Intramedullary enterogenous cyst of the spinal cord

Case report

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The occurrence of an intramedullary enterogenous cyst of the spinal cord is a very rare phenomenon. A review of the English literature yielded only four cases. The nomenclature of such a lesion varies from author to author, includes the terms “teratomatous cyst” and “enterogenous cyst,” probably reflecting uncertainty as to the etiology. We describe another such case.

Case Report

This 54-year-old man presented with an 8-year history of intermittent and recurrent episodes of low-back pain, diffuse persistent and painful dysesthesia from his scapulas radiating down to his legs, and progressive weakness and numbness from the mid-thoracic level downward. The symptoms were more severe on the left side. An episodic attack would last from 3 to 4 days to about 3 months. The symptoms would then subside slowly and spontaneously. Over the last 8 years, the patient experienced progressive neurological deficits accelerating over the final 12 months. The impairment of temperature sensation had resulted in him scalding his legs on several occasions. He also suffered from urge incontinence of urine and had been impotent for the last 5 years. He had been noted to have a progressive thoracic kyphoscoliosis since childhood.

Examination. The patient looked well and was normotensive. There was a small pigmented cutaneous patch over the point of maximum curvature of his kyphoscoliosis at the region of the T-7 vertebra. The left lower limb was grossly wasted and was paretic with a Grade I muscle power (MRC Scale 1944). The right lower limb, however, was normotonic with a Grade IV muscle power. There was hypoesthesia from the T-8 dermatome downward. Temperature perception was bilaterally affected, with gross impairment on the left side. Vibration sensation and proprioception were impaired only on the left side. The anal sphincters were intact. The reflexes were brisk bilaterally. Plantar response was upgoing on the left side, but no clonus was elicited.

A plain film of the vertebral column showed a hemivertebra at T-11, vertebral fusion of the mid-thoracic vertebrae with a kyphoscoliosis convex to the left, and a Grade II spondylolisthesis of L-5 on S-1. Metrizamide lumbar myelography showed a large filling defect at the T4-5 level due to an intramedullary lesion (Fig. 1). The cerebrospinal fluid contained 65 mg% protein.

Operation. A thoracic laminectomy was performed from T-3 to T-5. The dura was observed to be bulging and soft, and, upon opening the dura, a cystic swelling protruded through. There was no clear evidence of the cord. Milky fluid was aspirated from the cyst. Under the operating microscope, an incision of approximately 2 cm was made along the cyst with the carbon dioxide laser; this subsequently revealed a large cavity from which more fluid was aspirated. The cavity was smooth and opalescent at the base where
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the vertebral bodies were in close contact. It could be seen that the cyst was completely within the medulla, and the spinal cord was stretched around it. A full marsupialization was performed, and a small amount of cyst wall was taken for histological study. Histologically, the cyst contained collagenous tissues, and a portion of it was lined by a single layer of columnar epithelium. No mucus was identified and periodic acid-Schiff staining was negative (Fig. 2).

Postoperative Course. The patient made an uneventful recovery. On the 1st day after surgery, the muscle power in his left leg improved from Grade I to Grade II. By the 5th day, the grading had increased to III+, and the right leg had attained a Grade V power. Sensory recovery was slow, although subjectively he was satisfied that there was some improvement; however, objectively, none was detected. He was discharged 2 weeks postoperatively and within 2 months of surgery he was walking up to a mile at a time.

Discussion

A review of the English literature indicated that four such patients have been reported previously, two of whom were children. The age range was from newly born to 49 years. The sex distribution was 3:1 in favor of males. The lesions were found in the cervical and thoracic regions. It is interesting, and perhaps significant, to note that the children whose lesions were in the cervical spine also had the more severe form of associated congenital abnormalities. The two adult patients and our patient all had thoracic lesions, and their associated congenital abnormalities were less severe and less debilitating.

