

SPINAL ENTEROGENOUS CYSTS: A CLINICAL, MORPHOLOGICAL AND RADIOLOGICAL STUDY OF THREE CASES

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Enterogenous cysts of the central nervous system occur most frequently in the spinal canal, especially in the lower cervical and upper thoracic regions with intradural, extramedullary location.¹ Their intracranial occurrence is extremely rare.¹⁻⁴ They are benign epithelial lined cysts, with the lining resembling that of the alimentary canal.¹ They are often associated with developmental defects of the overlying skin and/or vertebral bodies.^{1,5} Occasionally they can have fistulous connection with similar mediastinal, thoracic or abdominal cysts, thus supporting an endodermal origin of these cysts.

Over the last two years we clinically observed three cases of this uncommon type of pathology and a complete study of them demonstrated important characteristics of their presentation. This made the observations of particular interest for the practice. The summary of data for the cases is shown in Table 1.

Case Reports

Case 1

This 16-year-old male patient was referred to the Department of Neurosurgery after being treated for low back pain radiating to the right lower limb, later associated with slow, progressive, bilateral lower limb weakness and incontinence. His complaints had started about three years ago with right leg pain, which persisted for one to two weeks and was relieved by medical treatment. About 10 days before admission, the low back pain with radiation to the right leg got seriously worse. Admitted to another hospital, his condition rapidly deteriorated, with numbness and severe weakness of the same limb. On admission to our department, his movements and sensation for the lower

TABLE 1. Summary of data for the cases.

Case	Age/sex	Level of lesion	Duration of symptoms	First symptoms
1	16 years/ male	T12-L1	3 years	Back pain
2	16 years/ male	L2-L3	4 months	Back pain, lower limb weakness
3	2 months/ male	Lower end at T8	Since birth	Spinal deformity, lower limb weakness, dyspnea

limbs were severely affected and he noticed urinary incontinence.

On examination, he was found to have paraplegia and a sensory level of affection from L3 dermatome caudally. There was complete loss of joint position and vibration sense in the distal parts of the lower limbs. Deep tendon reflexes were not present and plantar responses were also absent. The patient was admitted with a Foley catheter.

His hemogram and routine plasma biochemistry were within normal limits. Emergency myelography and postmyelography CT scanning revealed an extradural mass lesion, which was extending through the right neural recess at T12-L1. Asymmetry in the shape of the spinal canal indicated a slow-growing lesion. On the MRI study, an extradural (not affecting conus) cystic mass lesion was observed, which was not enhanced with gadolinium. The cystic nature of the lesion was obvious because of the fluid level detected inside (Figure 1).

Surgery was performed on the day of admission. Laminectomy of T12 and L1 with partial laminectomy of T11 and L2 was performed. An extradural mass lesion with posterolateral right location was found at the level of T12 and L1, extending to the intervertebral foramen. The lesion was cystic with a thick wall and was strongly adherent to the dura. After evacuation of the thick, gray-to-brown fluid contents, the cyst wall collapsed and it was dissected from the dura, followed by separation from the dural root sleeve entering the intervertebral foramen. On exploration, the extravertebral part of the cyst was not

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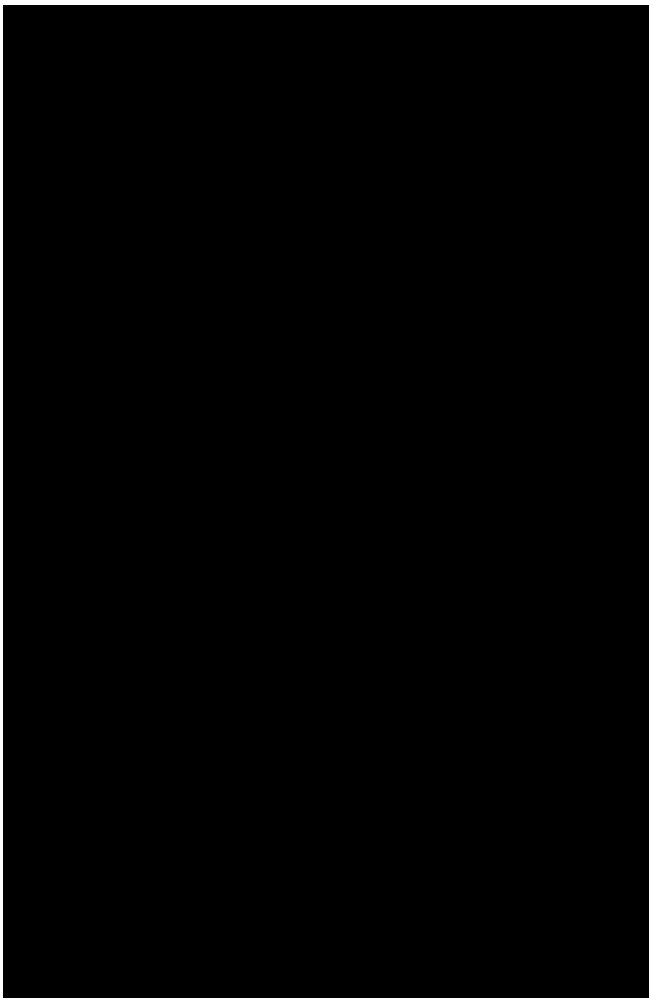


