

Case report: Intradural extramedullary bronchogenic cyst

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Cysts of the central nervous system (CNS) are usually benign and may be detected incidentally at autopsy. They become symptomatic either because of pressure, rupture, or secondary inflammation.^[1]

Spinal bronchogenic cysts are congenital in origin and are classified as enterogenous cysts.^[2] Enterogenous cysts have a lining epithelium similar to that of structures that originate from the primitive foregut, i.e., the esophagus, the respiratory tract, or the stomach. If the histological features of the lining epithelium resemble the respiratory epithelium, the cyst is termed a bronchogenic cyst.^[2] The present case report is of an enterogenous cyst (bronchogenic type) in the cervicodorsal region, with an intradural, extramedullary location.

Case Report

A 23-year-old male presented with complaints of gradually increasing numbness and weakness of both lower limbs for 22 days, along with one episode of incontinence of urine. On physical examination, he had a kyphoscoliotic deformity in the cervicodorsal region, which was confirmed with plain radiographs. Other vertebral anomalies could not be clearly discerned due to superimposition of shadows.

MRI of the cervicodorsal spine was then performed and showed a large, ovoid, thin-walled, homogenous lesion, extending from C5 to T1; it was hyperintense to cerebrospinal fluid (CSF) on T2W images [Figure 1] and isointense to the cord on the T1W images [Figure 2]. The lesion was intradural and extramedullary in location on the left posterior aspect of the spinal canal, compressing and displacing the cord anteriorly and to the right. On postcontrast images, there was no enhancement of the lesion [Figures 3 and 4]. Associated kyphoscoliosis was seen, with a hemivertebra at the T1 level [Figure 5]. In view of this combination of vertebral anomalies and a cystic lesion, the diagnosis of a neurenteric cyst was suggested.

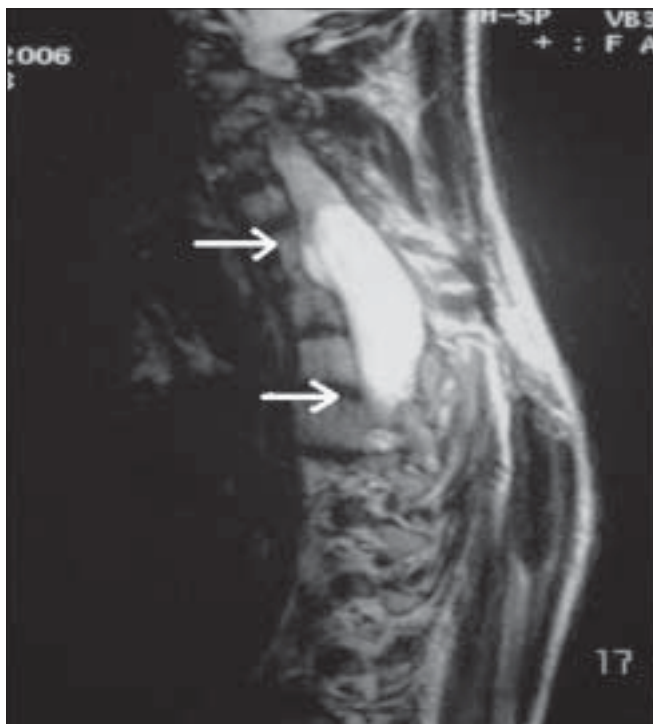


Figure 1: T2W sagittal MRI demonstrates a large oblong, well-defined homogeneous lesion (arrows) with a thin, barely perceptible wall extending from C5 to T1, hyperintense to CSF

The patient underwent surgery. Cyst fluid was aspirated and sent for examination. This proved to be negative for hydatid disease. The cyst wall was dissected and sent for histopathological examination. It showed features of a cystic structure lined by pseudostratified ciliated columnar epithelium, suggesting the diagnosis of a bronchogenic cyst [Figure 6].

