An Intramedullary Epidermal Inclusion Cyst of the Thoracic Cord Associated with a Previously Repaired Meningocele

Case Report

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The concurrence of failure of fusion of the midline structures with intraspinal neoplasms of cutaneous origin is well established.6,7,10 Reported anomalies include congenital dermal sinus, spina bifida, complete rachischis of the vertebral arches, cutaneous hypertrichosis, and capillary angioma. Since the relatively common meningocele is an unusual antecedent for the relatively rare intraspinal epidermal inclusion cyst, the following case is reported.

Case Report

History. This 6-year-old white boy was one of twins born 1 month prematurely. The pregnancy was otherwise uncomplicated and his twin sister has been normal since birth. Immediately after birth, the patient was found to have a mid-dorsal meningocele without evidence of hydrocephalus or spinal cord dysfunction. Eleven days after birth, the meningocele was repaired at another hospital, the brief operative note stated that the meningocele sac was dissected down to a narrow neck and ligated. No neural elements were found in the sac, and microscopic examination of the excised specimen showed "arachnoidal tissue."

The child walked at the age of 1 year, but his gait was described by his parents as "clumsy" and he fell frequently. Bowel and bladder function were regulated well until he was 6 years old, when his parents noted occasional urinary incontinence and increasing weakness of his right leg. These symptoms developed over a 3 month period prior to his first admission to St. Louis Children's Hospital. There was no history of back or leg discomfort.

First Admission to Children's Hospital. At the time of his first admission on June 17, 1964, he had a spastic gait with bilateral extensor Babinski responses. Pain sensibility was intact in the legs, but vibratory sensation was reduced on the left. The right leg was weaker than the left. A well-healed, non-tender, surgical scar was present in the mid-dorsal region. X-ray examination of the dorsal spine demonstrated widened spacing of the pedicles locally at D-7, 8 with minimal flattening of the medial aspects of the pedicles, and bifid laminar arches of D-7 and 8 (Fig. 1). A spinal puncture showed clear, colorless fluid with a protein of 52 mg. per cent. Manometrics demonstrated a complete block. The patient's signs and symptoms did not change and he was discharged on June 23rd.

Second admission. During the month after he was discharged from the hospital, the patient developed progressive neurological disability. He was readmitted on July 21st, unable to walk without support and was incontinent of urine. Marked hyperreflexia was present in the legs with bilateral Babinski signs. A Beevor's sign was present, and vibratory sensibility was reduced in both lower extremities. No pain deficit could be found. Laminograms of the thoracic spine were ob-

FIG. 1. Anteroposterior view of the dorsal spine showing widening of the interpedicular spaces at D-7 and D-8 with slight mesial flattening of pedicles. Spina bifida is faintly visible at these 2 levels.

1018
tained prior to myelography and excluded diastematomyelia and intraspinal calcification. Myelography demonstrated a complete block at the level of the 7th thoracic vertebral body with the characteristic features of an intramedullary mass (Fig. 2).

Operation. Immediately after the myelogram, a dorsal laminectomy was performed. Upon resection of the ligamentum flavum and epidural fat, a discrete ovoid mass was found fixed to the dorsal dura at the D-7 vertebral level. The mass consisted of granulation tissue containing suture material. The dura was then opened around this nubbin of tissue, and a stalk was found extending intradurally from the mass to an oblong intramedullary tumor (Fig. 3). The spinal cord in the region of the tumor had a chalk-white cast. Through a midline dorsal myelotomy, soft caseous material containing hair was evacuated from the tumor bed. The surface of the tumor had the texture of an orange peel and a pearly sheen. Except for a few shreds of tissue, the capsule stripped readily from the interior of the thoracic cord.

A small amount of tumor capsule was firmly attached to the cord and was not resected. Microscopic sections of the tumor showed keratin and degenerating squamous epithelium, but no hair follicles in the part of the excised tumor wall.

Postoperatively the patient has done well. Three weeks after surgery he was able to walk without help and had regained urinary continence. When seen in the clinic 3 months after surgery, his plantar responses were flexor. He was attending school and his gait was almost normal.

Discussion

There is considerable variation in the nomenclature of neoplasms derived from heterotopic dislocations of skin within the neural axis. The evolution of terminology and theories of pathogenesis of these tumors have been the subjects of several reviews. According to Willis, all neoplasms derived from sequestered skin should be considered as epidermal inclusion cysts, as subdivision into "dermoid" and "epidermoid" is arbitrary and leads to confusion. The gross and microscopic appearances of the tumor in this case serve to illustrate this contention. Grossly, the tumor contained hair, as well as desquamated skin, and thus should be classified as a "dermoid." Microscopic examination, however, revealed no hair follicles, but only desquamated and atrophic epithelium, consistent with an "epidermoid." This paradox was recognized and resolved by Bostroem in 1897, and again emphasized by List in 1941. Bostroem demonstrated that frequently only a small portion of the tumor capsule, which he termed the dermoid villus, contained

Fig. 2. Frontal view during myelography with the head tilted downward demonstrates total block at the level of the centrum of D-7 with widening of the medullary shadow characteristic of an intramedullary mass. Note the normal appearance at D-8 despite the widened interpedicular space.

Fig. 3. Operative photograph after exposure of the cord showing dural granulation tissue nubbin and connecting stalk directed cephalad into the cord. The small white dorsal aspect of the epidermoid cyst can be seen at the foot of the stalk, devoid of overlying arachnoid vessels.
hair follicles and a stratum granulosum. This nidus might easily be missed in histological sampling or be present in that portion of the tumor which is not resected. The terms “epidermoid” and “dermoid” are so deeply rooted in the clinical literature, however, that the infrequent encounters with these tumors will most likely still be reported under a variety of names.

The pathogenesis of epidermal inclusion cysts is usually considered to be a result of entrapment of cutaneous ectoderm within the neural axis during the 4th or 5th week of embryonic life.\(^2\)\(^,\)\(^12\) During this critical phase of development, any failure in coalescence of the neural ectoderm can set the pattern for a variety of dysraphic states that may also implicate other germinal derivatives. It is this embryonic stage in lower animals that is most favorable for study in the field of experimental teratology. A variety of externally applied injurious agents, ranging from heat to gamma-irradiation, will result in failures of neuro-ectodermal fusion.\(^13\)

The statistical analysis of congenital anomalies in twin pregnancies has led to conflicting conclusions regarding pathogenesis. One report states that concurrent anomalies in twins are more frequent in monozygotic rather than dizygotic twins.\(^5\) This has been interpreted to indicate a “genetic” fault as opposed to an environmental injury. Another study states that frequently only one of monozygotic twins is afflicted.\(^8\) Furthermore, it has been emphasized that since monozygotic twins share a common chorion and prenatal environment, they are more likely to be exposed to a common injurious environmental factor. Indeed, Willis has commented on the dangers of deductions regarding the pathogenesis of birth defects based on studies of twin pregnancies.\(^13\)

Clinically this case is of interest in that the patient has a thoracic meningocele several years before the advent of this cyst. This is an unusual anomaly to be associated with an intramedullary epidermal inclusion cyst. This case emphasizes the need for a careful intradural appraisal of the spinal cord in the surgical management of this unusual type of meningocele.

Summary

We have reported finding an intramedullary thoracic cord epidermal inclusion cyst in a dizygotic twin who had a thoracic meningocele repaired at birth. When 6 years old, he developed a paraparesis which improved following resection of the cyst. We have briefly discussed the related nomenclature as well as theories concerned with the pathogenesis of this type of inclusion cyst.

References