Lesson of the Week

Congenital dermal sinus presenting as anaerobic meningitis

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Congenital dermal sinus is a dysraphic condition resulting from incomplete separation of cutaneous structures and neuroectoderm in early gestation. Detailed accounts of its embryology, presentation, and treatment have been given elsewhere.^{1,2} The clinical importance of the lesion lies in its associated neurological complications, which may present at any time after birth, though usually in early childhood. We report a case of cervical dermal sinus resulting in anaerobic meningitis.

Case report

A previously well 5 year old white girl first presented to a district general hospital in December 1985 with a short history of cough and general malaise. She improved after a course of antibiotics and was discharged. Four weeks later she was readmitted with persisting malaise, intermittent fever, vomiting, and occasional vacant spells. Some improvement occurred over 10 days without specific treatment and she was discharged without a definitive diagnosis. By March 1986 she was reasonably well but underweight. In May she was readmitted with a three day history of cough and fever and was complaining of headache. She had become withdrawn and was at times confused. Examination disclosed neck stiffness but no focal neurological deficit. An initial diagnosis of upper respiratory tract infection with encephalitis was made. Computed tomography, however, showed a communicating hydrocephalus, so the child was referred for neurosurgical assessment.

On arrival in our unit she was drowsy and irritable with pronounced neck stiffness and a temperature of 38°C. There was no focal neurological deficit. A dermal sinus was noted in the cervical region (figure) and plain radiographs showed an underlying congenital fusion of the spinous processes of the second and third cervical vertebrae. The child's parents had been aware of the sinus for some time and had expressed from it sebaceous material. On seeking medical advice, however, they had been reassured.

Emergency treatment consisted of a ventricular tap, releasing purulent cerebrospinal fluid under high pressure. Microscopy disclosed Gram positive cocci subsequently shown to be *Peptococcus magnus*, sensitive to penicillin, chloramphenicol, and metronidazole. *Corynebacterium xerosis* was also cultured, sensitive to penicillin and chloramphenicol. The initial antibiotic regimen of penicillin and sulphadimidine was modified to penicillin and metronidazole.

Two days after admission she had an exploration of the sinus, which was found to extend through the dura and communicate with an intrathecal epidermoid. Both tract and tumour were excised. *Clostridium welchii* was

The spine and occipital region of neonates should be examined thoroughly and if a dermal sinus is detected the child should be referred for neurosurgical treatment before disastrous consequences ensue; the same applies if a sinus is discovered in an older child

cultured from the epidermoid. Postoperatively the child remained very ill and drowsy. A left sided weakness developed and she could no longer localise auditory and visual stimuli. An episode of apnoea signalled an acute rise in intracranial pressure necessitating ventricular drainage. Repeated blockage led to replacement of the drain on five occasions. Two courses of intraventricular vancomycin, supplementary to the intravenous antibiotics, were required for episodes of ventriculitis. Eventually the cerebrospinal fluid became sterile, permitting the insertion of an internal shunt. Ventriculography showed that the foramina of Monro were partially obstructed, so both lateral ventricles were drained into the peritoneal cavity.

Appearance of lesion on neck (arrowed). (Macular capillary naevus is also evident above sinus. This is a common finding on back of neck and not specifically associated with congenital anomalies of neuraxis.)

The child remained unresponsive for several weeks but with eyes open and crying on occasions. Gradually she became more alert and then able to respond to simple questions. Thereafter her language and motor functions continued to improve and eventually she was able to return home. By that stage her language had become appropriate for her age. A left hemiparesis was still evident but she could walk with a rolator. She could feed, wash, and dress herself with a little help. Her hearing was good but vision was difficult to assess, though she was attentive with apparently full visual fields.

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Discussion

The true incidence of congenital dermal sinus is unknown, as many remain asymptomatic and undetected. The lesion consists of an epithelial lined tract which may extend to any depth from the skin, but it frequently communicates with the epidural or intradural compartments. The cutaneous opening may be remote from the level of termination of the sinus. An epidermoid or dermoid tumour may be present anywhere along its length. Neurological damage may result from compression of the cord or cauda equina if a mass is present, or from infection if the fistula forms a portal of entry for organisms. A wide variety of bacteria may gain entry, including anaerobic species. A mixed flora may be present. Meningitis, myelitis, or epidural abscess may result. Sometimes both neural compression and infection occur in the same patient. The clinical presentation may therefore be acute, subacute, or chronic, depending on the underlying pathological mechanisms. In our patient the presentation of several months of non-specific illness was probably a manifestation of low grade (partially treated) meningitis, eventually developing into a fulminating anaerobic infection with hydrocephalus.

