which appeared tender (Fig 1). There was some erythema over the right posterior lower trunk and buttocks, extending into the inguinal region, with deviation of the gluteal cleft to the left; the anus was thought to be mildly displaced anteriorly. There was a tender, mildly fluctuant swelling (5 cm × 2 cm) over the superomedial buttock area on the right (Fig 2), which appeared to be separate from the swelling over the lumbar spine. The infant exhibited mild tenderness in the right renal angle with deep palpation. A provisional diagnosis of spinal epidural abscess (SEA) was made, but the gluteal swelling was unexplained. The infant underwent ultrasound scanning of the lumbar spine and urgent magnetic resonance imaging (MRI) of the pelvis. On ultrasound scans, the conus was at L1. The nerve roots of the cauda equina were bunched together, with reduced pulsation, and a paucity of surrounding cerebrospinal fluid was noted. On MRI scans, a presacral cystic mass measuring 5 cm × 2 cm was confirmed; this mass was pushing the rectum and bladder fora.
The mass extended through the greater sciatic notch on the right, into the right buttock region (Fig 3). Air was clearly visible within the mass. The lesion extended into the epidural space of the spinal canal, with air within the extradural space (Figs 5 and 6) reaching the upper cervical spine. Gadolinium enhancement of the tissue in the epidural space was noted, in keeping with local inflammatory changes. The bony elements of the sacrum were intact. The findings strongly suggested fistulous communication between the rectum, the presacral cyst, and the epidural space. Results of MRI scans of the brain were normal.

The infant underwent surgery with a combined pediatric surgical and neurosurgical team. The presacral collection was drained surgically through a posterior sagittal approach. Meconium-stained fluid was drained, and the rectum was defunctioned with a divided sigmoid colostomy. An L5-S2 laminectomy was performed. The epidural space contained purulent material, with striking greenish discoloration of the epidural fat (Fig 7). There was a separate collection in the right parasacral region, with a possible sinus tract extending anteriorly. There was no cerebrospinal fluid leak. Specimens for histologic examinations were taken from the epidural fat and from the parasacral cavity and its anterior extension.

The histologic examination of the right parasacral tissue showed large-bowel wall composed of mucosa, submucosa, and muscularis propria. The epidural material showed adipose tissue containing pigment-laden macrophages and material containing deeply pigmented hair shafts. No epithelial tissue was identified. The purulent material from the epidural space grew *E col i* and enterococcal species. The epidural tissue showed inflammation and the presence of meconium.

A gastrograffin enema on the 10th postoperative day showed a small fistulous tract from the upper rectum, above the levator ani muscle, extending posteriorly into the presacral space (Fig 8). The...
presacral space was not injected with contrast material; therefore, no communication with the spinal canal could be demonstrated.

The postoperative course was unremarkable. The infant continued to receive intravenous antibiotic therapy and was discharged from the hospital 3 weeks after surgery, to continue antibiotic treatment for an additional 3 weeks.

At 6 weeks, the presacral space was explored through combined abdominoperineal incisions. Inflammatory tissue that was firmly attached to the posterior wall of the rectum, 4 cm from the anus, was removed. This tissue was confirmed, in the subsequent histologic evaluation, to be the obliterated fistula and residual cavity. The postoperative course was unremarkable. At 3 months, results of a contrast examination of the rectum were reported as normal and the divided colostomy was closed.

DISCUSSION

Neuroenteric fistulas are rare congenital malformations thought to result from persistent communication between the embryonic neural tube and gut tissues.\(^2\) Isolated neuroenteric fistulas, without involvement of bone or soft tissues, are even rarer.\(^3\) By definition, a fistula is a communication between 2 epithelial surfaces. It may not involve the bone but it should therefore go through the soft tissues to reach the epidural fat. The biopsy of the right parasacral mass showed epithelial tissue and bowel muscle reaching the epidural space.