FIGURE 1. T2W sagittal imaging demonstrates sausage-like, well-capsulated extradural mass at Th12 and L1 level. The upper part of cystic fluid is hypointense; the dependent part shows high signal intensity, similar to CSF.

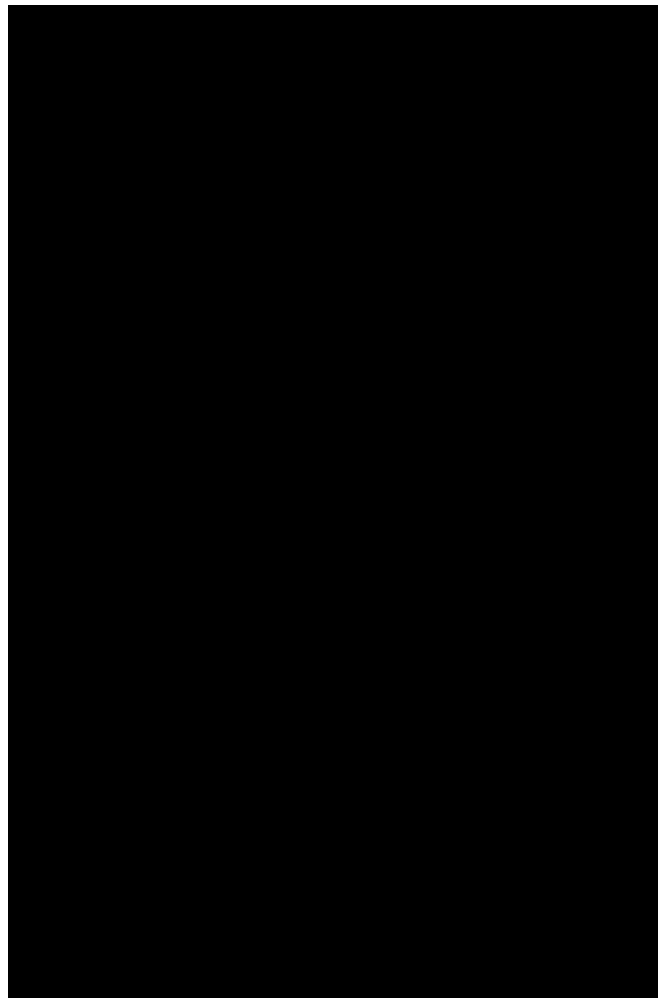


FIGURE 2A. Partial outline of an intradural mass at L3, widening of interpediculate distance of L1 and L2 vertebrae and displaced cauda equina elements.

found. No penetration through the dura was observed.

Postoperatively the patient experienced partial recovery. Weak movements of the toes appeared with partial recovery of pain sensation in the left lower limb. Unfortunately, the incontinence was still present until the 20th postoperative day, when the patient was transferred to another hospital for physical treatment and rehabilitation. Two years later he is able to walk with braces for the knee and ankle joints on the right side and is continent for urine and feces.

Case 2

This 16-year-old male was admitted to the Neurosurgical Department with progressive weakness of the right lower limb for four months, associated with low back pain radiating to the right leg. On examination there was

detectable wasting of the quadriceps muscle on the right side, with weak plantar and dorsal flexion of the foot. The right plantar reflex was weak and equivocal in type. There were no sphincteric disturbances. The straight leg raising test on the right was doubtfully limited.

Hemogram and serum biochemistry did not reveal any abnormality. Emergency myelography was carried out and this demonstrated incomplete block, outlining an ovoid intradural mass at L1, L2 and L3 vertebrae (Figure 2A). The extramedullary and cystic nature of the mass were confirmed on MRI: the lesion appeared slightly hyperintense to CSF on T1W images (Figure 2B) and showed high signal intensity on T2W sequences. CSF protein below the lesion was 905 mg/L.