The patient was discharged five days after surgery, by which time he had considerable improvement, with grade IV power in both lower limbs.

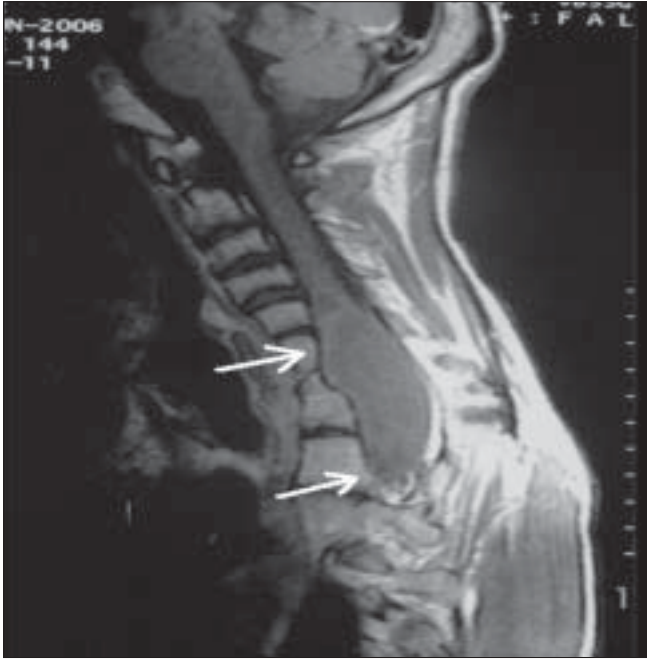


Figure 2: T1W sagittal MRI shows the lesion to be isointense (arrows)

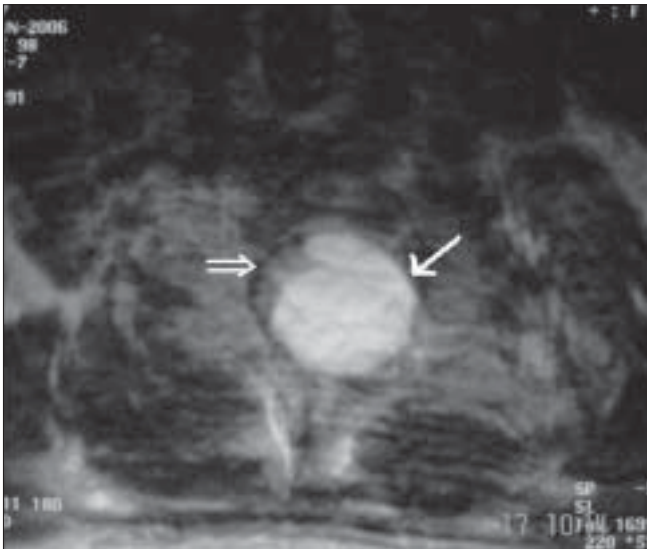


Figure 3: T2W axial MRI shows the lesion (arrow) compressing and displacing the cord towards the right anterior aspect (arrowhead)

Discussion

In 2005, Baumann *et al.* reported^[3] the first case of an intraspinal, intradural, extradurellary bronchogenic cyst located in the dorsolumbar region. Till date, only six cases of intradural, extradurellary, bronchogenic cysts have been reported.^[3]

Spinal neurenteric cysts are infrequently reported congenital abnormalities. They are believed to be derived from an abnormal connection between the primitive endoderm and ectoderm during the third week of life. Rauzzino *et al.*

