Cystic lesions within the spinal canal are generally the result of specific or nonspecific inflammatory lesions, tumors, traumatic hemorrhages, or hemorrhages from vascular malformations. True congenital intraspinal cysts appear to be very rare. On the basis of the teratogenic "determination period" and the germinal layers that are involved, it is possible to distinguish between intraspinal-extradural, intradural, teratoid, teratomatous, enterogenous, and arachnoid cysts. "Choroidoependymal cysts," and bronchogenic intraspinal cysts are rare and have only a curiosity value.

Enterogenous cysts occupy a special position among congenital abnormalities. Indirect evidence of their teratogenic determination period can be obtained from the histological findings. They are examples of the induction, determination, and organizing ability of the tissues. Although initially the cells still show a marked prospective potency, their development is later completed within quite narrow limits. As the prospective potency or plasticity of the tissue becomes smaller, their future significance is much greater, because their ability to replace defective tissue is much less. Enterogenous cysts represent inhibiting anomalies of the alimentary tract (that is, of the endoderm), which, because of the teratogenic determination period, may be found solely in the mediastinum, in both the mediastinum and the spine, or else purely intraspinally. In isolated intraspinal enterogenous cysts, attachment to the parent tissue (that is, the endoderm) has been lost.

The clinical symptoms associated with enterogenous cysts depend on the site of the lesion, and are not typical for all such cysts. A definitive diagnosis can only be made by biopsy and histological examination. In view of their rarity, we describe here a patient with this entity who has recently been under our care. Based on this case and a study of the literature, we have tried to indicate some of the special features of these anomalies.

Case Report

This 39-year-old woman had an episode of severe neck pains, associated with disturbance of sensation and fleeting paresis of the upper limbs 20 years before her current admission. Since then she had suffered further neck pains, with radiation into the right arm. Six weeks before admission, she experienced burning pains in the right arm, and later also in the left arm. She had increasing sensorimotor tetraparesis with bladder disturbances.

Examination. On admission, she exhibited sensitivity of the cervical spine to pressure and percussion. Neurologically there was a tetraparesis, more marked in the upper extremities, and also bilateral dissociated sensory disturbances below the C-4 vertebral level. A lateral x-ray film of the cervical spine showed possible erosion of the posterior margin of C-3 (Fig. 1 left). On
Enterogenous intraspinal cysts

FIG. 1. Left: Lateral film of the cervical spine showing a slight depression (dimpling?) of the posterior aspect of the body of C-3. The spinal canal appears slightly widened. Right: Myelogram with Amipaque injected from below. An atypical complete obstruction is apparent at the C-5 vertebral level.

the anteroposterior films there was a suspicion of a widening of the spinal canal between C-3 and C-5, with thinning of the pedicles. A lumbar puncture with a combined Queckenstedt test showed evidence of a complete block. Myelography with injection of Amipaque from below showed total obstruction at C-5 (Fig. 1 right), suggesting an intradural tumor.

Operation. At operation there was thinning of the laminae from C-1 to C-6, and the inferior margin of the arch of the atlas was eroded and atrophic. The dura was tense and did not pulsate. The spinal cord was expanded at the C3-4 level. On the left side a cystic tumor was seen which was grayish-white and showed yellowish flecks of calcification; its contents were turbid.

Histological examination of the cyst wall (Fig. 2) revealed an epithelial layer with no evidence of nervous tissue or any other tissues. The material was positive to periodic acid-Schiff (PAS) staining. A diagnosis of endodermal cyst was made.

Postoperative Course. Postoperatively, the neurological deficits showed a steady improvement. Five months after the operation the patient was again able to walk unaided.

Discussion

The pathogenesis of enterogenous anomalies is complicated, but it can be explained by the developmental history of the individual lesions. Their occurrence in association with a spina bifida is well known.\textsuperscript{58} The splitting of the notochord has been described as the “notochord syndrome” by Bentley and Smith,\textsuperscript{5} as “disruption of the ventral field” by Voth, \textit{et al.},\textsuperscript{82} and as “entodermal diastematomyelia” by Cecotto, \textit{et al.}\textsuperscript{13} These lesions can present in different forms, from the most severe anomalies that are incompatible with life\textsuperscript{13} to milder pathological abnormalities with diverse variations,\textsuperscript{24} as was shown by Rise\textsuperscript{69} and Bentley and Smith,\textsuperscript{5} in cases confirmed by biopsy and autopsy. They range from the most severe clefts of the medullary plate, the vertebral bodies, and arches, with herniation of the alimentary canal into the vertebral canal, to involution of the neuroenteric connection with merely a persistent strand of tissue between the intestine and neural tube (Fig. 3).

