

## Congenital Intraspinal Extradural Cysts (Intraspinal Meningocele)

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**SUMMARY:** *A case of congenital, intraspinal, extradural cyst is reported with pain as the only presenting feature. In the past, the lack of pain has been considered one of the characteristics of these rare lesions. In other reported cases, when pain has been present, it was minimal and never the presenting feature. The clinical and radiographic features and the surgical treatment are described. The pathogenesis is discussed and the literature reviewed. An addition to the surgical technique is given whereby one of the post operative complications may be obviated.*

**RÉSUMÉ:** *Un cas de kyste intraspinal extradural congénital est rapporté avec comme seul symptôme la douleur. L'absence de douleur est généralement considérée comme caractéristique dans l'image clinique de ces lésions rares. Dans les autres cas rapportés, où la douleur était présente, celle-ci était minime et n'était jamais le symptôme avant-coureur. Les faits cliniques et radiologiques ainsi que le traitement chirurgical sont décrits. La pathogénèse est discutée et la littérature revue. Une addition à la technique chirurgicale est présentée par laquelle les complications post-opératoires peuvent être évitées.*

Congenital extradural cysts of the spinal canal are rare, benign lesions of unknown origin with a good prognosis following surgical extirpation.

The first verified case was that of Marks and Collins in 1915, followed by a case report of Mixter's in 1932. Elsberg reported four cases in 1934, postulating a congenital basis for these lesions. He described a clinical and roentgenological syndrome which has since been quoted widely.

"The individual is in adolescence with the history and symptoms of progressive spastic paraplegia. '*Pain is absent or is not a prominent feature.*' The objective disturbances of sensibility are slight and their upper level is in the midthoracic region, usually the sixth or seventh dermatome. Manometric tests demonstrate a subarachnoid block with the characteristic spinal fluid changes of cord compression. Measurements of A-P x-ray films show that the interpedicular spaces of three or more vertebrae somewhere between the fourth and ninth thoracic vertebrae are enlarged. Pedicles of the affected vertebrae, especially those of the sixth, seventh and eighth are narrowed and atrophic." (italics ours).

In 1935 Lehman, added two other characteristics, Schuerman's epiphysitis of vertebral bodies in the region of the lesion and marked variability of signs and symptoms of spinal cord compression as compared to steady deterioration with other types of space occupying lesions.

Good, Adson and Abbot in 1944, and Dastur in 1963, added the appearance of paravertebral rounded soft tissue shadows to the syndrome. Dastur saw these in his three cases as well as the classic widening of the spinal canal diameters and believed

they were due to lateral extensions of the cyst through the foramina.

Relief has been reported following lumbar puncture, (Davis, 1949; Good et al., 1944; Smith and Chavez, 1958). Such relief probably indicates a freely communicating cyst. With a non-communicating cyst the possibility of worsening the situation following a lumbar puncture must be considered.

Of the sixty-two cases described in the literature, four were in the cervical region, the remainder were thoracic or thoraco-lumbar. Tarlov's (1938) nerve root cysts and traumatic cysts are not included (Swanson and Fincher, 1947).

Our case is unusual in that pain was not only prominent, but was the only symptom.

### CASE REPORT

Mr. A. J., a 16 year old six foot six inch healthy student was admitted to the Winnipeg General Hospital in June 1973 with a one and one-half year history of gradually progressive, dull, nonradiating thoraco-lumbar pain. His past and family history were unremarkable.

Aside from a minimal scoliosis of the thoracic spine and a cafe-au-lait spot on his right chest, general and neurological examinations were entirely normal. Radiograms of his thoracic and lumbar spine confirmed a mild scoliosis with convexity to the left. There was widening of the interpedicular distance with associated thinning of the pedicles from T-11 to L-2, and scalloping of the posterior surfaces of the same bodies (Fig. 1).