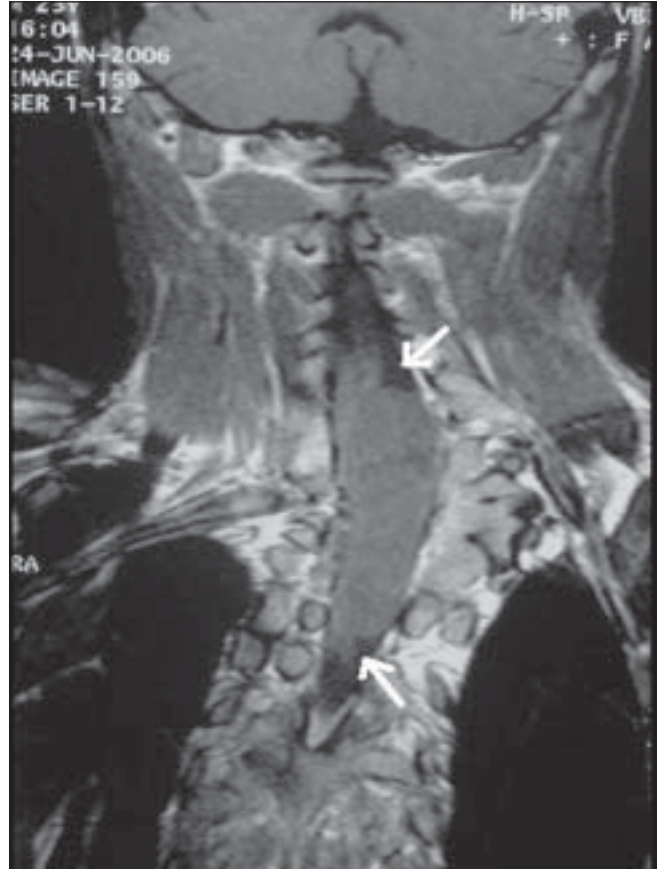


Figure 4: Post-contrast T1W coronal MRI shows that the lesion (arrows) displaces and compresses the cord with widening of the CSF space (arrowheads), suggesting an intradural extradurellary lesion without any enhancement

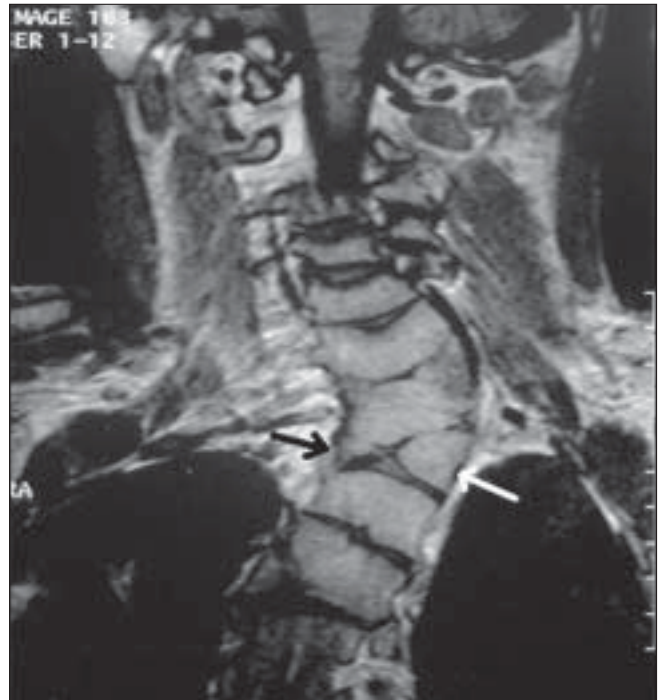


Figure 5: T1W coronal MRI shows a hemivertebra at T1 (arrow), the right fragment being smaller, leading to a scoliotic deformity

