Recurrent Meningitis Associated with Congenital Lumbo-Sacral Dermal Sinus Tract

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Recurrent episodes of meningitis in infants and children frequently constitute a frustrating and distressing problem, both in determination of cause and in treatment. In the absence of infection elsewhere in the body, prior cranial or spinal surgery or trauma, or spontaneous cerebrospinal fluid rhinorrhea or otorrhea, unexplained recurrent meningitis has often been the cause of progressive morbidity and occasionally of mortality.

It has long been known that one of the sources for such recurrent infections is a congenital dermal sinus tract extending from the surface of the skin in to the meninges. The two most common locations for the external opening of these persistent dermal sinus tracts are in the occipito-parietal midline of the scalp and in the lumbo-sacral midline of the back.2

In 1951, Matson and Ingraham3 called attention to the intracranial complications arising from congenital dermal sinus tracts persisting in the area of the cranial epithelial ectoderm. They pointed out that a cranial sinus tract always passes inward in a caudal direction, so that if it enters the cranial vault, it does so inferior to its origin and extends within the posterior fossa still more caudally to the region of the 4th ventricle or cisterna magna. Our primary purpose in this paper is to make one point that appears to need emphasis; namely, that the same sort of embryological reasoning explains the location, extent and complications of congenital dermal sinus tracts in the lumbo-sacral region.

A series of recent cases has shown very vividly, and sometimes tragically, that the inner terminus of a congenital dermal sinus tract may be expected to extend to the neuro-ectodermal segmental level corresponding to the epithelial-ectodermal level (dermatome) of its origin. In other words, if a dermal sinus tract arising anywhere in the lumbar, sacral, or coccygeal area of the midline skin of the back extends inward all the way to its ultimate embryological level in the central nervous system, this will be to the conus medullaris. Thus, the sinus tract will always pass inward in a cranial direction, often for many vertebral segments. It may become attenuated and stop or it may expand at its inner terminus to form a cystic mass. However, if there is a history of meningitis or if there is any evidence of involvement of the nervous system, the sinus tract must be expected to extend intrathecally as far cranially as the 12th thoracic—1st lumbar vertebral level. Since the tract intervening between the external skin opening and a dermoid cyst at the inner terminus may sometimes be small and tenuous, it is essential that the surgeon treating these patients always be prepared to expose the conus medullaris if intrathecal dermoid cysts and sinus tracts associated with recurrent episodes of meningitis are not to be overlooked.

The association of bacterial meningitis with the persistence of a congenital dermal sinus extending from the skin surface to the subarachnoid space has already been well described and does not need to be discussed here; 3-7 the case material in this report will speak for itself as additional evidence.

Material

The present report is based on 8 patients recently treated on the neurosurgical service of the Children's Hospital Medical Center in Boston. The common denominator in 7 of these patients was the occurrence of intrathecal infection that was initially inappropriately managed because of failure to recognize the embryological significance of the combination of a midline lumbo-sacral dermal sinus tract and meningeal infection. Since we often learn most from a searching review of the poor results in our own experience and that of others, it seemed important to make this material available.

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In this series of patients, one had a large intraspinal abscess with profound neurological deficit. Six patients came under our care because of a history of recurring episodes of acute meningitis. The 8th patient had a congenital dermal sinus tract extending all the way from the sacral area to the conus medullaris. This lesion had been identified and completely excised prior to any meningeal infection.

In 5 of the 6 children with recurrent meningitis a previous incomplete resection of a dermal sinus tract had been carried out in another hospital. Five of these 6 children eventually proved to have an intrathecal dermoid cyst at the inner terminus of the sinus tract; the other patient with recurrent meningitis proved to have an intradural extension of the sinus tract ending at the level of the filum terminale without cyst formation.

Case Reports

Case 1. D. C. (47-06-02) was a 2½-year-old girl admitted to the Children's Hospital Medical Center, March 16, 1958, with a 3-month history of a draining sinus in the lumbar area. At the age of 20 months it was noted that she had a large distented urinary bladder, and a diagnosis of "neurogenic bladder" was made at another hospital. During the next 4 months the patient gradually became paraplegic. Incision and drainage of an abscess in the lumbar area was carried out. The patient was then transferred to the Children's Hospital Medical Center.

Examination. The child was paraplegic. There was a purulent discharge from the site of the incision and drainage. In addition to facetous paraplegia, there was fecal and urinary incontinence. A lower lumbar cutaneous angioma was noted; it was 4 cm. in diameter and had long blond hairs protruding from a midline sinus opening. There was another granulating, draining sinus to the right of the midline and pressure on the right posterior iliac crest caused the purulent drainage to increase.

Hemoglobin at the time of admission was 6.8 grams. White blood count was 13,900, 82% of which were polymorphonuclear leukocytes. Roentgenograms of the lumbo-sacral region demonstrated spina bifida of the upper sacral segments.

Operation. The right flank abscess was further incised and drained at the time of the hospital transfer. Three days later lumbar laminectomy was performed with subtotal excision of a large infected intrathecal dermoid cyst. At the time of surgery, there were 2 sinus tracts. A large amount of infected granulation tissue was noted in the extradural space. The sacral nerve roots were edematous and bound down by adherent inflammatory tissue. The midline dermal sinus tract continuous with the infected intrathecal dermoid cyst was found to extend cranially to the conus medullaris (T-12 level). The cyst wall and its contents were excised as completely as possible.

Pathological examination demonstrated stratified squamous epithelium with multi-loculated tracts lined by inflammatory granulation tissue.

Postoperative Course. The child remained severely paraplegic with only minimal recovery of sensory and motor function.

Comment. This is a most tragic result of a benign condition which progressed to severe central nervous system damage while this child was actually under hospital treatment, because of failure to recognize the significance of the cutaneous sinus associated with infection and with bladder dysfunction.

Case 2. S. E. (58-30-15) was an 8½-year-old girl admitted to the Children's Hospital Medical Center on December 8, 1964. At birth the child had had a small rounded, epithelialized cyst in the region of the 4th and 5th lumbar vertebrae. Immediately above this was a small darkened area with hair protruding from a dermal sinus tract. The patient developed normally until the age of 6 years when a pustule appeared at the opening of the dermal sinus tract. This was associated with drainage of fluid, culture of which grew B. proteus. She was hospitalized elsewhere and started on penicillin, sulfadiazine and chloramphenicol, following which she had surgical repair of what was said to be a meningocele associated with a dermal sinus tract. She did well until September, 1963, when she was admitted again to the same hospital because of headache, vomiting and limitation of motion of her lower back. Lumbar puncture at that time revealed cloudy cerebrospinal fluid. Cultures of the fluid grew 3 to 4 colonies of bacteroides. She was placed on penicillin, chloramphenicol and sulfadiazine. Even while she was on antibiotic therapy, temperature elevation associated with nuchal rigidity occasionally occurred. Intermittent episodes of twitching of the right leg were noted as well as mental confusion. Electroencephalograms demonstrated a left parieto-occipital slow wave focus. She was readmitted in March, April, and October, 1964, with repeated seizures due to bacteroides. She was transferred to the Children's Hospital Medical Center in December, 1964, for further study.

Examination. The patient was a nervous, irritable child, well developed, but in emotional distress. She demonstrated moderate nuchal rigidity and had a waddling gait. She held her spine in rigid extension and was unable to bend forward or laterally to the left. There were only 15° of straight leg raising bilaterally. The previous operative incision, although it was tender, appeared to be well healed and there was no other evidence.