Intraspinal Enterogenous Cyst

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Congenital intradural cysts of the spine, derived from the embryonic endodermal layer, are extremely rare, and a brief review of the literature elicits no surgical recoveries. It seems fitting therefore to render a report of the surgical removal of an enterogenous intradural cyst resulting in clinical recovery.

Embryology. Such enterogenous cysts are lined with a single layer of embryonic endodermal cuboidal cells and are completely transparent, being filled with a clear, colorless and watery fluid. But the majority of congenital spinal cysts are derived from the other two primary germinal layers: the ectoderm and the mesoderm,24-10,13,14 coming under the category of dermoid and arachnoidal cysts. Even in those teratomas derived from all three layers, the endodermal layer tends to be overgrown by the other two, and may disappear entirely.2

The rare enterogenous cysts frequently are associated with spinal vertebral defects6 and mediastinal cysts,7 because of the original close contact of notochord and endoderm15 in the third week of embryonic life.1,5,13,10 Later this notochord, derived from the notochordal plate, is surrounded by the paraxial cellular mass of the mesoderm, segmenting into the spinal column; and the endoderm reforms into the primitive alimentary canal. The result may be separate or connecting cysts in the gut, in the mediastinum, or in the spinal canal, all following exfoliation of notochordal cells and possible engulfing of endodermal cells, preventing completion of the interposed spinal column.11

Historical. Fallon et al.8 have offered a most complete survey and bibliography with special emphasis on embryonic developmental factors on this subject. Guillery2 reported a case of mediastinal enteric cyst with a partial connection by funnel to a smaller enteric cyst within the spinal canal. Rhaney and Barclay16 reported 3 cases of combined deformities of gut, mediastinum, cord and spine accompanied by enterogenous cysts. Only a few cases of intraspinal enterogenous cysts have come to light7,8,11,16 and only 1 cyst was limited to the spinal canal.8

Case Report
J.G., a 28-year-old white Peruvian male, was flown to the Hartford Hospital from Peru on June 3, 1960 because of a rapidly progressing quadriplegia. Following a minor spinal injury 2 years previously pain and numbness developed down the inner aspect of the left arm. Twenty months later clear evidence of compression of the lower cervical cord had developed and he was admitted to the British American Hospital under the care of one of the authors (F.C.), who demonstrated a complete myelographic block between the 6th and 7th cervical vertebrae. He carried out a cervical laminectomy with a preoperative diagnosis of a central herniation of the intervertebral disk, and found what appeared to be a spondylosis or central herniation one space higher than the block. He "evacuated this disk transdurally," leaving the dura mater open because of "dorsal bulging of the cord." Temporary marked improvement of the quadriplegia followed but it progressed again within 3 weeks, and by the time the patient had reached the Hartford Hospital 6 weeks postoperatively he had deteriorated rapidly.

Examination. There was a nearly complete quadriplegia with spastic paresis of legs, ankle clonus, and possible Babinski's sign, urinary retention, flaccid weakness of arms, sensory loss to pin prick below the 6th dorsal level on the right, and 7th on the left, and partial posterior-column sensory loss in the legs only. Arm reflexes showed a combination of loss of upper and lower neurones with absent triceps bilaterally, absent biceps on the left, and bilateral positive Hoffmann's reflex. There were no cerebellar or bulbar signs and no fasciculations or wasting.

Laboratory tests included a normal count of blood cells; electrolyte studies, including alkaline phosphatase; and a blood sedimentation rate of 39. The spinal fluid contained total proteins of 320 mg. per cent, and sugar of 54 mg. per cent. The previously inserted Pantopaque dye was again run upwards revealing a block at the level of C6-C7 having a scalloped appearance, and a laminectomy defect centering at a level one space higher.

Operation. An emergency procedure was carried out with the patient in an upright position under local anesthesia, again with a preoperative diagnosis of probable central ruptured disk. This time bilateral facetectomies at C6-C7 interspace were carried out with electric large conical and small spherical dental drills and the laminectomy was extended downwards. The previously removed disk with ventral dural adhesion was situated about 1 cm. higher than the block and no additional protrusion of disk was found, either at the previous site or below it. The pseudodura mater, which had formed over the previously uncovered dorsal cord, was incised and opened and adhesions were cut meticulously. The cord again was found to bow sharply dorsally at the site of the block and yet no extra- or intradural solid mass was found ventrally. Rather, a small grape-sized, transparent, nearly invisible cyst presented itself ventral and lateral to the cord, filled with clear colorless
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Fig. 1. Photomicrograph (X430) of wall of the cyst showing the columnar epithelial lining. The nuclei are in a basal position. The cytoplasm contains material compatible with mucin as demonstrated by the periodic-acid-Schiff stain.

