Generalised hypotonia following erosive vertebral osteomyelitis in an infant

Neonatal cervical osteomyelitis is an extremely rare condition. It is a potentially dangerous condition with high fatality and disastrous consequences. Hence, it is important to recognise it early to prevent long-term morbidity and sensory-motor disabilities. It can have an indolent course conspicuous with the absence of pyrexia. We report a four-week-old infant with erosive osteomyelitis involving C1 and C2 and retropharyngeal abscess resulting in neurological abnormalities.

Jayanta Banerjee
MB BS, DCH, MRCPCH, Paediatric Specialist Trainee Year 5
docjayanta@hotmail.com

Sheetal Bhojani
MB BS, DCH, MRCPCH, Paediatric Specialist Trainee Year 5

Veerasingam Nerminathan
MB BS, MRCP, FRCPCH, Consultant Paediatrician

Paediatrics Department, Southend University Hospital NHS Foundation Trust, Essex

Keywords

hypotonia; osteomyelitis; neonate; cervical spine

Key points

Banerjee J., Bhojani S., Nerminathan V. Generalised hypotonia following erosive vertebral osteomyelitis in an infant. *Infant* 2011; 7(1): 16-19.

1. Neonatal cervical osteomyelitis is a rare condition, which can have high morbidity and mortality.
2. Osteomyelitis should be considered in any infant presenting with neurological signs, even in the absence of pyrexia.
3. Early clinical suspicion, prompt radiological investigations and broad-spectrum antibiotic cover can prevent disastrous consequences, while delayed diagnosis can lead to permanent paralysis and death.

Osteomyelitis, a potentially serious infection, is very rare in neonates[1,2]. It can follow an open bone injury, but more commonly occurs as a result of haematogenous spread of micro-organisms[3-5], and mostly involves the long bones of the upper or lower limbs[4]. In neonates, vertebral osteomyelitis accounts for only 1-2% of osteomyelitis[2].

To our knowledge, only seven cases of cervical spine osteomyelitis in a neonate or early infant have been reported in the literature[1,6-10]. None of these had involvement of the first cervical vertebra (C1). We report a four-week-old infant with erosive osteomyelitis involving C1 and C2 and retropharyngeal abscess resulting in neurological abnormalities.

Case report

A four-week-old male infant with no significant antenatal or peri-natal problems presented with 'floppiness' and difficulty in feeding. He was born by normal delivery with Apgar scores of 9 at 1 minute and 10 at 5 minutes. Pregnancy was uneventful and there were no risk factors or clinical features of maternal or neonatal sepsis.

According to parents he had been well since birth, establishing feeds and had normal movement of all four limbs until two to three weeks of age. In the following week, he was noted to feed poorly and there were reduced spontaneous movements in the right arm, prompting parents to seek medical attention. The paediatrician who saw the infant at that stage diagnosed it to be birth-related right-sided Erb's palsy. When reviewed a week later, he was unable to suck or swallow feeds from a bottle, prompting admission to the paediatric ward.

On admission, he was afebrile and pale. There was a small soft lump (1.5 cm x 1.5 cm) localised over the right side of his rib cage (7th-8th rib). Neurological examination revealed reduced tone in all four limbs with normal truncal tone. Bilaterally, his arms were held in an internally rotated position with flexion of the wrists and his lower limbs were abducted in a frog-like position. There was an absence of spontaneous movements and reflexes in all four limbs.

Initial blood tests showed: white cell count 33 x 10^9/L (neutrophils 26 x 10^9/L and lymphocytes 4 x 10^9/L), platelets 737 x 10^9/L and C reactive protein of 301 mg/L.

Urgent cranial and spinal MRI scans revealed a large retropharyngeal multi-cystic lesion encasing the anterior arch of the C1, consistent with multi-loculated abscess with phlegmon (**FIGURE 1**). C1 was subluxated with marked narrowing at the cranio-vertebral junction and effacement of the CSF showing evidence of cord compression. In addition there was a high intensity linear structure in the vicinity of the straight sinus suggesting straight sinus thrombosis.

Subsequently, the infant was transferred to the tertiary neurosurgical unit for further management. The retropharyngeal abscess was incised and drained via pharyngeal route by the ENT and spinal surgeons.