The most common clinical presentation was that of an intermittent course of minor complaints stretching over periods of months, or maybe years, until finally compensatory mechanisms failed, resulting in severe neurological deficits. The presenting symptoms were nonspecific back pain, paresthesia, and weakness of the affected limbs. The clinical course of the five cases is summarized in Table 1, and a histological classification is given in Table 2.

The pathogenesis and embryogenesis of enterogenous cysts in the spinal cord have been the subjects of much debate in the past; however, no consensus has been established with regard to the etiology. Rhaney and Barclay suggested that the intraspinal enterogenous cyst was a product of abnormal separation of germ layers. Early in embryonic development, the primitive mesoderm, which gives rise to the notochord, comes to lie in close contact with the endoderm, a process called "intercalation." At a later stage when separation occurs ("excalation"), groups of endodermal cells may be carried back with the mesoderm and give rise to the enterogenous cyst. Hamilton, et al., stated that the ectoderm of the primitive streak is capable of forming both endoderm and paraxial endoderm. If and when detachment of the cells from Hensen's node occurs during its caudal migration, then maturation of these cells could give rise to bronchogenic or enterogenous cysts. Rhaney and Barclay, however, did not consider these enterogenous and bronchogenic cysts as teratomas. The types of lesions described by Hamilton, et al., as far as they are concerned, were distinct from intramedullary enterogenous cysts both from the embryological and from the clinical points of view. Hoefnagel, et al., examined the observations made by Hunter and Lennox on nuclear sexing of teratomas. In female patients there was always a Barr body in the teratomatous cells, whereas in male patients there may or may not have been a Barr body in the cells. The presence of a nuclear sex chromatin in the cells of male patients would be significant. Hoefnagel, et al., determined the nuclear sex of the cells in their case of spinal cyst in a female patient and found Barr bodies to be present. With the aid of cross-references from authors of four previously published cases in male patients, they obtained a result of two out of four positive findings. (The cases selected were not specified as intramedullary enterogenous cysts, but as spinal cysts.) They accordingly considered these cysts to be teratomatous in nature.

Fig. 1. Metrizamide lumbar myelogram showing the intramedullary lesion as a large filling defect.
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Fig. 2. Photomicrographs of the cyst showing a portion lined with ciliated columnar epithelium. Left: H & E, x 249. Right: H & E, x 480.

Table 1
Clinical summary of five cases of intramedullary cyst of the spinal cord

<table>
<thead>
<tr>
<th>Authors, Year</th>
<th>Age (yrs), Sex</th>
<th>Level of Lesion</th>
<th>Nature of Cyst Fluid</th>
<th>Histology of Cyst</th>
<th>Associated Congenital Abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rhaney &amp; Barclay, 1959</td>
<td>neonate, M</td>
<td>upper cervical</td>
<td>not recorded</td>
<td>gastric wall lining</td>
<td>Arnold-Chiari malformation; lumbosacral spina bifida &amp; meningomyelocele; hydrocephalus; microgyria; congenital heart disease</td>
</tr>
<tr>
<td>Rewcastle &amp; Francoeur, 1964</td>
<td>12, M</td>
<td>C2-T2</td>
<td>black fluid</td>
<td>pseudostratified columnar epithelium; mucus-secreting</td>
<td>11 ribs on lt side; hemivertebrae T-4 &amp; T-6; multiple bone abnormalities, C-3 to T-3; mediastinal enterogenous cyst; gastrointestinal abnormalities</td>
</tr>
<tr>
<td>Silvernail &amp; Brown, 1972</td>
<td>34, F</td>
<td>T-10</td>
<td>cloudy</td>
<td>stratified cuboidal epithelium; mucus-secreting</td>
<td>none</td>
</tr>
<tr>
<td>Rosenbaum, et al., 1978</td>
<td>49, M</td>
<td>T-9</td>
<td>cloudy</td>
<td>stratified ciliated columnar epithelium; not mucus-secreting</td>
<td>anomalous vertebrae, L4–S1</td>
</tr>
<tr>
<td>Kwok &amp; Jeffreys, 1982</td>
<td>53, M</td>
<td>T4-5</td>
<td>cloudy</td>
<td>simple ciliated columnar epithelium; not mucus-secreting</td>
<td>rt progressive midthoracic scoliosis; hemivertebra T-11; spondylolisthesis, L-5 on S-1</td>
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Intramedullary enterogenous cyst of the spinal cord