Bentley and Smith\textsuperscript{5} separated enterogenous anomalies into: 1) visceral adhesions; and 2) spinal and ventral nervous anomalies. Among the visceral anomalies are fistulas, sinuses, diverticula, and enteric cysts. Among the spinal ventral and nervous anomalies are splits in the dorsal tissues of the embryo, including a split notochord.

Feller and Sternberg\textsuperscript{21} also described the following variations of enterogenous cysts: 1) The alimentary canal leads through a split vertebral body, the cord, and the vertebral arches to the skin surface, where an “area medullo-vasculosa” is recognizable (Fig. 3a).\textsuperscript{5,10,13,14,31,41,52,57,64,65,70,71,81} 2) The alimentary canal drags dura through the split vertebral body and the
FIG. 3. Diagrams showing the various possible appearances of enterogenous anomalies. Squared shading indicates endoderm, broken lines mesoderm, and diagonal lines ectoderm.

card is also split, like a diastematomyelia (Fig. 3b, c, and d). The gut is closed and there is merely a strand of tissue extending through the vertebral body; the neural tube is open. Histologically the strand may consist of ectodermal and endodermal tissue (Fig. 3c).

4) The gut and the neural tube are closed. There is merely a cleft in the vertebral body. Within this cleft there may be or else a diverticulum between the closed gut and the closed neural tube (Fig. 3i).

If there is no strand of tissue but merely an intraspinal cyst, the cyst can lie in variable relationship to the spinal cord (Fig. 3e, f, and g). Sometimes enterogenous cysts are found only in the mediastinum with or without an associated vertebral defect, such as fused vertebrae or splits in the vertebral bodies.

Both diverticulum of the esophagus and duplication of the alimentary tract have a later teratogenic determination point.

Figure 3 summarizes these various possibilities. Quite often true endodermal intraspinal cysts are confused with other congenital cystic anomalies, which are composed of other embryonic layers, as for example the cystic teratoids or the cystic teratomas. A true enterogenous cyst only shows endodermal elements, so that the epithelium can show those structures that are also identifiable during the normal development of the epithelium. If there is histological evidence of one other germinal layer, a teratoid should be considered, and if there is evidence of all three layers, a teratoma may be present. The inductive potentiality, the prospective plasticity, and the particular determination periods of the three germinal layers play a decisive role in determining the nature of the lesion.

Oehlcker stated in 1909 that the particular teratogenic determination period for spina bifida anterior must be looked for in the 1st month of embryonic life (that is, in the early stage of the cartilaginous vertebral anlage). In 1911, Budde believed that persistence of the neurenteric canal was the cause of spina bifida anterior. It could interfere with the formation of the notochord and hence with the closure of the anlage of the vertebral bodies. The faulty separation between the endoderm and the notochord can be regarded as the chief characteristic of the split vertebral bodies. The teratogenic determination period is therefore in the presomite phase. It appears that at this stage a sort of adhesion exists between the endoderm and the notochord. During the first 3 weeks of embryonic life, the cells of the notochord are in intimate contact with the endoderm (primitive foregut) and separation occurs slowly. However, if this separation of the two embryonic layers is not complete, the mesoderm (that is, the somites) meets with resistance, so that a small piece of the primitive gut can become trapped in the developing spinal canal. According to Hamilton, et al. such endodermal anomalies must be the result of multiple adhesions between the amnion and yolk sac during the 3rd week of development.

Other authors also place this critical teratogenic determination period in the 3rd week, and implicate as the cause of the endodermal anomaly strips of cells that migrate further dorsally and hence cause faulty closure of the somites. As the prospective potency of the germinal layer is most marked in the cervical region, incomplete fusion results from the inclusion of the foreign tissue (endodermal tissue), and consequently, a cyst develops. Further caudally, the organizational ability of the tissue diminishes, and the endodermal cell nests lead to diastematomyelia, as is undoubtedly shown by a case reported by Keen and Coplin in 1906. According to Feller and Sternberg, these endodermal cell nests must be in the region of the primitive node (that is, at the particular place where all three germinal layers are found). The disparity in level between the intraspinal and extraspinal portions of the endodermal cysts, which may be demonstrated at autopsy or operation, is accounted for by the fact that, during the course of development of the mediastinum, a caudal migration takes place. The intrathoracic portion of the cyst migrates with the mediastinum, while the vertebral anomalies and the intraspinal portion are "left behind." Persistent primitive anatomy is said by some
Enterogenous intraspinal cysts