Blood cultures showed no growth after five days. Pus culture grew *Staphylococcus*

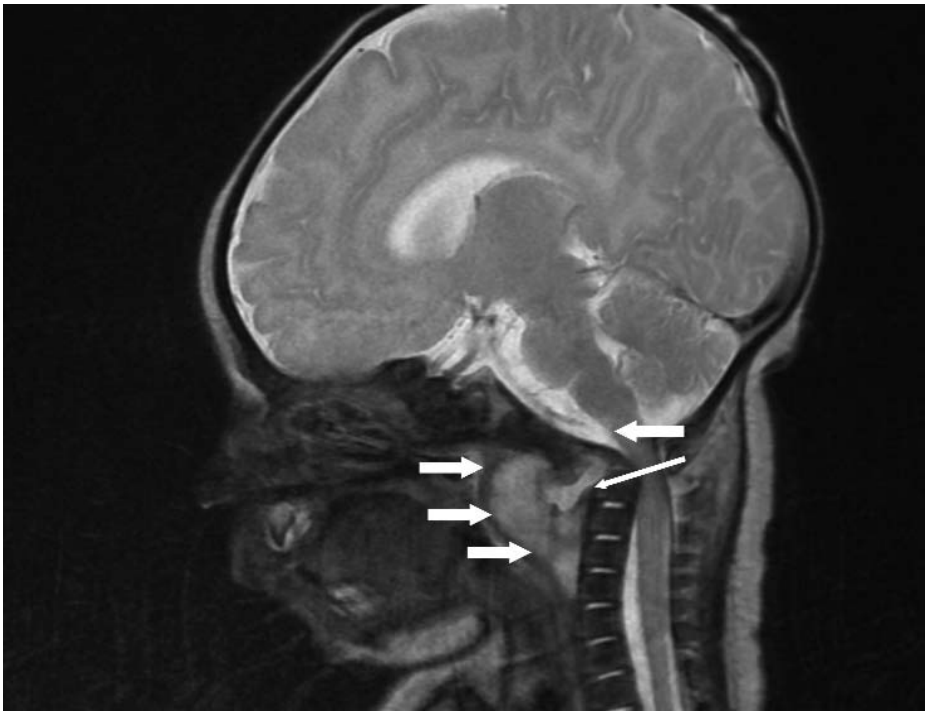

FIGURE 1 MRI scan showing subluxation of the C1 over C2 (thin arrow) with disintegration of the odontoid process and retropharyngeal abscess (three broad arrows) pressing over the oropharynx. There is effacement of CSF noted above the foramen magnum (single broad arrow).

aureus sensitive to amikacin, flucloxacillin, gentamicin, erythromycin and vancomycin. He was already commenced on rifampicin and clindamycin following consultation with the infectious disease team according to the local guideline for osteomyelitis. He showed significant improvement with this treatment.

Since the location of osteomyelitis indicated a haematogenous route of infection (in the absence of evidence of trauma), several investigations were carried out including an echocardiogram, an abdominal ultrasound scan, a bone scan and scan of the lump over the right side of his chest. Bone scan showed increased uptake in the region of the right anterior 7th/8th rib, proximal left humerus and distal left femur correlating with potential further sites of infection. The swelling over his 7th/8th rib subsequently disappeared following an eight week course of antibiotics. His echocardiogram did not reveal any signs of endocarditis and ultrasound scan of the abdomen did not reveal any signs of abscess or free fluid.

Subsequently, he had repeat brain and cervical spine MRI scans with contrast, MRA neck and venogram CT; all showing a reduction in the size of the retropharyngeal abscess. The summary of the findings of the various investigations indicated the pre and paravertebral phlegmon was still present with fluid tracking between the anterior arch of C1 and C2 with no thecal compression. The lateral masses of the C1 and C2 were slightly demineralised, worse on the left side. The CT venogram revealed reduction of the thrombus in the straight sinus compared to the previous MRI scan.

Since his MRI findings suggested potential instability of the cervical spine, a hard cervical brace was fitted. Regular physiotherapy was started and splints were fitted to his wrists, to counteract the developing hypertonia. He had further investigations to rule out underlying immune deficiencies which were all negative. He continues to have regular follow ups with physiotherapy, paediatric and neurosurgical teams. He received low molecular weight heparin for a total of six months for his sinus thrombosis and over this period continued to have improve-ment of tone and power in all four limbs.

Discussion

Historical perspective

According to Wilensky (1927) the term osteomyelitis was coined by Nelaton in 1844[4]. In 1921, Hobo postulated a hypothesis for the specific localisation of the bacteria in the metaphysis of the long bones. In 1959, Wiley and Trueta demonstrated that there were two potential routes for the organisms to reach the spine via haematogenous route: the nutrient arteries and the paravertebral venous system[4,5]. In 1979, Ogden provided definite histological support for an initial metaphyseal focus in human neonatal osteomyelitis[4].

Pathophysiology and microbiology

Vertebral plexus of the veins and the inter-metaphyseal communicating artery has an important role in bacterial colonisation and subsequent vertebral osteomyelitis[3-5]. Osteomyelitis of the cervical spine can lead to neurological deficits due to collapse, fracture dislocation or subluxation of the vertebrae and development of extradural abscess involving the spinal cord or nerve root[2-4].