A pantopaque myelogram demonstrated a communication at T-12 to a long accessory extension of the subarachnoid space, extending primar-

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*Figure 1a*—Anterior posterior radiogram showing thinning of the pedicles and widening of the interpedicular spaces of T-10, T-11, T-12 and L-1.



*Figure 1b*—Lateral radiogram showing scalloping of the posterior surfaces of the 12th thoracic and first lumbar. (T-11 not adequately visualized.)

ily to the right. There were lateral extensions of this accessory sac through the intervertebral foramina at T-12, L-1, and L-2 (Fig. 2).

#### Operation

A thoraco-lumbar laminectomy from T-11 to L-2 inclusive was carried out removing the spines and laminae en bloc with the connecting ligaments. A long tubular cyst grossly indistinguishable from the dura was found lying on the posterior surface of the dural tube (Fig. 3). The communication between the cyst and dural sac at the level of T-12 was ligated and divided. The lateral extensions of the cyst through the intervertebral foramina were adherent to the nerve roots with numerous fine vascular connections. The entire cyst was stripped out and removed. The block of spines and laminae were replaced and wired in situ followed by closure of the wound in layers.

The eight day post operative course was uneventful and the patient was discharged symptom free. Post operative x-rays taken nine months later revealed good union of the replaced neural arches and measurable restitution of the interpedicular spaces and of the pedicle thickness (Fig. 5).

#### Pathology

Macroscopically the cystic structure measured 9.5 cms by 3.5 cms by 0.5 cms. The inner lining was smooth and the outer wall membranous. Microscopically the wall was composed mainly of collagen bundles, suggesting dura and arachnoid. There was no inflammation or hemorrhage and no mesothelial lining. (Fig. 4.)

#### DISCUSSION

Elsburg et al., (1934) considered the cysts congenital diverticulae of the dura or protrusions of arachnoid through a congenitally weak place in the dura. Nugent et al., (1959) also thought the cysts were congenital arising from cystic dilatation of arachnoid structures associated with the nerve root sleeves. Rexed (1947) found a "more or less intense proliferation of a root arach-

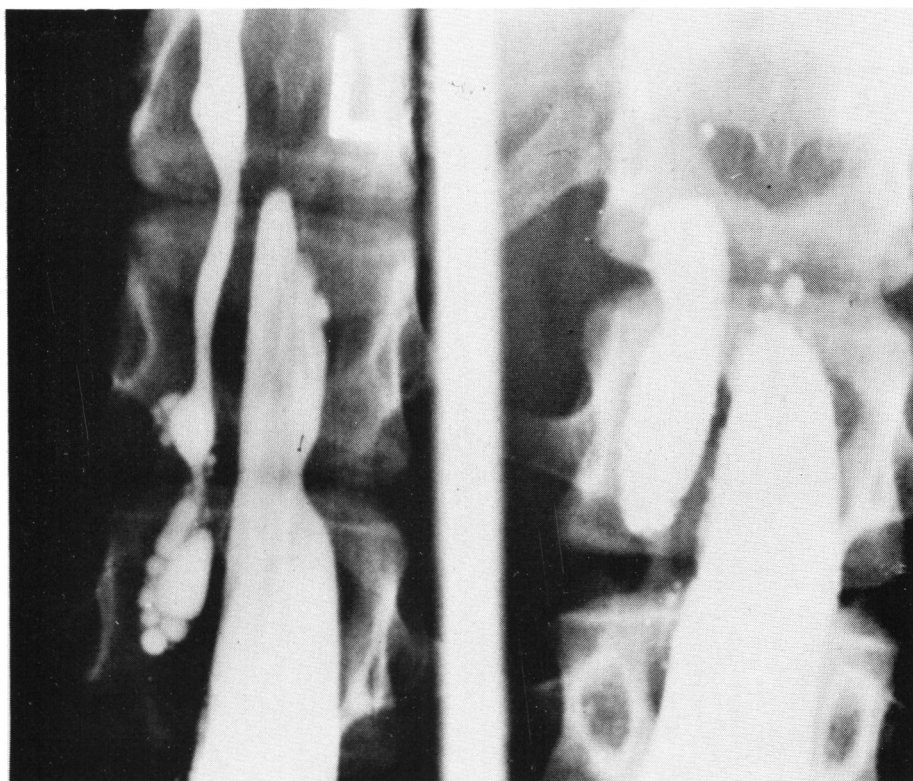


Figure 2 a—Myelogram demonstrating the two separated columns of contrast material with the cyst to the right. (Marker "L" indicates the left side in all myelograms).