**TABLE 1**

*Summary of reported cases of enterogenous intraspinal cysts*

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Age (yrs), Sex</th>
<th>Location of Cyst</th>
<th>Neurological Findings</th>
<th>Vertebral Changes</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Puusepp, 1934</td>
<td>27, M</td>
<td>subdural, C-3</td>
<td>gradual tetraplegia</td>
<td>none</td>
<td>cured</td>
</tr>
<tr>
<td>Adams &amp; Wegner, 1947</td>
<td>41, M</td>
<td>intradural, T11-12</td>
<td>paraparesis</td>
<td>none</td>
<td>improved</td>
</tr>
<tr>
<td>Case 1</td>
<td>29, M</td>
<td>subdural, C3-4</td>
<td>gradual paraparesis</td>
<td>none</td>
<td>improved</td>
</tr>
<tr>
<td>Harriman, 1958</td>
<td>33, M</td>
<td>intradural, T5</td>
<td>gradual paraparesis</td>
<td>none</td>
<td>died</td>
</tr>
<tr>
<td>Adams, 1960</td>
<td>6, F</td>
<td>extradural, C5-6</td>
<td>gradual hemiplegia</td>
<td>none</td>
<td>cured</td>
</tr>
<tr>
<td>Hoefnagel, et al., 1962</td>
<td>28, M</td>
<td>intradural, C6-7</td>
<td>progressive paraparesis</td>
<td>yes</td>
<td>cured</td>
</tr>
<tr>
<td>Case 1</td>
<td>38, M</td>
<td>intradural</td>
<td>paraparesis</td>
<td>none</td>
<td>cured</td>
</tr>
<tr>
<td>Case 2</td>
<td>11, M</td>
<td>intramedullary</td>
<td>disturbance of sensibility</td>
<td>scoliosis</td>
<td>improved</td>
</tr>
<tr>
<td>Case 3</td>
<td>5, M</td>
<td>intramedullary</td>
<td>diastematomyelia</td>
<td>yes</td>
<td>improved</td>
</tr>
<tr>
<td>Case 4</td>
<td>7, M</td>
<td>intradural</td>
<td>monoparesis</td>
<td>none</td>
<td>spinal</td>
</tr>
<tr>
<td>Rewcastle &amp; Francoeur, 1964</td>
<td>15, M</td>
<td>intradural, T12-S2</td>
<td>intermittent monoparesis</td>
<td>none</td>
<td>cured</td>
</tr>
<tr>
<td>Case 2</td>
<td>18, M</td>
<td>intradural, T11-L3</td>
<td>intermittent paraparesis</td>
<td>none</td>
<td>cured</td>
</tr>
<tr>
<td>Case 3</td>
<td>27, M</td>
<td>intradural, T11-L1</td>
<td>gradual paraparesis</td>
<td>none</td>
<td>improved</td>
</tr>
<tr>
<td>Case 4</td>
<td>49, M</td>
<td>intradural, T11-L3</td>
<td>intermittent paraparesis</td>
<td>yes</td>
<td>improved</td>
</tr>
<tr>
<td>Case 5</td>
<td>15, F</td>
<td>intradural, C5-6</td>
<td>progressive paraparesis</td>
<td>yes</td>
<td>improved</td>
</tr>
<tr>
<td>Case 6</td>
<td>34, F</td>
<td>intramedullary</td>
<td>progressive paraparesis</td>
<td>none</td>
<td>improved</td>
</tr>
<tr>
<td>Levin &amp; Autin, 1964</td>
<td>39, F</td>
<td>intradural</td>
<td>gradual paraparesis</td>
<td>yes</td>
<td>cured</td>
</tr>
<tr>
<td>Németh, 1965</td>
<td>8 days, M</td>
<td>upper thoracic</td>
<td>diastematomyelia</td>
<td>yes</td>
<td>died</td>
</tr>
<tr>
<td>Brun &amp; Saldeen, 1968</td>
<td>2, M</td>
<td>intradural</td>
<td>unknown</td>
<td>none</td>
<td>died</td>
</tr>
<tr>
<td>Kahn, et al., 1971</td>
<td>28, M</td>
<td>intradural, C3-4</td>
<td>tetraparesis</td>
<td>none</td>
<td>improved</td>
</tr>
<tr>
<td>Case 2</td>
<td>6, F</td>
<td>intradural, C6-T1</td>
<td>tetraparesis</td>
<td>yes</td>
<td>improved</td>
</tr>
<tr>
<td>Case 3</td>
<td>37, M</td>
<td>intradural, T11</td>
<td>paraparesis</td>
<td>none</td>
<td>improved</td>
</tr>
<tr>
<td>Klump, 1971</td>
<td>6 wks</td>
<td>intradural, C5-T1</td>
<td>hemiparesis</td>
<td>yes</td>
<td>improved</td>
</tr>
<tr>
<td>Silvermail &amp; Brown, 1972</td>
<td>12, M</td>
<td>intradural intramedullary, T2</td>
<td>gradual tetraparesis</td>
<td>none</td>
<td>improved</td>
</tr>
<tr>
<td>Desphande, et al., 1972</td>
<td>intraspinal</td>
<td>unknown</td>
<td>unknown</td>
<td>unknown</td>
<td>unknown</td>
</tr>
<tr>
<td>Palma &amp; Di Lorenzo, 1976</td>
<td>Case 1</td>
<td>20, M</td>
<td>intradural, C7-T2</td>
<td>gradual paraparesis</td>
<td>none</td>
</tr>
<tr>
<td>Case 2</td>
<td>21, M</td>
<td>intradural, C5-6</td>
<td>paraparesis</td>
<td>none</td>
<td>cured</td>
</tr>
<tr>
<td>Case 3</td>
<td>43, F</td>
<td>intradural, C3-5</td>
<td>paraparesis</td>
<td>none</td>
<td>improved</td>
</tr>
<tr>
<td>Case 4</td>
<td>48, F</td>
<td>intradural, C5-6</td>
<td>paraparesis</td>
<td>none</td>
<td>cured</td>
</tr>
<tr>
<td>Case 5</td>
<td>18, M</td>
<td>intradural, cervical</td>
<td>tetraparesis</td>
<td>none</td>
<td>cured</td>
</tr>
<tr>
<td>Fabini &amp; Adams, 1979</td>
<td>54, F</td>
<td>intradural, C1-2</td>
<td>gradual tetraparesis</td>
<td>none</td>
<td>improved</td>
</tr>
<tr>
<td>Agnoli, et al., 1984</td>
<td>39, F</td>
<td>intradural, C1-6</td>
<td>gradual tetraparesis</td>
<td>yes</td>
<td>improved</td>
</tr>
</tbody>
</table>