On the day after admission, laminectomy of L1 and L2 was done. After opening the dura along the midline, a rounded lesion was found, involving cauda equina and

displacing the conus medullaris tip upwards. It was reaching caudally to the upper border of the L3 vertebral lamina. The lesion was cystic and through a puncture its turbid and viscous content was evacuated. The collapsed wall was then dissected from the surrounding roots and removed completely. The operative wound was closed in the usual way.

There were no postoperative complications and the motor power of the right lower limb improved over the following weeks, when the patient started to do physical treatment and rehabilitation.

Case 3

A male infant, aged two months at admission, was observed to have episodes of respiratory distress and fever after birth. The movements of his legs were apparently weak. For these findings the child was referred to the

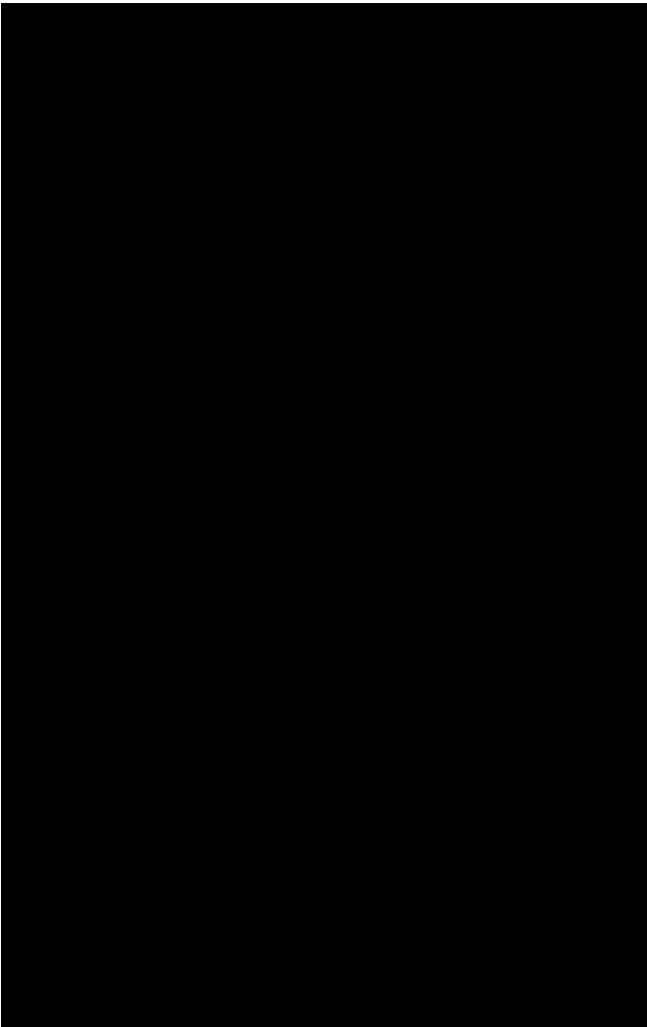


FIGURE 2B. Sagittal T1W MRI. Note widening of A-P diameter of the spinal canal at L1 and L2.
neurosurgical department.

On examination the baby was tachypneic, distressed, his circulation was stable, blood gases were showing compensation and he was on nasogastric feeding. General examination of all systems did not reveal major abnormalities, except minimal basal crepitations on chest auscultation, compensated heart, pulse 120/min and respiratory rate 50/min. The neurological examination revealed decreased spontaneous and provoked movement activity, spasticity and deep tendon hyperreflexia of the lower limbs. A hard subcutaneous swelling along the midline was palpated at a level from C7 caudally to the lower third of the thoracic spine. Hemogram, serum biochemistry and blood gas analysis were within normal limits.

Chest x-ray demonstrated a posterior mediastinal mass and the vertebral x-rays detected several malformed vertebrae in the upper and middle thoracic regions. The patient's condition was reviewed by a chest surgeon for the posterior mediastinal mass. MRI revealed a tandem intraspinal-posterior mediastinal cystic mass with somewhat dorsal extraspinous extension (Figure 3A). The interconnection appeared as a midline sinus tract extending through a bony spinal defect. The deficient vertebrae, as well as the lack of communication between the mass and CSF spaces, was best disclosed on myelography (Figure 3B). There was sufficient evidence to suggest an enterogenous cyst rather than an anterior meningocele.