Staphylococcus aureus is the most commonly isolated organism of osteomyelitis[1,2,8]. Another organism commonly implicated in neonatal osteomyelitis is *Group B beta-haemolytic Streptococcus*[6,9]. Before the introduction of antibiotics, acute haematogenous osteomyelitis had a mortality of approximately 20%, while long-term morbidity occurred in 50% of the cases[2,4].

Vertebral osteomyelitis can be the result of spread from the adjoining structures such as the pharynx, or may be due to haematogenous spread which can result from a variety of infections including arthritis, osteomyelitis of long bones, sinusitis, endocarditis, respiratory tract infections and skin infections, eg paronychia. However, in up to 70% of the cases no primary infective focus is identified[2,4,5,11]. The main sites of vertebral osteomyelitis are the lower thoracic and lumbar spine[3,4]. Cervical spine involvement is rare, more so in the neonatal age group, where only seven cases have been reported in the literature (**TABLE 1**).

Clinical presentation

Infants can present with a localised swelling, retropharyngeal abscess or cellulitis[3,12-15]. The presenting complaint can include respiratory distress, stridor, irritability and persistent crying[1,6,12]. There have been four cases of cervical osteo-myelitis in neonates reported in the literature who presented with unilateral/bilateral upper limb paralysis or Erb's palsy around three weeks of age[1,2,9,10]. The clinical course can be completely indolent as in our case where there was no fever; faltering

Author	Age at presentation	Organism isolated and probable primary site of infection	Presenting complaints and MRI scan findings	Intervention	Outcome
Ram et al[1] 2002	4 weeks	*Methicillin resistant Staph aureus*, Paronychia of right fifth toe	Stridor and bilateral upper limb weakness. MRI – retropharyngeal abscess C3-C4 region, extending posteriorly into epidural space and spinal cord moderately displaced, partial destruction of the body of C3	Neck stabilisation, 48 hours of ventilation, 12 weeks antibiotics with vancomycin and cefotaxime	Follow up MRI – gradual healing of infected area; after 12 weeks of antibiotics, patient was able to lift both arms above shoulders
Sharma et al[2] 2000	6 weeks	*Staph aureus,* No obvious infective focus	Weakness of left shoulder and elbow, 5 x 5 cm swelling in posterior triangle of the neck. CT showed destruction of C6, mild compression of thecal sac at C6 level	11 weeks of antibiotics, incision and drainage of abscess	6 months follow-up – full neurological recovery and new bone formation at the site of vertebral destruction on CT scan
Barton et al[6] 1996	1 month	*Group B Streptococcus,* Primary focus unknown	Feeding difficulty, posterior thoracic mass. MRI showed a 4 x 4 cm left paraspinal mass at T8-9, eroded vertebral bodies with wedge compression	Broad spectrum antibiotics, surgical debridement at 3 months of age	One year later the child remained on external body brace with good lower extremity motion and learning to stand
Ein et al[7] 1988	3 weeks	*Staph aureus,* Septic arthritis	Cyanotic spell requiring ventilation. CT scan showed a mediastinal mass extending from C7 to the carina, possible neuroenteric cyst	Antibiotics, incision and drainage of abscess	Walking since 2 years of age
Martijn et al[8] 1992	3 weeks	*Staph aureus,* Paronychia of both hands	1 week history of decreased movement of arms and legs, absent reflexes. MRI showed involvement of C2-4, prevertebral mass compressing and displacing the spinal cord posteriorly	Intravenous antibiotics	Recovery of neurological symptoms and MRI showing decreasing abscess size and residual changes in the body of C2-4
Ammari et al[9] 1992	3 weeks	*Group B Streptococcus* Probable maternal spread	Floppiness at birth progressing to reduced movement of upper extremities. MRI – mass at C5-6 with anterior collapse, kyphosis C6	Antibiotics for 28 days, surgical debridement	Discharged at 7 weeks of age with normal Moro reflex, upper limb power 4/5 bilaterally, improved grasp
Colville et al[10] 1975	3 weeks	*Staph aureus* Osteomyelitis of left femur	Bilateral shoulder and upper limb paralysis, swelling of left lower thigh and femur. X-ray – widening of the paravertebral space with destruction of C5	Antibiotics, immobilisation of the cervical spine and lower limbs, blood transfusion	Severe sepsis, bacterial endocarditis, cardiac failure leading to death

TABLE 1 Published cases of vertebral osteomyelitis in the neonatal age group (includes cases published in English only).

growth, poor feeding and neurological signs were the presenting features, which pointed towards a diagnosis of infection of the spine.

