Congenital Intraspinal Extradural Cyst*

Report of Three Cases in One Family

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Elsberg, et al.,3 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 Bucy1 expanded Elsberg's description and discussed the common association of Sheuerman's changes in the adjacent vertebral bodies. Wise and Foster6 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 Elsberg2 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.

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.

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**Fig. 1.** Drawing of a spinal cross section to show the dural defect and communication with the subarachnoid space.
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Fig. 2. Drawing of an intraspinal extradural cyst of the type in Case 2 shows the small projections in the intervertebral foramina.

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

Fig. 3. Myelogram in head-down position. Pantopaque has entered the cyst and demonstrates the upper margin of the cyst at T-2; pantopaque also is demonstrated in an intervertebral foramen on the left (Case 1).
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Case 3. This 2½-year-old sister of the two other patients came to our attention on routine examination of the parents and the remaining siblings. Repeated testing has not revealed any neurological deficits, but she does tire more readily than her playmates. Distichiasis is present, but there is no evidence of lymphedema. Plain x-rays of the spine show unequivocal widening of the thoracic spinal canal with erosion of the pedicles (Fig. 5). Myelography was advised but parental permission has not yet been obtained.

Discussion

The similarity of these three cases to each other and to the previously recorded cases seems obvious. In summarizing his description of the clinical syndrome, Elsberg noted:

"The individual is an adolescent with the history and symptoms of a progressive spastic paraplegia. Pain is absent or is not a prominent symptom. The objective disturbances of sensibility are slight and their upper level is in the mid-thoracic region, usually at the sixth or seventh thoracic dermatome. The manometric tests demonstrate a subarachnoid block with the characteristic spinal fluid changes of cord compression. Measurements on antero-posterior x-ray films show that the interpedicular spaces of three or more vertebrae are enlarged. The pedicles of the affected vertebrae are narrowed and atrophic."

Among all cases reported, 70% of the cysts were in the midthoracic area, and 33% were freely communicating with the subarachnoid space. Following operation, the majority of patients showed progressive improvement in neurological function. The average age was 22 years and the average duration of symptoms before diagnosis was 3 years. In most, the spastic paraplegia seemed disproportionate to the minimal sensory loss and led to errors and delays in diagnosis. Several patients had periods of remission, often follow-

Fig. 4. Myelogram in head-down position. The pedicles are eroded and the spinal canal widened. A partial block is noted at T-9. Pantopaque has entered the cyst and demonstrates the upper margin of the cyst at T-1 (Case 2).

Fig. 5. The pedicles are eroded and the spinal canal is widened from T-1 to T-7 (Case 3).
FIG. 6. These graphs depict the interpedicular distances as measured radiographically on all members of this family. Both parents and the third child have normal curves. The first, second, and fourth child demonstrate an increase of the interpedicular distance in the thoracic area. The dotted curve of the second child was obtained from review of x-rays taken 9 years before operation, and the thoracic interpedicular distances were enlarged at that time. (*C* = cervical; *T* = thoracic; *L* = lumbar)

Fig. 7. The family tree denotes familial occurrence of distichiasis, Milroy’s disease, and intraspinal extradural cysts.
ing a lumbar puncture, which led to an erroneous diagnosis of multiple sclerosis. A few had multiple operative procedures before the diagnosis of an intraspinal extradural cyst was established.

Previous authors have stressed the value of plain x-ray films of the thoracic spine and measurement of the interpedicular distances. Fig. 6 graphically illustrates their value.

Conclusion

Although a familial occurrence of intraspinal extradural cysts has not been recorded in the cases reported heretofore, our experience suggests a genetic etiology. This hypothesis is further supported by the family history of associated Milroy's disease and distichiasis (Fig. 7), both of which have been established as genetic diseases.

References