Large intramedullary abscess of the spinal cord associated with an epidermoid cyst without dermal sinus

Case report

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An intramedullary abscess of the spinal cord (IASC) represents a rare disease associated with a potentially devastating outcome. Few cases involving children suffering from an IASC have been reported in the neurosurgical literature. In the majority of the reported pediatric cases there were either congenital abnormalities, such as a dermal sinus, or signs of local infections leading to a secondary hemopoietic spread. The authors report the case of an 18-month-old girl with an extensive IASC associated with an epidermoid cyst extending from T-11 to S-2 without evidence of a dermal sinus or history of clinically apparent systemic infection. To their knowledge, this is the first case report of an IASC without a condition facilitating either direct contamination via a dermal sinus or hemopoietic spread from an infectious focus outside the central nervous system. Signs and symptoms, the clinical course, and imaging features are discussed and the relevant literature is reviewed. (DOI: 10.3171/SPI-07/09/357)

Key Words • abscess • epidermoid cyst • magnetic resonance imaging • pediatric neurosurgery • spinal cord

An IASC is a rare disease and has a poor prognosis unless diagnosed and treated promptly. In 1830 Hart reported the first case, which involved an adult patient, and since that time fewer than 100 cases have been described in the literature (for review see the article by Simon and colleagues17). Largely these reports were focused on adults; fewer than 40 reports of IASC involved children. The pathogenesis of the disease in adults is hemopoietic spread of a systemic infection, mainly from a cardiopulmonary source. In children the majority of cases of IASC are associated with spinal cord developmental abnormalities, namely a dermal sinus, or previous surgical repair of a spina bifida aperta. Thus, an intramedullary abscess is predominantly due to direct contiguous spread of the infection from the skin. Similar to IASC in adults, the hemopoietic spreading of systemic infections can also occur. We report the case of an 18-month-old girl harboring an IASC without evidence of a dermal sinus tract, history of surgery, or clinical signs of a previous or concurrent infection.

Case Report

Examination. This 18-month-old girl was admitted to the children’s hospital with a recent history of walking difficulties, as well as a fever of 38˚C that began the evening before admission. On admission she was unable to walk or stand because of pain in her hips. Laboratory investigations on admission revealed an increased erythrocyte sedimentation rate (32/64) and leukocytosis (12,500/μl), whereas the C-reactive protein was negative. Initial urine examination had shown a high number of leukocytes. On physical examination she was in no apparent distress, afebrile, and interactive. Initially a coxitis fugax was thought to be the cause of her hip pain, but the patient’s symptoms rapidly progressed to paraplegia 5 days after admission to the hospital. Emergency spinal MR imaging demonstrated no lower-extremity muscle atrophy. We did observe gluteal muscle atrophy and a diminished sphincter tone causing urinary and fecal incontinence. The Babinski sign was negative. The patient’s neurological status was otherwise normal. Tonsillitis, sinusitis, and urogenital tract infection were excluded by both physical examination and laboratory investigations. Inspection and palpation of the spine showed a well-demarcated 25-mm-diameter nevus in the lower lumbar region (Fig. 1); aimple or a tuft of hair—

Abbreviations used in this paper: IASC = intramedullary abscess of the spinal cord; MR = magnetic resonance.
suggestive of a dermal sinus or other abnormalities associated with spinal dysraphism—was absent. Magnetic resonance imaging revealed an intramedullary lesion spanning the spinal cord from C-6 to the conus medullaris (Fig. 2a–e). Injection of intravenous contrast demonstrated an extramedullary part of the lesion in the lumbar canal involving the cauda equina and extending to S2–3 (Fig. 2c and f). The lower part of the lesion appeared polycystic, with bright contrast enhancement on the border and almost no solid portion.

Operation. The patient was immediately taken to the operating room following the MR imaging. A T11–L4 laminotomy was performed. After incision of the dura mater, we observed a lesion resembling a tumor in the region of the conus medullaris, displacing the nerve roots bilaterally. There was no clear cleavage plane with respect to the conus, but the nerve roots could be dissected free with the aid of a microscope. The capsule of the tumor was opened to obtain a tissue sample for a frozen section. The tumor tissue was partially solid but we also noted a yellow fluid that was purulent in nature. A median myelotomy was performed to allow dissection of the tumor in the conus region, and after we opened a cystic cavity the tumor could be dissected from the cord. At this stage of the operation a complete resection was considered to be possible, and the surgical field was enlarged by performing an L-5 laminectomy and by opening the sacral spinal canal. After meticulous dissection, the tumor was completely separated from the cauda equina on the left side, whereas some minor tissue remnants had to be left on the right side due to incorporation of the nerve roots into the tumor capsule. During resection, the lesionlike material resembling an epidermoid was removed. Following dural closure the osseous defect was reconstructed by fixation of the lamina, and the wound was closed in a standard fashion.

Pathological Examination. Several specimens for microbiological investigation were collected intraoperatively, and examination revealed Proteus mirabilis and Escherichia coli. Antibiotic treatment with clindamycin and cefotaxime was initiated. Because an allergic exanthema occurred, treatment was switched to meropenem. Histopathological investigation excluded the diagnosis of a malignant tumor but showed squamous epithelium with keratin. Furthermore, Gram staining revealed Gram-positive cocci, and therefore the material was classified as an epidermoid with partial intramedullary growth was the primary pathological entity. The epidermoid might have developed in utero by inclusion of ectoderm during closure of the neural tube. One of the two detected bacteria was E.