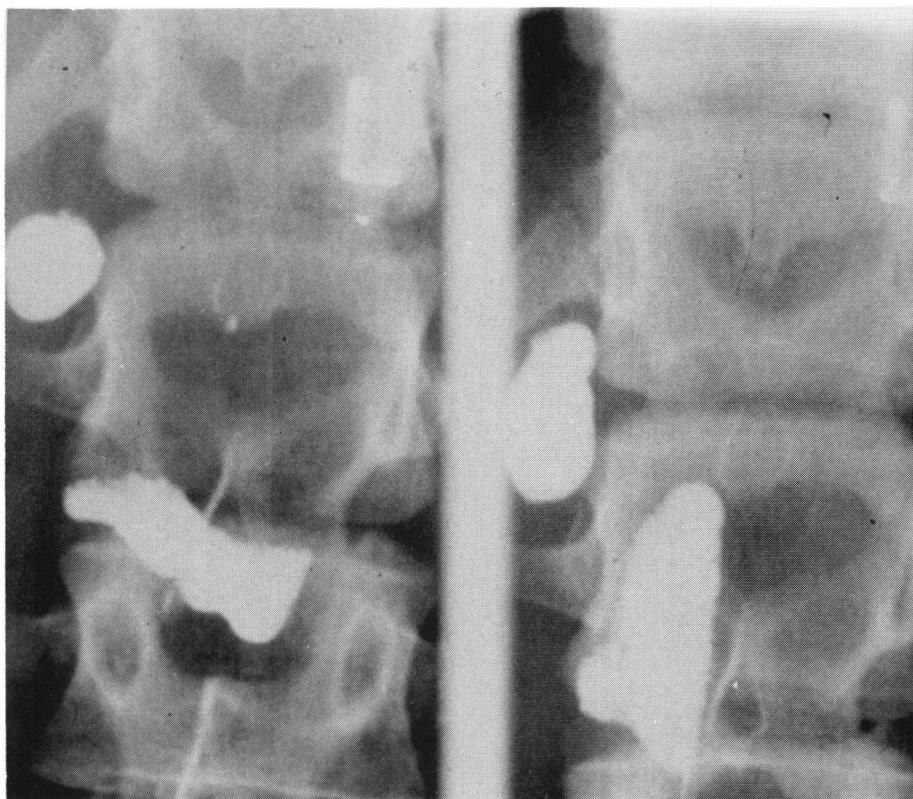


Figure 2 b—Shows the contrast material going across horizontally from the normal subarachnoid space into the cyst demonstrating the neck between the two sacs.

