Congenital Intraspinal Extradural Cyst*  
Report of Three Cases in One Family

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ELSBERG, et al., in 1934, described four cases of congenital intraspinal extradural cyst noting the rarity of the lesion (1% of all spinal cord tumors in their series) and the difficulty of preoperative diagnosis. Cloward and Bucy expanded Elsberg's description and discussed the common association of Sheuerman's changes in the adjacent vertebral bodies. Wise and Foster in 1955 found 34 cases recorded in the world literature.

Although a familial history has not been recorded in the previous cases, the patients in this report are three of four siblings.

The pathology as described by Elsberg is shown in Fig. 1. The cyst which usually causes erosion of the pedicles and overlying laminae visible on x-ray, is attached to the subarachnoid space by a small stalk that penetrates a dural defect. Although valves in the stalk have been described in some cases, more often the cyst communicates freely with the subarachnoid space, and the contained fluid has the chemical characteristics of cerebrospinal fluid. The cyst wall is composed of arachnoid, and in some instances the cyst itself has contained arachnoid granulations. As it enlarges, the cyst projects into the adjacent spinal foramina (Fig. 2) and, with further enlargement, compresses the dura and spinal cord, causing neurological deficits.

Case 1. This 12-year-old girl was admitted to the New York Hospital in 1960 because of progressive weakness of the legs. She had been treated previously in the outpatient clinic for chronic conjunctivitis secondary to distichiasis.

Received for publication July 19, 1967.
* Presented in part to a joint session of the New York Neurological Society, the New York Neurosurgical Society and the Section on Neurology and Psychiatry of the Academy of Medicine, January 11, 1966. This work was supported in part by a Special Fellowship (1-F11-NB-1684-01) from the U. S. Public Health Service, National Institute of Nervous Diseases and Blindness.
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Examination. On admission there was a slight percussion tenderness in the midthoracic area, weakness of both legs, more marked on the left, hyperactive knee and ankle jerks bilaterally, and extensor plantar responses. An incomplete loss of pin and touch sensation was present below the T-4 level, more marked on the right. Vibration and position sense were diminished below T-4. Plain x-rays showed widening of the thoracic pedicles; a myelogram (Fig. 3) showed an incomplete block at the T-10 level with contrast medium passing readily into a dorsal cyst that extended from T-3 to T-10.

Operation. At laminectomy the cyst was found to be completely extradural and attached to the arachnoid by a small midline stalk at T-8. It contained spinal fluid that communicated freely with the subarachnoid space. The cyst was removed intact and the dural defect secured with interrupted silk sutures.

The postoperative course was uneventful.

Fig. 1. Drawing of a spinal cross section to show the dural defect and communication with the subarachnoid space.
and now, 7 years later, there are no neurological deficits.

**Case 2.** This 12-year-old boy, a brother of the first patient, had been followed since infancy for asthma, Milroy’s disease, and chronic conjunctivitis secondary to distichiasis. Progressive weakness of both legs prompted a third admission to the New York Hospital in 1965.

**Examination.** The patient had a broad-based gait, weakness of both legs, more marked distally, hyperactive knee and ankle jerks bilaterally, and extensor plantar responses. There was also slight diminution of pain and touch sensation below the T-4 level. Vibration and position sense were markedly impaired in both legs. Percussion did not elicit spinal tenderness. Plain x-rays showed widening of the spinal canal in the midthoracic area. Myelography demonstrated an intraspinal extradural cyst partially filled with contrast material (Fig. 4).

**Operation.** A thin-walled, translucent cyst with small projections extending out of the intervertebral foramina (Fig. 2) extended from T-2 to T-10 and measured 16 cm in length and 3 cm in diameter. It contained spinal fluid and communicated freely with the subarachnoid space at T-8. Following removal of the cyst the dura was incised; the cord appeared compressed but no other abnormalities were noted.

The postoperative course was uneventful and now, 2 years later, there has been nearly complete return of neurological function. (Note: An over-penetrated chest plate had been made in our second case 9 years before this hospitalization, and retrospective analysis of the film confirms that abnormal widening of the spinal canal had occurred even at that time. This indicates that the cyst had been