Fluid and covered by a membrane thin as tissue paper, grossly resembling an echinococcus or arachnoidal cyst. It consisted of two cyst lobules surrounding one of the motor roots. As it was being teased gently from the root it collapsed and its membrane was then removed subtotally from the ventral dura mater, surface of the cord, and motor root. The cord promptly fell forward into a normal position showing no further bulge, and yielded no fluid when pierced with the finest of hypodermic needles. Closure was carried out with fine and heavy stainless-steel sutures in the dura mater, muscles, subcutaneous layers and skin.

Course. The patient made a rapid recovery, being able to walk on the 1st postoperative day with nearly complete recovery of sensation on the 2nd day, clearing of pyramidal signs within 1 week, and discharge on the 10th postoperative day.

One month after the operation, as an outpatient, the Pantopaque was removed by the lambar route, through a 20 gauge needle, after first demonstrating an absence of the previously demonstrated block. Neurologically he showed a nearly normal gait; moderate increased reflexes of the legs without Babinski’s sign; and moderate weakness of hand grips. Neurologic findings 15 months postoperatively were entirely normal except for slight diminution of the left triceps. He had returned to Peru to full-time work within 6 weeks of his operation and writes of his continuing recovery to date.

Pathological Report. Gross examination of the material received revealed a 2×2 cm. irregular, pink-white, thin-walled membranous structure compatible with the ruptured wall of a cyst. The watery fluid contents had extruded previously. Microscopic sections (Fig. 1) demonstrated a simple columnar epithelium lining the cyst. The nuclei were round or oval and located near the base of the cells in an orderly row. No anaplasia or mitotic activity were noted. Periodic-acid-Schiff stains were positive for material compatible with mucin in the cytoplasm. Among the mucin-containing cells were occasional characteristic goblet cells akin to those seen in the intestinal tract. The epithelium in no way resembled ependyma, ruling out the possibility of an ependymal cyst.

Discussion and Conclusions

This case is of unusual interest to both pathologists and neurosurgeons. It yielded a rare enterogenous cyst, especially rare because of the absence of the usual accompanying abnormalities of spine, cord, or body cavities. Although congenital it was asymptomatic for 28 years. Clinically, it was indistinguishable from a central cervical ruptured disk and was so diagnosed by two neurosurgeons. Pathologically, grossly it resembled the more common arachnoidal or parasitic cysts; but microscopic sections revealed that the wall of the cyst was composed of a sharply defined single layer of columnar epithelium which could come only from the endodermal germinal layer. Functionally, it seems extraordinary that a cyst of such delicacy,
and near invisibility, had sufficient cohesion and strength to remain intact 28 years and ultimately
to cause a quadriplegia. Surgically, it is unusual
to effect a clinical cure (continuing 3 years to the
time of this writing) by the simple expedient of
puncturing the wall of a cyst and rather incom-
pletely tearing it from its congenital moorings.

Summary

1. Congenital intradural cysts of the spine
derived from the endoderm are extremely rare.*
2. Such cysts are lined with a single layer of
embryonic endodermal columnar cells and are
nearly invisible, being filled with clear colorless
fluid.
3. A report is made of such a grape-sized trans-
parent cyst causing rapidly progressive quadri-
paresis in a 28-year-old Peruvian man.
4. Subtotal surgical removal has resulted in a
cure to date.

* Since acceptance of this article for publication the
senior writer has attended the joint meeting of the
Neurosurgical Society of America and the Society of
British Neurological Surgeons, held June 27–29, 1963,
in London, where Charles Langmaid and Robert Jones
presented an excellent survey of 4 enterogenous cysts in
males aged 5 to 38 years, 3 verified at operation, and
only 1 limited to the spinal canal. The others were ac-
companied by bony or extraspinal anomalies or masses.
They cited an additional reference (Hoefnagel, D.,
Psychiat., 1962, n.s. 25: 159–164) to a case report of an
enterogenous cyst under the terminology of “terato-
matous cysts.”

Jack Small observed that he previously had reported
8 cases of “pre-axial enterogenous cysts” occurring in
front of the spinal axis from the cocus to the poas, at
the meeting of the Society of British Neurological Sur-
geons at Derby, November, 1961. All were removed
surgically. He compared their histology with that of
posterior mediatinal and mesenteric cysts. His only
knowledge of a published successful removal was that
described by Knight et al.14 under the terminology of
“gastrocystoma.”

Matson, Bering et al. at the Children’s Hospital,
Boston, in personal communications stated that they
have removed such cysts in some 6 to 8 children. These
cysts contained clear or cloudy fluid within a thin mem-
brane situated in or anterior to the cord, of which
roughly one-half were associated with retropleural or
cervical extensions through holes in the vertebrae. They
will be published in a monograph on intraspinal tumors
in children. One case of thoracic neurocystic cyst has
already been published by Holcomb, G. W., Jr. and

The senior writer has included this rather lengthy
footnote to correct certain false impressions mistakenly
offered in the first paragraph. In spite of rather sparse
publications and confusing nomenclature, “enteroge-
nous cysts” probably are not too rare. Successful surgical
removal is not infrequent. Only one-half of such cysts
were similar to that described in the present report; the
remainder were of quite different consistency, location,
and terminology.—W.B.S.

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