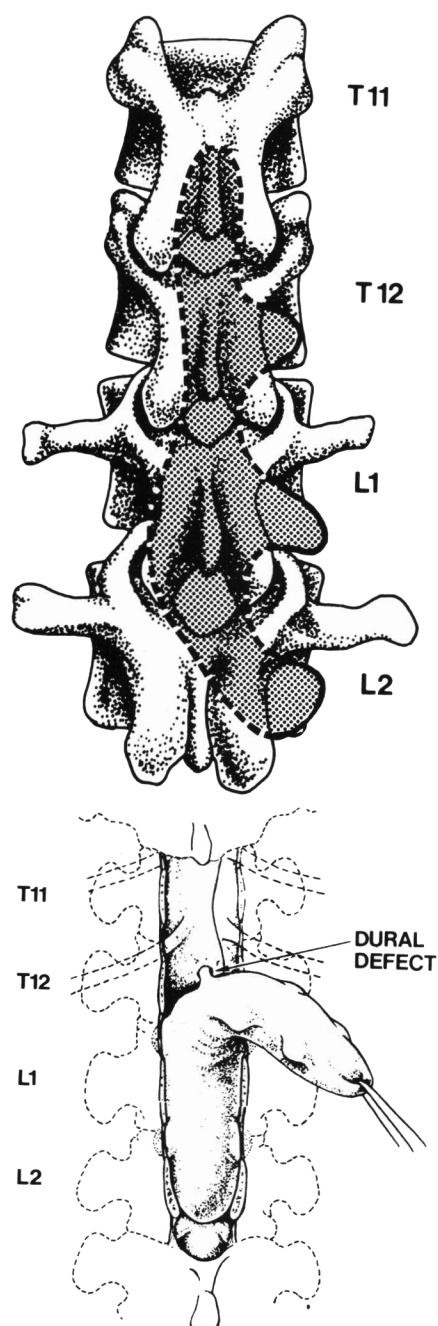


Figure 3 a & b—Artists drawings made from exposure at surgery and photographs. Figure 3 a shows the relationships of the cyst to the bony canal and figure 3 b shows its relationships to the normal dural sac which it had displaced to the left and posteriorly. The dural defect establishing a communication between the two went almost straight off to the right as the cyst lay in situ but is rotated around towards the mid line as the cyst is retracted. The myelographic connection (figure 2 b) is a more accurate representation of the direction of the communicating defect.



Figure 5—AP radiogram 9 months post operatively showing the intact neural arches wired in place, the restitution of near to normal thickness of the pedicles of T-10, 11, 12, L-1 and the restoration of the interpedicular spaces to nearly normal. The pre and post op interpedicular measurement at L-2 was identical at 28 millimeters indicating that the radiogram exposure is comparable in the pre and post operative pictures. T-10 has come from 28 to 27 millimeters, T-11 from 31 to 28, T-12 from 36-31 and L-1 from 40 to 36.



Figure 4 a—Photomicrograph of the cyst wall consisting of collagen and elastic tissue with no endothelial lining, hematoxylin and eosin X25.