Radiology

Plain lateral radiographs of the cervical spine can be a useful initial investigation, which may reveal vertebral destruction, collapse of vertebrae, loss of bony trabeculation or a paraspinal soft tissue mass[2,8,11]. CT scans are more sensitive than plain X-rays and can offer useful additional information regarding the extent of the infective lesion. Gadolinium-enhanced MRI scans are particularly useful for detecting spinal cord involvement and extradural abscesses[11]. Radionuclide bone scans are sensitive and have been reported to detect up to 90% of pyogenic vertebral osteomyelitis. However, initial bone scans can be unremarkable as illustrated by the case described by Martijn et al[8].

Management

The mainstay of management of cervical spine osteomyelitis is broad spectrum antibiotics. The majority of cases in the literature have preferred a 10-12 week course which we feel could be decided individually in discussion with the microbiologist.

Surgical intervention in vertebral osteomyelitis is warranted in:

- wide-spread osseous involvement
- clinical sepsis which does not respond to antibiotics
- progressive neurological deficit due to spinal cord compression,[2] as seen in our patient.

One crucial aspect in the management of cervical osteomyelitis is the stabilisation of the spine, which can be achieved by a brace. This is followed by gentle mobilisation with external bracing using halo braces, moulded spinal jackets or spinal intervention[2]. Physiotherapist and occupational therapist should be involved to aid with mobilisation.

The long-term neurological prognosis depends on the degree and duration of spinal cord compression and hence remains largely unpredictable.

Conclusion

In any infant presenting with neurological signs *viz* unilateral or bilateral upper limb paralysis, stridor, irritability and difficulty in feeding, cervical spine lesions including osteomyelitis should be considered, even in the absence of pyrexia. Early clinical suspicion, prompt radiological investigations and broad-spectrum antibiotic cover can prevent disastrous consequences while delayed diagnosis can lead to permanent paralysis and death.

Acknowledgement: The authors would like to thank Dr Marc Tebruegge, Honorary Clinical Research Fellow, Infectious Diseases Unit, The Royal Children's Hospital Melbourne and Department of Paediatrics, The University of Melbourne Postgraduate Scholar, Murdoch Children's Research Institute Fellow, for his help and encouragement while writing this paper.

References

1. **Ram R., Reaveley A., Dorling J., Eason J.** Bilateral upper limb weakness and stridor. *Arch Pediatr Adolesc Med* 2002;156:941-43.
2. **Sharma R.R., Sethu A.U., Mahapatra A.K. et al.** Neonatal cervical osteomyelitis with paraspinal abscess and Erb's palsy: A case report and brief review of literature. *Pediatr Neurosurg* 2000;32: 230-33.
3. **Resnick D., Niyawama 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;2619-46.
4. **Nade S.** Acute haematogenous osteomyelitis in infancy and childhood. *J Bone Joint Surg Br* 1983;65:109-19.
5. **Wiley A.M., Trueta J.** The vascular anatomy of the spine and its relationship to pyogenic vertebral osteomyelitis *J Bone Joint Surg Br* 1959;41-B: 796-809.
6. **Barton L., Villar R., Rice S.** Neonatal group B streptococcal vertebral osteomyelitis. *Pediatrics* 1996;98:459-61.
7. **Ein S.H., Shandling B., Humphreys R., Krajbich I.** Osteomyelitis of the cervical spine presenting as a neurenteric cyst. *J Pediatr Surg* 1998;23:779-81.
8. **Martijn A., van der Vliet A.M., van Waarde W.M., van Aalderen W.M.C.** Gadolinium-DTPA enhanced MRI in neonatal osteomyelitis of the cervical spine. *Br J Radiol* 1992;65:720-22.
9. **Ammari L.K., Offit P.A., Campbell A.B.** Unusual presentation of group B Streptococcus osteomyelitis. *Pediatr Infect Dis J* 1992;11:1066-67.
10. **Colville J., Jeffries I.** Bilateral acquired neonatal Erb's palsy. *Ir Med J* 1975;68:399-401.
11. **Modic M.T., Feiglin D.H., Piraino D.W. et al.** Vertebral osteomyelitis: assessment using MR. *Radiology* 1985;157:157-66.
12. **Abdullah V., Ng S.K., Chow S.N. et al.** A case of neonatal stridor. *Arch Dis Child Fetal Neonatal Ed* 2002;87:F224-F25.
13. **Bourgeois F.T., Shannon M.W.** Retropharyngeal cellulitis in a 5-week-old infant. *Pediatrics* 2002;109:E51.
14. **Coulthard M., Isaacs D.** Neonatal retropharyngeal abscess. *Pediatr Infect Dis J* 1991;10:547-49.
15. **Morrison J.E. Jr, Pashley N.R.** Retropharyngeal abscesses in children: a 10 year review. *Pediatr Emerg Care* 1988;4:9-11.