Neurocutaneous fistulas may be present at any level of the neuraxis, usually in the midline. The commonest sites are the lumbosacral and suboccipital regions, presumably as a consequence of the delayed closure of the neural canal at these levels.³ They are less often seen in the cervical region, in common with spinal dysraphism in general.⁴ In a recent report of a single case only six other cases could be added from published work.⁵ The age range at presentation in the seven cases was from 10 months to 35 years. Three patients presented with progressive motor difficulty in all limbs, one with an intramedullary abscess, and one with paralysis and recurrent meningitis. Two were neurologically normal.

The sacrococcygeal dimple is similar in appearance to the dermal sinus but occurs at the caudal end of the spine. This common anomaly, present in about 3% of live births, arises as a result of failure of separation of the ligamentum caudale from the skin. It is usually distinguishable from a dermal sinus by virtue of its site over the coccyx and its caudal direction. If there is any doubt about its nature, however, exploration should be carried out.

The threat posed by the potential infective sequelae of a

congenital dermal sinus was emphasised as early as 1934.⁶ Nevertheless, patients still die as a result of this complication. Treatment of infection therefore needs to be prompt and aggressive and to include early surgical exploration of the sinus. In any child presenting with meningitis it is important to look for a sinus as an underlying cause, particularly when an unusual organism is isolated or in cases of recurrent meningitis.

Clearly, however, the ideal is to detect a sinus before the onset of complications. Inspection of the spine forms part of the routine examination of a neonate but the lesion may remain undetected because the external opening may be very small, requiring magnification to be seen, or it may be hidden by hair. Indeed, the neonatal notes of our patient disclosed that the spine had been inspected but no abnormality had been detected. Doctors conducting such examinations should look specifically for these lesions; more obvious deformities will have been noted by the midwife. Unfortunately, as with our patient, even if the lesion is noted the potential danger may not be appreciated by a medical attendant.

We present this case to draw attention to the potential hazard that this congenital abnormality presents to a patient. It is not possible to determine clinically whether a sinus reaches as far as the dura and the absence of bony abnormalities in a radiograph does not rule out such communication. Should a sinus be noted in the neonate or older child the family doctor or paediatrician should refer the patient for surgical exploration before complications occur.

References

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(Accepted 29 April 1987)

MATERIA NON MEDICA

Tie me kangaroo down

In England until 1877 if you were unfortunate enough to be convicted of any one of a large number of crimes you might well have been sentenced to transportation. If you survived incarceration in the notorious hulks, and then the long voyage in appalling conditions to Australia, you could have landed up in the penal settlement of Port Arthur in Tasmania.

Nowadays it is a very picturesque site, and the buildings—unfortunately damaged by bush fires at the beginning of this century—bear tribute to the fact that there must have been gifted architects and craftsmen among the transported convicts. The interdenominational church, the four storey penitentiary, and the 100 bed hospital are good illustrations of their architectural and building skills. In the penitentiary are preserved some of the menus available to the convicts, and it seems that always provided you were able to work you could expect three substantial meals a day; and in this respect you were probably better off than some of the free settlers trying to scratch a living from the land.

Geographically Port Arthur was ideal from the point of view of ensuring that prisoners did not escape, for it is a large peninsula joined to the main island by a narrow strip of land known as Eaglehawk Neck. Across this narrow isthmus fierce dogs were tethered on chains with little space between them, and in addition soldiers and constables patrolled the area, and the "Pommie" convicts were led to believe that the surrounding seas were teeming with sharks. As a result escapes were very rare. One ingenious prisoner managed to obtain some kangaroo skins in which he dressed himself and at dusk one evening started hopping through the surf out of range of the guard dogs. He had nearly reached the safety of the main part of the island

when he heard one of the armed guards shout to another, "Look at that old Jack [kangaroo]. Why not have a pot at him?" To the great surprise of the guards the kangaroo suddenly shouted, "Don't shoot—it's me."—ROBERT HOUSE, retired general practitioner, Tewkesbury.

What is the latest first aid advice on administering fluids to an injured person?

There is no change in the orthodox first aid advice not to give fluids by mouth to a person who has sustained an injury that may be serious and when an anaesthetic may be administered. Although there has been a new appreciation of the place of oral fluids in counteracting dehydration in cholera, the position is quite different after major trauma. This is because trauma, especially trauma severe enough to cause considerable loss of blood, causes massive stimulation of the sympathetic outflow to the abdomen. This greatly reduces the motility of the gut so that fluid reaching the stomach is likely to remain there. Secondly, it causes constriction of both the arterioles and the venules of the gut as a result of which absorption is reduced virtually to zero. If a patient needs fluid before reaching hospital it should be administered intravenously. The other possible first aid measure not requiring the skill of erecting an intravenous infusion is the application of shock pants.—WILLIAM RUTHERFORD, consultant in accident and emergency, Belfast.