SECTION EDITOR: ABRAHAM BERGMAN, MD

Bilateral Upper Limb Weakness and Stridor

Raja Ram, MBBS; Anne Reaveley, MBBS; Jon Dorling, MBChB; Julian Eason, MHSc

Clinical Presentation: A 4-week-old girl had difficulty breathing, reduced feeding, and upper limb weakness. She was born at 39 weeks' gestation by forceps after a failed ventouse. At birth, minimal resuscitation was needed, with suction and facial oxygen. Results of subsequent examinations after birth were normal. At 2 weeks of age, she developed a dry cough, which gradually worsened, and on the day of admission, her breathing became increasingly noisy and she developed apneic episodes. There had been no exposure to cigarette smoke or pets.

Dr Eason: The history of cough developing in the preceding 2 weeks and subsequent lower respiratory tract signs culminating in apnea is highly suggestive of respiratory syncytial virus bronchiolitis. Apnea is a common clinical finding in young infants with bronchiolitis. Noisy breathing could, however, be confined to the upper respiratory tract, and the change in breathing pattern suggests laryngomalacia. Laryngomalacia occurs when the larynx is soft and flexible; it is present from birth and exacerbated by upper respiratory tract infection. While it is a possibility in this case, the lack of stridor in the first few weeks of life makes it unlikely.

Other causes of stridor include an enlarging subglottic hemangioma or a vascular ring; however, these would not give rise to neurologic signs. Bacterial infection by *Haemophilus influenzae* type B needs to be considered, but epiglottitis is a rare event now since immunization and uncommon at this age. Bacterial supraglottitis and tracheitis should also be considered.

Physical Examination: Vital signs were all normal, with a temperature of 36.9°C, oxygen saturations of 98% in air by pulse oximetry, and heart rate and blood pressure within normal limits. An examination of the respiratory system revealed inspiratory and expiratory stridor, with tachypnea (respiratory rate, 62/min) and respiratory distress. Neurologic examination showed flaccidity of the upper limbs when both arms were held in a "waiter's tip" position, indicative of an Erb palsy. There was normal grip and finger power. The lower limbs were normal except for a small paronychia on her right fifth toe.

Dr Eason: The examination results suggest an upper airway obstruction despite some lower respiratory signs. The presence of bilateral shoulder muscle weakness (Erb palsy) is both confusing and of concern and will need further investigation. It implies the possibility of severe birth trauma, with damage of the phrenic nerve (cervical spinal nerve roots C3, C4, and C5) as well as the upper brachial plexus roots (cervical spinal nerve roots C5, C6), causing bilateral Erb palsy and vocal cord paralysis. In the absence of a history of shoulder dystocia, this seems very unlikely. Photographs taken in the first few days of life also rule out this diagnosis by showing the patient's ability to lift her arms.

Other possibilities include a type 2 Chiari malformation with downward displacement of the cerebellar vermis and the medulla to cause foramen magnum impaction syndrome. This can lead to abnormal brainstem and nerve root function. It is rare except as a complication of spina bifida and hydrocephalus. Neither a spinal lesion nor a large head circumference is present on examination. Bulbar palsies, due to abnormal function of lower motor neurons (eg, spinal muscular atrophy) may display similar signs if the vocal cords are involved. On examination, one would expect to see a lax open mouth with jaw muscle atonia, weakness, and wasting, particularly of the tongue, which would also show fasciculation. Infection with diphtheria or Epstein-Barr virus should be detectable on examination of the throat by the appearance of an adherent membrane or inflamed tonsils covered in exudate.

Although there is diagnostic uncertainty, the priority is to secure the airway and commence artificial ventilation. The presence of a pediatric anesthesiologist is advisable, as intubation could be difficult. Once the infant is stabilized, a postintubation chest radiograph, complete blood cell count with differential and platelet counts, blood levels of electrolytes, blood urea nitrogen, and creatinine, blood culture, and a throat swab should be obtained. She will need to be transferred and to receive ongoing care in an intensive care unit.

Laboratory Data: A complete blood cell count detected an elevated white blood cell count of $30.6 \times 10^3/\mu L$, with a marked neutrophilia of $24\,200 \times 10^6/\mu L$, and an elevated level of C-reactive protein (33 mg/L). Although a blood culture was negative for organisms, both a nasopharyngeal aspirate and a throat swab culture indicated methicillin-resistant *Staphylococcus aureus*. A swab from the paronychia grew a *S aureus* that was fully sensitive to flucloxacillin. Cranial ultrasonography results were normal, and a barium swallow showed no evidence of a vascular ring. A lateral soft tissue x-ray film and a magnetic resonance image (MRI) of the cervical spine were obtained (**Figure 1** and **Figure 2**).

Ultrasonography of the neck confirmed the presence of a well-defined retropharyngeal collection. This was not amenable to percutaneous drainage due to the

From the Jenny Lind Children's Department, Norfolk and Norwich University Hospital Trust, Norwich, England.