Discussion

Intramedullary abscess of the spinal cord in children is a rare pathological finding; only about 40 cases have been described in the literature. The pathogenesis of IASC includes hemopoietic, contagious, and idiopathic spread. Direct contagious infection can originate from the mediastinum, peritoneum, and retroperitoneal space. Alternatively, IASC is disproportionately found in children with congenital abnormalities. A dermal sinus is the leading cause of IASC in children. Simon et al. recently reported the case of a 13-month-old boy with an IASC due to a dermal sinus tract and presented an analysis of 38 pediatric cases. In this review 53% of the children either had prior anatomical defects including dermal sinus (17 patients), isolated myelomeningocele (1 patient), an unspecified lumbar defect (1 patient), or a sacral decubitus ulcer (1 patient).

Fourteen patients had previously undergone surgical correction of the anomaly. A preexisting anatomical defect was significantly more common in children younger than 5 years of age than in children older than 5 years. The risk posed by a congenital dermal sinus, which can potentially transmit any superficial infection into the intrathecal space, has been stressed by many authors. Furthermore, the inner end of a dermal sinus extending into the intradural space may form a dermoid or epidermoid tumor, as reported by Morandi et al., in whose review congenital dermal sinus accounted for approximately 20% of IASCs and an epidermoid was encountered in four cases.

The present case differs from all other previous cases in that the patient did not have any of the previously described congenital abnormalities. The only dermal abnormality was the cutaneous nevus in the lower lumbar region (Fig. 1). No dermal sinus or any other anomaly was found during clinical, MR imaging, or intraoperative examination during abscess drainage and removal of the lesion. The presence of a very small fistulous connection as a cause of transmitted infection, however, cannot be definitely excluded. Based on the histological features, an intraspinal epidermoid with partial intramedullary growth was the primary pathological entity. The epidermoid might have developed in utero by inclusion of ectoderm during closure of the neural tube. One of the two detected bacteria was E.

Fig. 1. Photograph showing a small cutaneous nevus but no signs of a dermal sinus or other abnormality. This photograph was obtained after surgery. No dermal sinus was seen prior to or during surgery.
coli, which commonly causes urinary tract infections. Thus, the epidermoid might have been infected via a hemopoietic spread from a clinically asymptomatic urinary tract infection. This was suspected because urine testing revealed abundant leukocytes. A fistulous connection between the urogenital tract and the intraspinal space was excluded. Common hemopoietic sources for IASC include urogenital infection, infective endocarditis, and infective pulmonary diseases, but this route is more common in adults than in children. In our patient no clinical signs of systemic infection were present; in particular, there was no evidence of meningitis, which was atypical given the findings of Morandi et al., in whose study most of the patients presented with signs of meningitis or systemic infections. Very few investigators have described a chronic intramedullary abscess, which may mimic an intramedullary tumor, as we assumed in our patient. Guzel et al. reported a case of a 14-month-old girl with thoracolumbar IASC. In this case no tumor was proven and there was no evidence of any organism on the Gram or Ziehl–Neelson staining. In some previous studies, organisms such as Staphylococcus, Streptococcus pneumoniae, Haemophilus, Proteus, Listeria, and Actinomyces were isolated from IASC specimens, with Staphylococcus and Streptococcus being the most frequently isolated organisms. However, delayed cultures of IASC are sterile in microbiological examination in 28% to 39% of cases. Most of the children (55%) in one study were infected with a single organism, whereas 17% of the patients had mixed bacterial flora. In the present case, Proteus mirabilis was detected in addition to E. coli, and therefore a mixed bacterial flora was present. Proteus mirabilis was most frequently found to be associated with IASC and dermal sinus in previous reports.

The most commonly involved region in pediatric patients is the thoracolumbar spine. This is in agreement with our patient, but MR imaging showed signal changes up to the cervicothoracic spine, resembling a holocord astrocytic tumor. In a few previously reported cases of IASC the authors observed that once the infection developed in the spinal cord, it tended to spread longitudinally along the fibers of the cord itself. The MR imaging findings in our patient are similar to the reported MR imaging features of myelitis due to bacterial meningitis and, as such, this entity should have been included in the differential diagnosis. In a report by Kastenbauer et al., which included three adults, the authors described hyperintensities on T2-weighted images...
that predominantly involved the gray matter and extended from the cervical to the lumbar cord. Leptomeningeal and discrete nodular intramedullary enhancement on T1-weighted images were detected in one patient.

Once an IASC is suspected, the mainstays of treatment involve immediate surgical decompression, myelotomy, and abscess drainage followed by intravenous antibiotic therapy. For surgical treatment complete excision is not necessary because the lesion is sometimes poorly delineated and extends throughout the spinal cord. Clinical improvement is usually seen, as was the case in our patient.4

Conclusions

To our knowledge, this is the first report of an intraspinal epidermoid and an IASC without congenital abnormalities, previous surgery, or clinically apparent systemic infection. Although the MR imaging features may resemble those of a tumorlike lesion, which may not be considered for resection, an IASC should be considered and immediate surgery is recommended to establish a histological diagnosis and microbiological results. Early surgical intervention and antibiotic treatment are the prerequisites for neurological improvement.

Acknowledgment

We thank Dr. Charles L. Rosen, M.D., Ph.D. (Department of Neurosurgery, West Virginia University), for his editorial help in writing this manuscript.

References

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Accepted May 23, 2007.

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