Embryologically, the spinal cord is formed from 2 parts. The segment from the medulla to the midthoracic region is formed by primary neurulation. The more distal segment is formed by a process called “canalization and retrogressive differentiation.”\(^4\) The notochord arises from the notochord process on day 15 of embryonic life. It causes the overlying ectoderm to differentiate into the neural plate, the lateral parts of which thicken to form the neural folds, which eventually close to form the neural tube (neurulation) on day 17. When neurulation is complete, the neural tube separates from the overlying ectoderm. The posterior neuropore (caudal end)
closes at ~27 days. Canalization is the process by which the neural tube elongates caudal to the posterior neuropore. During this process, the notochord and neural epithelium fuse, forming a caudal cell mass. Canalization of this cell mass and cellular necrosis of parts of it (retrogressive differentiation) lead eventually to formation of the conus medullaris and filum terminale. At ~25 days, the notochord separates from the gut and the neural tube and, with the surrounding mesenchymal cells, forms the somites. Each somite has a medial sclerotome, which eventually forms the vertebral bodies, and a lateral myotome, which forms the paraspinal muscles. Notochordal remnants between the vertebrae become the nucleus pulposus of the intervertebral disk.

A persistent connection between the endoderm and ectoderm is thought to result in splitting of the notochord. In the most severe form of split-notochord syndrome, communication exists from the intestinal cavity through the spinal canal and into the skin of the back. Any part of that fistula may become obliterated, leaving less complete fistulas and cysts. A persistent distal end of this fistula forms an enteric sinus, a persistent middle portion forms a neuroenteric cyst, and a persistent proximal end may result in a neuroenteric fistula (Fig 9). Feller and Sternberg described the split-notochord syndrome, which is characterized by the triad of vertebral anomalies, a posterior mediastinal mass, and intestinal duplication. The majority of neuroenteric fistulas occur in the thoracic region and involve the foregut or the midgut. Rarely, they can arise in the lumbosacral region and involve the hindgut.

In our case, the presacral cyst clearly communicated with the rectum; that communication was demonstrated with the gastrograffin enema. The failure to demonstrate communication between the presacral cyst and the epidural space could have been the result of insufficient pressure within the presacral cyst, because the contrast material was injected into the rectum and not into the cyst. The striking meconium staining of the epidural fat, the mixed intestinal flora within the epidural space, the pigmented macrophages, and the demonstration of colonic mucosa and submucosa in the tissue taken from the right parasacral region are strong indicators of the existence of such fistulous communication. The hair that was demonstrated in the epidural tissue was most likely swallowed with amniotic fluid during embryonic life and reached the epidural space through the neuroenteric communication. However, teratomas within fistulas have also been described. The air in the epidural space could have been intestinal gas entering through the fistulous communication, although gas produced through bacterial metabolism cannot be excluded as an alternative cause.

We were surprised at the early colonization of this breastfed infant, with extensive deep infection with both enterococci and \textit{E coli} by 20 hours of life. Although this is unusual for Western countries, it would be a common observation in rural communities such as those in Pakistan.

SEAs are uncommon among children and, to our knowledge, a SEA has not been observed previously in a newborn infant. Nejat et al\textsuperscript{1} described a thoracic epidural abscess in a 20-day-old infant. In the series reported by Auletta and John,\textsuperscript{13} the youngest patient was 5 weeks of age and the average age was 8 years. None of their 8 cases or the other 26 cases they extracted from the literature had a recognized associated neuroenteric cyst, although the 5-week-old infant was described as having the Curran triad (anorectal malformation, sacral bony anomaly, and presacral mass). Many of those cases were reported before the advent of MRI, which has allowed better recognition of associated anomalies.

The predominant organism in SEAs in most series is \textit{Staphylococcus aureus}, consistent with a different route of infection. The diagnosis of SEA should trig-
ger a search for the underlying cause, and the presence of coliforms should suggest the possibility of a connection to the colon.

Early diagnosis and aggressive treatment of this condition are essential, because delay can lead to catastrophic results, including sepsis, spinal deformity, permanent neurologic deficits, and death. A diverting colostomy is an important part of the treatment of children with neuroenteric fistulas. This prevents contamination of neural tissues and enables the physician to control the infection while awaiting definitive surgical treatment of the fistula.

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