©2002 American Medical Association. All rights reserved.
Downloaded From: http://archpedi.jamanetwork.com/ on 05/03/2015

Figure 1. Magnetic resonance image of the cervical spine. A large retropharyngeal abscess can be seen at the level of the C3-C4 vertebrae. There is extension posteriorly into the epidural space and the spinal cord is moderately displaced. Partial destruction of the body of C3 is shown anteriorly.

overlying blood vessels, so a transpharyngeal biopsy and aspiration were performed. Histologic examination results confirmed inflammatory cells and gram-positive cocci, but the culture was negative.

Dr Eason: It is likely that the paronychia on her right fifth toe led to hematogenous spread of bacteria. The bacteria seeded into the rich capillary bed of the cervical discs, causing staphylococcal discitis. Local spread into the cervical vertebra then occurred, leading to osteomyelitis, with subsequent extension into the retropharynx. Her neck was stabilized in a soft collar and she was treated with intravenous antibiotics for more than 12 weeks. It is good

Figure 2. Lateral radiograph of the cervical spine. Large retropharyngeal mass with loss of the fourth cervical vertebral body demonstrated.

practice to use broad-spectrum cover, such as cefotaxime sodium and flucloxacillin until culture results are available. In this case, methicillin-resistant *S aureus* was cultured, so the treatment was changed to intravenous vancomycin hydrochloride and cefotaxime.

Hospital Course: The patient was ventilated for 48 hours, and once she was stabilized and receiving antibiotic treatment, she was allowed to go home in between doses. A follow-up MRI scan showed gradual healing of the infected area. After 12 weeks of antibiotic therapy, our patient showed a good recovery and was able to lift both arms up and above her shoulders, with no muscle weakness on examination.

COMMENT

Cervical osteomyelitis in newborns is very rare. Only 5 cases have been reported.[1-5] Clinical presentation with paresis and paralysis occurs in less than 1% of cases.[6] We have found only 2 patients with cervical osteomyelitis and bilateral upper-limb paralysis reported in the literature.[4,5] Both newborns had definite weakness of the arms around 3 weeks of age. Neither of them was reported to have had stridor.

Martijn et al[1] described a 3-week-old girl with cervical osteomyelitis, quadriplegia, and paronychia of both hands. Our patient had a paronychia of her right toe, from which *S aureus* was isolated. *Staphylococcus aureus* is the most frequent organism responsible for pyogenic vertebral osteomyelitis.[1-3,5] The culture of the retropharyngeal biopsy specimen was negative but antibiotics had been given prior to the biopsy. Sharma et al[3] obtained a positive culture from a trocar biopsy specimen of the verte-

©2002 American Medical Association. All rights reserved.
Downloaded From: http://archpedi.jamanetwork.com/ on 05/03/2015

bral body or the involved soft tissue in only 27% of patients who were already taking antibiotics .

Stridor or airway obstruction is commonly seen in children younger than 7 years with a retropharyngeal abscess.[7] Retropharyngeal abscesses rarely occur in newborn infants, with only 13 cases having been reported since 1966.[8] One series of neonatal cases diagnosed as having retropharyngeal abscess described 3 infants who had stridor on initial examination. In addition, they all had external swelling on the neck, which was absent in our patient.[9]

Lateral radiograph of the cervical spine and MRI are used to confirm this rare infection.[1-5,9] Magnetic resonance imaging is helpful in detecting spinal cord involvement and extradural abscess, and may allow earlier detection of osteomyelitis than the plain films alone.[3] Ultrasonography of the neck[8] is also suggested for diagnosing and assessing a suspected retropharyngeal abscess.

The mainstay of treatment is antibiotics.[1] Depending on the degree, extent, and site of spinal instability, patients may also need spinal immobilization for several weeks.[3] For our patient, a soft cervical collar was used to stabilize the neck. Rarely, a patient may need surgical decompression and stabilization. Recovery from any neurologic deficit depends on the degree and duration of spinal cord compression.[3]

Accepted for publication April 10, 2002.

Drs Ram and Reaveley wrote this article in conjunction with Drs Dorling and Eason.

Corresponding author and reprints: Jon Dorling, MBChB, Jenny Lind Children's Department, Norfolk and Norwich University Hospital Trust, Colney Lane, Norwich, England NR4 7UY (e-mail: jondorling@supanet.com).

REFERENCES

1. Martijn A, van der Vliet AM, van Waarde WM, et al. Gadolinium-DTPA enhanced MRI in neonatal osteomyelitis of the cervical spine. *Br J Radiol.* 1992;65:720-722.
2. Ein SH, Shandling B, Humphreys R, et al. Osteomyelitis of the cervical spine presenting as a neurenteric cyst. *J Pediatr Surg.* 1988;23:779-781.
3. Sharma RR, Sethu AU, Mahapatra AK, et al. Neonatal cervical osteomyelitis with paraspinal abscess and Erb's palsy: a case report and brief review of the literature. *Pediatr Neurosurg.* 2000;32:230-233.
4. Ammari LK, Offit PA, Campbell AB. Unusual presentation of group B *Streptococcus osteomyelitis. Pediatr Infect Dis J.* 1992;11:1066-1067.
5. Colville J, Jeffries I. Bilateral acquired neonatal Erb's palsy. *Ir Med J.* 1975;68: 399-401.
6. Resnick D, Niwayama J. Osteomyelitis, septic arthritis and soft tissue infection: the axial skeleton. In: *Diagnosis of Bone and Joint Disorders.* 2nd ed. Philadelphia, Pa: WB Saunders; 1988:2619-2646.
7. Morrison JE Jr, Pashley NR. Retropharyngeal abscesses in children: a 10-year review. *Pediatr Emerg Care.* 1988;4:9-11.
8. Gaglani MJ, Moise AA, Demmler GJ. Neonatal radiology casebook: retropharyngeal abscess in the neonate. *J Perinatol.* 1996;16:231-233.
9. Coulthard M, Isaacs D. Neonatal retropharyngeal abscess. *Pediatr Infect Dis J.* 1991;10:547-549.