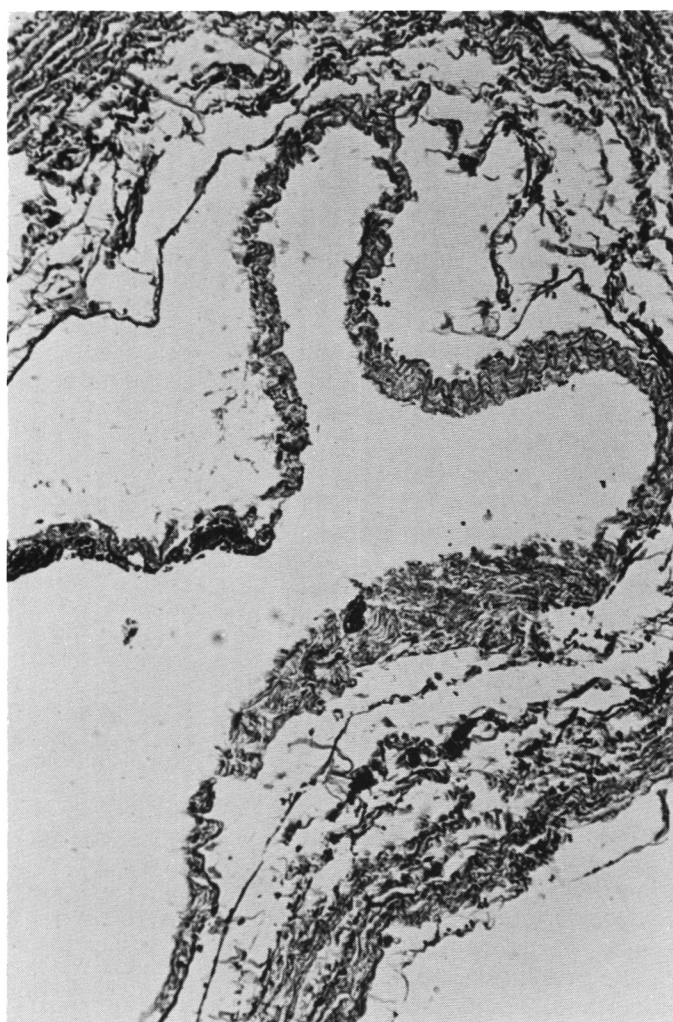


Figure 4 b—Masson trichrome X 64.

noidea", which was in some cases connected with cysts in the roots themselves. Hyndman and Gerber (1946) suggested that misplaced cell nests may be the origin.

The structure of the cyst wall and the absence of reactionary fibrosis and hemorrhage cast doubt on a traumatic origin although Mayfield (1942) and Meredith (1940), reported cases with a history of trauma and hemosiderin deposits found in the cyst wall. Hoffman (1973) documented a cyst extending from C-4 to T-1 which probably began as a nerve root avulsion but, otherwise resembled these extradural congenital spinal cysts.

Communication with the subarachnoid space has been demonstrated

in one third of the cases and in others a dural opening or attachment was noted suggesting that a communication had pre-existed and had been obliterated (Cloward and Bucy, 1937; Hyndman and Gerber, 1946; Nugent et al., 1959; Lake et al., 1974). Elsberg et al., (1934) suggested this possibility although they could not find a communication.

The dorsal midline attachment of the cyst in our patient suggests the possibility of a congenital fusion defect, ie: a lesion that might have become a classic meningocele.

Very few reported cases were associated with other congenital malformations. Balestrieri's (1958) case had an associated Dandy-Walker cyst and a bony defect of the post-

erior arch of C-4. Strang and Tovi (1961) reported an associated bony defect in the arch of the atlas. One of Gortvai's (1963) cases had an associated subluxation of the odontoid process. Cohen (1945) described an associated extradural neuroblastoma. Bergland (1968) described three siblings all with districhiasis and Milroy's disease, all with widened spaces and narrowed pedicles and two with verified cysts, the third having refused surgery.

Sex incidence of those reported is two to one, male, female. The longest verified presence of an untreated cyst is twenty-five years (Hyndman 1946).

Clinical manifestations are presumably due to varying degrees of

spinal cord compression. The intracystic pressure would exceed the intrathecal pressure only if there was a ball-valve or flap valve type of communication, or a higher protein count or a secretory lining within the cyst. Only Gortvai's case (1963) mentions a high protein, 720 milligrams percent. Secretory lining has never been described but such a lining might become attenuated and fibrosed from stretching and increased pressure.

Contrary to Elsberg's classic picture, pain is not uncommon. With the exception of our case it has usually been radiating and never the sole neurologic feature. Smith and Chavez (1958) thought the lumbar cysts should be grouped under a separate heading being different in age, sex distribution and clinical presentation from the thoracic cysts.

Cloward and Bucy (1937) and Adelstein (1941) cautioned against the possible development or worsening of dorsal kyphosis following surgery. This is a well known potential hazard from extensive laminectomy for any cause and for this reason we replaced the laminae and spines, wiring them in situ.

The present case emphasized the importance of persistent back pain and one should not await neurologic deficits before at least x-raying the spine. Definitive diagnosis of the nature of the intraspinal mass may be obtained only by careful myelography and then, only if the communication exists.

In as much as these lesions are entirely benign, surgical removal should be undertaken even though the lesion may extend over many vertebrae. With all slowly progressive extra-medullary cord lesions eventual complete recovery may be anticipated even with severe long tract disturbance. To avoid the

hazard of aggravation or development of a dorsal kyphosis replacement of the spines is suggested as a useful adjunct in the surgical management of such patients.

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