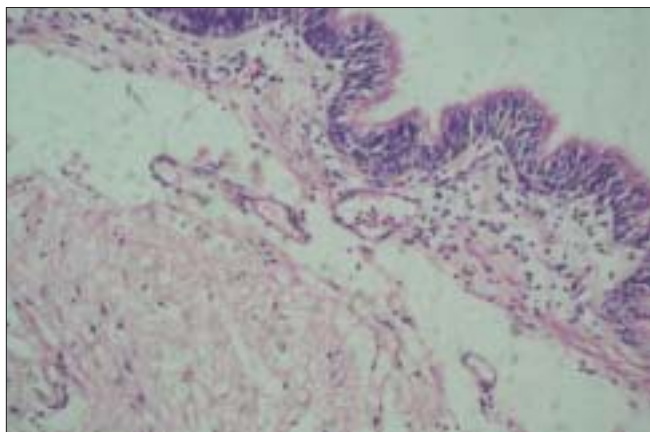


Figure 6: Photomicrograph of the cyst wall showing bronchogenic epithelium

reported a series of 13 patients of neurenteric cysts treated over a 50-year period.^[4] In their series, the cysts most commonly occurred as intradural, extramedullary masses in the thoracolumbar region, situated dorsal to the spinal cord, as was seen in our patient as well.

Intraspinal neurenteric cysts represent 0.3–0.5% of all spinal 'tumors.'^[4] However, they are not tumors (a fact that differentiates them from teratomas) but are more similar to hamartomas — displaced nests of endodermally-derived tissue.^[4] The findings may vary from a simple isolated intraspinal cyst with no other abnormalities to that of the so-called split notochord syndrome, as proposed by Bentley and Smith, in which there may be multiple visceral and vertebral anomalies.^[5]

Such cysts have been reported to occur at any site from the posterior fossa to the lumbosacral region, typically occurring in the dorsal spine or cervicodorsal junction, as the cephalic end of the notochord develops first and is the part most sensitive to disturbances during embryogenesis.^[2]

Embryological Development

Bremer postulated that congenital spinal anomalies, ranging from neurenteric cysts to diastematomyelia, could be explained by the persistence of a neurenteric canal.^[4] Bentley and Smith expanded Bremer's theory, postulating that the splitting of the notochord is the primary event.^[4] The subsequent deficiency in the overlying neural plate could allow for an endodermal diverticulum to herniate through the spinal column and make contact with the surface ectoderm.^[4] The persistence of the neurenteric connection that occurs in such a situation may be transient or permanent, and partial or complete. A number of scenarios may potentially result from this^[4]; for example:

1. If only the ventral portion remains, a duplication of the alimentary canal, without associated spinal abnormalities, can occur.

2. If only the terminal dorsal portion persists, a cutaneous abnormality, possibly associated with a sinus, might result.
3. If the mid-portion persists and the cleft in the vertebral body is incompletely obliterated, anterior spina bifida and, possibly, diastematomyelia may result.
4. If the cleft in the vertebral body is transient and incomplete, an intraspinal cyst can occur in the absence of permanent anterior vertebral anomalies.

Any combination of the aforementioned scenarios may occur.^[4]

Sundaram *et al.*^[1] reviewed 145 cases of cysts in the CNS of which five were neurenteric cysts. All five cases presented with cord compression. One of their cases was associated with hemivertebrae and all of them were intradural and extramedullary — two cervical and three thoracolumbar.

Wilkins and Odom^[6] classified these cysts into three groups according to the histological features. The simplest type of enterogenous cyst, group A, is lined by a single layer of cuboidal or columnar epithelial cells, with or without cilia, as seen in all of the four cases described by them. The more complex type, group B, has additional elements that are found along the course of the gastrointestinal tract or tracheobronchial tree, including mucous glands and smooth muscle, as was seen in our case. Group C has ependymal or glial tissue in addition to the features seen in group B. Immunohistochemical studies confirm the endodermal origin of these cysts.^[6,7]

Neurenteric cysts may also be associated with anterior or posterior spina bifida, widened vertebral bodies, fused vertebrae, hemivertebrae, and diastematomyelia.^[8] A review of 33 histologically verified enterogenous, intraspinal cysts by Agnoli *et al.*^[8] showed that 18 were located in the cervicodorsal spine; 80% were intradural, extramedullary cysts and 12% were intramedullary cysts.

Our case was an intradural, extramedullary bronchogenic cyst; it was dorsally located and was associated with vertebral anomalies.

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