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 rachischisis 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.
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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 ovoid 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.

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.

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. 5,7,11 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. 12 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." 4,9 Microscopic examination, however, revealed no hair follicles, but only desquamated and atrophic epithelium, consistent with an "epidermoid." 9 This paradox was recognized and resolved by Bostroem 6 in 1897, and again emphasized by List 4 in 1941. Bostroem demonstrated that frequently only a small portion of the tumor capsule, which he termed the dermoid villus, contained