Submissions

The aim of "Clinical Problem Solving" is to demonstrate how master clinicians approach complex problems. Contributions for the section are encouraged; informal inquiries via e-mail to the editorial office are advised prior to preparing and submitting a manuscript.

©2002 American Medical Association. All rights reserved.
Downloaded From: http://archpedi.jamanetwork.com/ on 05/03/2015

Inadequate Recognition of Education Resources Required for High-Risk Students With Sickle Cell Disease

Students with chronic illness, such as sickle cell disease (SCD), diabetes mellitus, cancer, or asthma are at risk for poor academic attainment. Students aged 5 through 19 years with SCD have an additional burden, as this disease is associated with multiple painful episodes resulting in hospitalizations and strokes.[1]

In 1997, we reviewed the status of 17-, 18-, and 19-year-olds with SCD followed up by the Hematology/Oncology Center at St Louis Children's Hospital (St Louis, Mo). Only 4 (15%) of 26 patients were on target to completing high school. Given the poor education attainment, we expected that most of these students would have received educational support and/or had Individual Education Plans (IEP)[2] to which they were entitled through the Individuals with Disabilities Education Act.[3] The poor high school graduation rate for these young adults prompted us to consider the adequacy of educational support for students with SCD who are considered to be at increased risk for poor academic attainment, defined as those with strokes or multiple painful episodes requiring hospitalization in a 12-month period. As a result of the initial survey, we systematically assessed the academic performance, school attendance, and availability of IEPs for students with SCD believed to be at high risk for academic difficulties.

Thirty-nine high-risk students—24 with strokes and 15 with 3 or more hospitalizations for pain in 1 year—were identified in 1999. Twenty-eight percent (stroke, n=8; pain, n=3) of the students had been retained at least 1 grade before the 1999-2000 academic year. Students missed an average of 15.5 and 38.4 school days from the stroke and pain group, respectively. Seventeen (70%) of 24 of the stroke group and 2 (13%) of 15 of the pain group had been evaluated for or had an IEP. All students in both groups would have been expected to have received an assessment for an IEP based on either documented cognitive impairments or high absentee rates. Fifteen students were between the ages of 14 and 18 years and in high school. Only 5 (33%) of 15 were scheduled to graduate or had graduated from high school within 4 years. Two students with strokes received certificates of completion indicating that they had attended high school, but had not met the minimum academic requirements for graduation. No student was scheduled to complete a vocational training program.

Students with SCD and strokes or multiple admissions for pain are particularly vulnerable for poor academic achievement. Inadequate educational resource allocation for this group and the necessary steps to optimize academic potential has not been previously recognized. We believe that a multidisciplinary approach that includes partnerships with students, parents, medical staff, and educators is essential for the early identification and prompt educational assistance for students with SCD at risk for academic failure.

Sonya Herron, MSSW, LCSW
Stephen J. Bacak, MPH
Allison King, MD
Saint Louis, Mo
Michael R. DeBaun, MD, MPH
Washington University School of Medicine
660 S Euclid Ave, CB 8208
St Louis, MO 63110
(e-mail: debaun_m@kids.wustl.edu)

This work was supported by the Episcopal Foundation of St Louis, and the Doris Duke Clinical Scholars Foundation, New York, NY.

1. Miller ST, Macklin EA, Pegelow CH, et al. Silent infarction as a risk factor for overt stroke in children with sickle cell anemia: a report from the Cooperative Study of Sickle Cell Disease. *J Pediatr*. 2001;139:385-390.
2. Office of Special Education and Rehabilitation Services US Department of Education. A guide to the individualized education program, 2000. Available at: http://www.ed.gov/offices/OSERS/OSEP/Products/IEP_Guide. Accessed August 1, 2001.
3. ASPIIRE and ILIAD Partnerships. Law and regulations: IDEA '97 Law and Regs, 1998. Available at: http://www.ideapractices.org/index.htm. Accessed August 1, 2001.

Correction

Error in Measurement. In the article titled "Bilateral Upper Limb Weakness and Stridor," published in the September issue of the ARCHIVES (2002;156:941-943), on page 941, second column, under "Laboratory Data," neutrophilia should have been given as 24 200/μL.

©2003 American Medical Association. All rights reserved.
Downloaded From: http://archpedi.jamanetwork.com/ on 05/03/2015