Intramedullary Epidermoid Tumors of the Thoracic Spinal Cord
Report of Two Cases

WARD W. STEVENS, M.D.,* AND EDWARD B. SCHLESINGER, M.D.
Department of Neurological Surgery, Neurological Institute, Columbia University,
College of Physicians and Surgeons, New York, New York

Epidermoid tumors involving the spinal cord are quite rare.1,5,6,9,11,13 The opinion of the majority of authors is that these tumors arise from displaced normally-developing somatic cells. Recently, Van Gilder and Schwartz14 reported a method of producing epidermoid and dermoid tumors in the albino rat by direct skin implantation along the neuraxis; this evidence supports the concepts of a congenital origin or subsequent development after repeated lumbar puncture.

The vast majority of intraspinal epidermoid tumors are intradural and extramedullary. They are commonly associated with a dermal sinus and occur usually in the lumbar-sacral segments. In 1962, Manno, et al.,9 in a review of all reported cases, found only five tumors that they classified as being intramedullary. However, a critical review of these cases1-3,5,10,12 revealed that these were confined to the conus medullaris and extended down the dural sac; hence, they were not truly intramedullary. Gross6 reported a case in which the tumor initially appeared intradurally and upon reoperation years later was found within the substance of the cord. King,7 in 1957, reported a case quite similar to the ones we are about to describe and stated that he was unable to locate a similar case in the literature. In none of these series has a long-term follow-up been reported giving the results of operative management of a recurrent tumor or the effect of radiotherapy upon such a lesion. We are able to provide these data in the following cases.

Case Reports

Case 1. A 40-year-old man was admitted to the Neurological Institute in 1952, with an 18-month history of pain and numbness in the left leg. Approximately 12 months before admission he had noted the insidious onset of occasional spasms in the left leg at night, decreased libido, and infrequent bowel incontinence. He had also begun to drag his left foot and, on two occasions, burned the left leg without being aware of any pain.

First examination. There was slight dragging of the left foot, weakness of the distal muscles of the left leg, bilateral extensor plantar responses, and hyperactive reflexes. There was loss of perception of pain, temperature, and light touch below T-6 on the right, and bilateral decreased vibratory sense. A lumbar puncture revealed normal manometrics and a total protein content of 83 mg% in the cerebrospinal fluid. Thoracic spine films showed a mild scoliosis to the right with possible thinning of the pedicles at T-3 and T-4.

First operation. The spinal cord was explored at T-4 on the basis of the clinical localization. A midline mass was found lying almost completely within the substance of the cord, beneath the pia and arachnoid. When a small opening was made in the mass, a soft, green, sebaceous material came out; this material was gradually and carefully removed, and no hairs or skin as such were seen. It was thought wise not to remove the thin capsule which was adherent to the cord.

Pathological findings. Microscopic examination of the specimen showed cholesterol clefts and strands of keratinized desquamated material. Pathological diagnosis was epidermoid tumor.

Second examination. The patient was readmitted in 1960 with a 6-month complaint of a slowly increasing weakness and numbness in the right leg, and spasticity in both legs. He had returned to work a few weeks after his first operation with no appreciable change in his deficit. Neurological examination revealed an unsteady gait, mild

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* Present address: 2037 Crystal Spring Avenue, S.W., Roanoke, Virginia 24014.
spasticity, and bilateral leg weakness, with extensor plantar responses and sustained clonus. Sensory examination was the same as in 1952, except for an additional loss of position sense in the legs. Myelography revealed an obvious recurrence of the cyst with a partial block at the T-5 level. The cerebrospinal fluid protein was 140 mg%.

Second operation. Reexploration of the previous laminectomy revealed a thick milky-white arachnoid. Upon incising the arachnoid, the underlying cyst became apparent; the spinal cord appeared wrapped around the cyst in a thin mantle. Again, the contents of the cyst were evacuated without difficulty, leaving the capsule adherent to the neural tissues.

Third examination. The patient was readmitted in 1966 with a 6-month history of slowly increasing weakness of the right leg, fecal incontinence, and a decreased sensation of the need to void. He had been discharged 3 weeks after the second operation, able to walk without assistance and experiencing no bladder or bowel difficulty. Neurological examination revealed that the patient now had a paraparetic gait, requiring canes to maintain balance. There was marked weakness of the right leg and about 50% weakness of the left leg. Reflexes and sensory examination were as in 1960. Myelography showed evidence of a recurrence of the cyst without a significant block (Fig. 1). The cerebrospinal fluid protein was 54 mg%.

Third operation. At reexploration, essentially the same findings as in the second operation were noted. The cyst was evacuated without difficulty and a cavity was again left within the cord substance. The patient experienced an uneventful convalescence and was discharged 3 weeks later. Four months after discharge the patient was back at work and not experiencing any bowel or bladder difficulty.

Comment. This patient with an intramedullary epidermoid cyst of the thoracic cord has been followed for 15 years. Three times the cyst has been evacuated but the patient has maintained adequate functional use of the legs and remained gainfully employed. In this instance, recurrence of cystic material took about every 7 years to reproduce a significant neurological deficit. Thus we were able to effectively treat the tumor on a long-term basis by reoperation and careful intracapsular removal of the recurrent keratin material. The problem of contamination of the subarachnoid space with the contents of the cyst and the usual subsequent chemical meningitis were fortunately not encountered in this case.

The hazards of total removal of the densely adherent capsule of an intraspinal epidermoid has been well stressed in the literature.6,7,9 Most reports state that these tumors are slow-growing, and a satisfactory long-term result may be expected after a subtotal removal. However, most of the reported lesions were in the lumbo-sacral segments rather than the smaller confines of the thoracic canal.

Case 2. A 41-year-old man was first admitted to the Neurological Institute in 1956 with a 5-year history of progressive unsteadiness of gait and a more recent onset of urinary incontinence, loss of libido, numbness in both legs, and increased sensitivity to cold in the right leg.

First examination. The patient had a slightly spastic gait with bilateral extensor plantar responses and sustained clonus. A loss of the sense of pain, temperature, light touch, vibration, and position was demonstrated bilaterally below T-6. Myelography revealed an intramedullary tumor at the T2-T3 level without a block of the subarachnoid space. The cerebrospinal fluid protein was 49 mg%.

A provisional diagnosis of an intramedullary astrocytoma was made and the patient given a total tumor dose of 3200 r over a

Fig. 1. Case 1. Incomplete block at T-4 by intramedullary lesion just before third operative evacuation of the cyst.