TABLE 2

<table>
<thead>
<tr>
<th>Type</th>
<th>Classification</th>
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<tr>
<td>Type I</td>
<td>simple or pseudostratified columnar or cuboidal epithelium with or without cilia</td>
</tr>
<tr>
<td>Lying on a basement membrane</td>
<td></td>
</tr>
<tr>
<td>Type II</td>
<td>Type I plus mucus glands, serous glands, smooth muscle, fat, cartilage, bone, elastic fibers, lymphoid tissues, or nerve ganglion</td>
</tr>
<tr>
<td>Type III</td>
<td>Type II plus ependymal or glial tissue</td>
</tr>
</tbody>
</table>

* Classification is modified from Wilkins and Odom. 9

Rewcastle and Francoeur 9 made the observation that intraspinal cysts were associated with mesodermal abnormalities such as hemivertebrae, spina bifida, and similar cysts in the thoracic cavity, which led them to believe that the anomalies were not simply displacement of normal developing somatic cells but were teratomatous in origin. Their determination of nuclear sexing of their series of four male patients also yielded positive results in two cases.

Rosenbaum, et al., 7 agreed with the suggestion by Hoefnagel, et al., 3 and Rewcastle and Francoeur 5 that these cysts were teratomas. Their proposal for differentiating the two types of cyst could be based upon the histological findings: if the epithelial lining contained no cilia, the diagnosis of a teratoma could not be made; in other words, teratomatous cysts are lined by ciliated epithelium only. Therefore, they retained their reservation that perhaps enterogenous cysts and teratomatous cysts were separate entities. However, the differentiation ought to be viewed in relation to the embryogenesis rather than from the histological variants, as ciliated columnar epithelium can be formed in the rostral end of the yolk sac endoderm.

The similarities in the presentation and in the histology of these intramedullary cysts led to our belief that intramedullary teratomatous cysts and intramedullary enterogenous cysts were a single entity. In the past, both intramedullary and extramedullary cysts were classified together, hence the confusion in nomenclature. Embryologically, despite the histological similarity, intramedullary and extramedullary enterogenous cysts must have derived from separate tissues and, therefore, it is not justifiable to classify them together as the same entity. The evidence derived

![Fig. 3. Diagrammatic illustration of a midline section through an early embryo showing the neuroenteric canal extending forward beneath the neural plate dorsally and the endoderm ventrally. (Reproduced with permission from Harrison RG: Development of the vertebral column, in Owen R, et al (eds): Scientific Foundations of Orthopaedics and Traumatology. London: William Heinemann Medical Books, 1980, p 163.)](image-url)
from nuclear chromatin studies by various authors was based on intraspinal intramedullary cysts, and their results suggested that these were teratomatous in origin. However, one could not say that this would also apply to the intramedullary cysts.

For a cyst to be located inside the spinal cord, the tissue constituting the cyst most likely derived from the spinal cord itself. The most plausible explanation was proposed by Rhaney and Barclay in 1959. Inspection of a midline section (Fig. 3) through an early embryo reveals the relationships of the various structures. The close contact between the neuroenteric canal and the endoderm of the developing yolk sac would give rise to the formation of an enterogenous cyst if excalation of the two germ layers were not 100% complete.

The relationship of the associated congenital abnormalities to the age of presentation and the level of the lesion seems to be interesting and perhaps significant. If, as is most probable, the patients with more congenital abnormalities had presented for evaluation early in the process, this would leave the cases of late onset with fewer and relatively minor congenital abnormalities. Cervical lesions seem to be associated with more severe congenital abnormalities due to the fact that the rostral neuroenteric canal is in closer contact with the proximal foregut and the thoracic structures. This would explain any anomalies closely relating to the structures.

Acknowledgments

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References


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