* Synopsis of all cases found in the literature in which PAS reaction was positive and the intraspinal cysts contained mucin.

Authors to be the cause of an abnormal attachment between the endoderm and ectoderm, with possible splitting of the cord. Defective migration of the neurenteric canal or an additional neurenteric canal with forking of the notochord ("split notochord") has also been proposed as a cause of endodermal cysts.

These reported causes of endodermal cysts, some hypothetical and some based on operative findings, show how diverse their manifestations can be. One must therefore, as already mentioned, distinguish between cysts arising from three, two, or only one of the germinal layers. The multivarious manifestations of these cysts are reflected in their nomenclature. Kubie and Fulton, et al., Rewcastle and Francoeur designated these anomalies as "teratomatous cysts." Knight, et al., referred to these lesions as "gastrocytoma," and Hosoi and Puusepp as "intestinoma;" only Neuhauser, et al., called them "archenteric cysts."

Bucy and Buchanan used the term "teratoma," in order to emphasize particularly their make-up from the three germinal layers. Adams and Wegner likewise grouped these tumors among the teratomas. Rewcastle and Francoeur, on the other hand, used the designation "teratomatous cysts," for the histologically true endodermal cysts. The term "intraspinal enterogenous cyst" originates from Harriman.

In Table 1 we have summarized true histologically verified intraspinal endodermal cysts reported in the literature. The histological demonstration of mucin is highly suggestive of endodermal tissue and at the same time the PAS reaction should also be positive. According to these criteria,
the first case in the literature was described by Puusepp in 1934. The patient was a 27-year-old woman with a 17-year history of symptoms. The start of the illness coincided with an injury and it continued with repeated episodes associated with pains. On admission, the patient had a tetraparesis. There was no evidence of any abnormality in the spinal column. The cyst was situated subdurally between C-3 and C-4, and myelography showed an incomplete obstruction. After operation there was a complete recovery of function. Histologically there was evidence of mucous glands, submucous muscle fibers, and a cylindrical epithelium. This report is strikingly similar to our own case. However, endodermal cysts can be an incidental finding at autopsy, after having been completely asymptomatic during life. In about one-half of the cases, the progress of the illness is episodic so that it may be difficult to distinguish this disorder from multiple sclerosis. Males show a 3:2 predominance over females. The lesions are more frequently found in the cervical than in the thoracic or lumbar regions. The cyst usually extends at the most over three segments. In about half of the cases there is vertebral involvement such as spina bifida or fused vertebrae. Evidence of secondary changes in the vertebrae such as widening of the spinal canal is uncommon. The myelogram is not diagnostic and merely suggests an intradural space-occupying lesion.

In about 70% of cases there is complete regression of the neurological deficits after a successful operation. Operative death is very rare. Endodermal cysts are benign congenital tumors, and so far only rare cases of metastasis or recurrence has appeared in the literature.5

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A. L. Agnoli, A. Laun and R. Schönmayr


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Address reprint requests to: Agnolo L. Agnoli, M.D., Neuroradiological Department, University Kliniken Giessen, D-6300 Giessen, Federal Republic of Germany.