Recurrent Meningoencephalitis and Ascending Myelitis Caused by Dermal Sinus Tract of Extraordinary Length

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Summary. The case of an infant with a dermal sinus tract, recurrent meningitis, ascending myelitis and a fatal outcome is described. The extraordinary extent of the cord affection with abscesses up to the level of the lower brainstem, and the presence of squamous epithelium within them, is difficult to explain on the basis of current concepts of the formation of dysraphic dermal sinus tracts. Morphogenetic, clinical and radiological aspects are discussed.

Key words: Ascending myelitis – Meningoencephalitis – Dermal sinus tract

Bacterial meningocerebrophalomyelitis is a major medical problem in infancy and childhood. Proper recognition is essential since any delay in the diagnosis, or inadequate treatment, may result in serious and even fatal consequences. The incidence of postmeningitic hydrocephalus requiring shunt procedures is still considerable. Reports from European countries give figures of up to 20% [8, 9, 13]. A recent report from Cape Town [6] suggests a higher incidence in less developed countries, viz. 30%–40%. The above figures do not include the unrecognized cases with hydrocephalus of a lesser degree which have spontaneously reached a state of balance. Damage to the cortex through the cerebritic process as well as through infarction caused by vasculitis [2, 5, 15] produces irreversible mental and neurological defects in 8%–30%. The spinal cord is also invariably involved, more or less, becoming covered by mats of densely packed mono- and polymenuclear cells, in the same way as the brain [2]. Root sleeves may become infiltrated and obliterated. The myelitis seldom dominates the clinical picture, however, and intramedullary abscess formation is exceptional. Gram-negative bacteria, such as E. coli, invade the CSF space either by way of a septicemia or they use direct dermal-subarachnoid communication. This latter path consists of a developmentally preformed canal, a pilonidal or dermal sinus tract. The present report concerns an instance of fatal outcome in which the terminal course was dominated by an ascending myelitis.

Case Report

A 2-month-old male infant had recurrent fever after variola vaccination. He was then admitted to a hospital with a tentative diagnosis of meningitis. Cerebrospinal fluid (CSF) was cloudy, the cell count 660/mm³, glucose 20 mg% and protein 124 mg%. Treatment with Ampicillin 250 mg × 4 i. v. for one week did not result in any improvement. A CSF culture yielded growth of Gram-negative "bacteroides". The diagnosis of bacterial meningitis was then abandoned and a tuberculous etiology regarded probable. Treatment was changed accordingly and various combinations of tuberculostatic drugs, including streptomycin administered. This therapy was given for 2 months during which time the clinical condition of the infant improved. However, stiffness of the neck persisted and the CSF had not become normal. CSF protein had decreased to 46 mg%. Glucose was still low, 31 mg%, and there were 10 cells/mm³ at the lowest, predominately polymenuclear cells. The supposed tuberculous etiology of the condition had not been proved by cultures or biological tests. Repeated bacterial cultures were constantly negative.

The child was then dismissed home. A few days later, in spite of continued antituberculous therapy, a recurrent bout occurred. Fever rose high. A repeat lumbar puncture yielded a highly pathological CSF.
Fig. 1. Gas myelography. Only the cervical part of the spinal cord could be outlined by gas. Thoracic and lumbar parts of spinal canal were completely occluded by swollen spinal cord.

Fig. 2. Spinal cord in situ after removal of posterior arches and dura. The cord is yellow-green and markedly thickened throughout its entire length.

The cell count was now 2900/mm³ (60% polynuclear cells), protein was 120 mg% and sugar 20 mg%. A culture yielded E. coli, bacteroides and anaerobe streptococci. High dose therapy with ampicillin, sulfonamide and chloromycetin was instituted. The infant responded favorably and became afebrile, but 4 weeks after the institution of the antibiotic therapy a new recurrence took place; paresis occurred in his right leg. These then rapidly spread to include the bladder and abdominal musculature, the anal sphincter, the left leg and finally the right arm, in that order. The infant was then transferred to this hospital.

On inspection there was a discrete pit in the skin over the lumbosacral region, with a punctate opening at its bottom. The area was dry and there were no more striking changes indicating a dysraphic state. However, X-ray examination of the lumbosacral spine showed median arch defects of L₅ and S₁, 1 cm wide.

Because of the preponderance of the paresis on the right, a pathological EEG over the left hemisphere and a tense fontanelle an abscess in the left hemisphere was suspected in addition to the obvious myelitic affection which had caused the paresis of the anal and bladder sphincters. The latter having turned into a neurogenic bladder.

*Left carotid angiography* was performed. There were no signs of any hemispheric mass, abnormal vascularization or signs of damage of the blood-brain barrier. The lateral ventricles were not dilated and there was no subdural hygroma. A nuclear brain scan was also negative.

*Gas myelography* via the lumbar route could not be performed since only a few droplets of coli-smelling pus could be withdrawn by aspiration. A *suboccipital gas insufflation* showed the changes shown in Fig. 1. Further treatment included intensive antibiotic therapy and partial extirpation of the dermal sinus tract. Death occurred 1 week thereafter. *Postmortem examination* revealed the following findings:

**Central Nervous System**

The meninges showed increased redness due to hyperemia and vascular dilatation in the meninges and there were patches of pus, especially on the base...