Surgery was considered to be the only option to control the progressive dyspnea and arrest the neurological deterioration. It was performed in two stages, by two teams, six days after admission. After a left thoracotomy, the lesion located in the posterior mediastinum was removed. Its nature was cystic, full of dark, turbid, mucous fluid, and its isthmus part was ligated deep inside a canal-like defect between several malformative vertebral bodies. Through a separate approach, an upper and midthoracic laminectomy was done and an enlarged fusiform dural sac was exposed. A midline incision of dura revealed a cystic lesion integrated with the spinal cord in one portion and extending ventrally through the defect between the vertebral bodies. The cyst was evacuated and it contained the same type of fluid as the mediastinal part. The wall was dissected from the spinal canal and resected. A small part, integrated with the continuity of the spinal cord (there was no detectable border between the cyst and the cord under the surgical microscope), was left in place and the internal layer of the cyst coagulated and excised. The postoperative period was uneventful for the first two days. After that, the patient developed obstruction of the right principal bronchus, later on complicated with a bronchopleural fistula. The patient's condition after the ninth postoperative day deteriorated severely and despite the rethoracotomy, the

patient expired because of cardiac arrest and failed resuscitation.

Pathological Findings

The tissue removed at surgery was fixed in 10% buffered formalin. Sections were routinely processed and embedded in paraffin. Five micron sections were cut and stained with hematoxylin and eosin (H&E), periodic acid Schiff stain (PAS) and mucicarmine stains.

The histological findings were similar in all three cases. The cysts showed a fibrous wall with a single layer of mucin-secreting columnar epithelial lining. There were glandular structures resembling gastric glands. Some goblet cells were seen and a few cilia. The cyst wall contained two layers of muscle coat. No ependymal or glial tissues were seen (Figure 4). The epithelial cells were

positive for PAS and mucicarmine. The diagnosis was enterogenous cyst (Group B).

Discussion

Enterogenous cysts of the central nervous system, also called neuroenteric cysts⁶ or gastrocytomas,⁷ were first reported in 1934 by Pussep,⁸ who treated a case of intestinoma of the cervical spinal cord. These cysts within the spinal cord are not common, with only 51 cases having been reported with histological confirmation.^{1,3,9} They generally arise from the lower cervical to the upper thoracic region of the spinal cord, causing symptoms of spinal compression.^{1,5,6,10} Most of these cysts are found with intradural extramedullary location and intramedullary lesions are very rare.^{1,7,11} About half of these cases are associated with spinal deformities such as spina bifida,

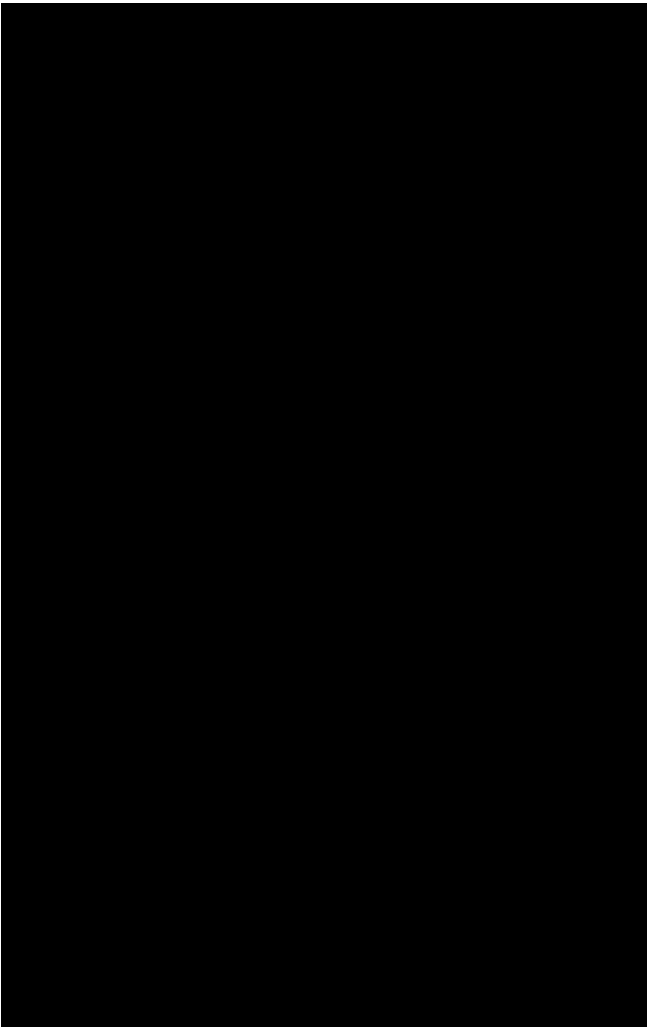


FIGURE 3A. T1W sagittal MRI. The intercommunication shows a flow void phenomenon, probably because of close proximity of the mediastinal portion of the cyst to the heart and great vessels.

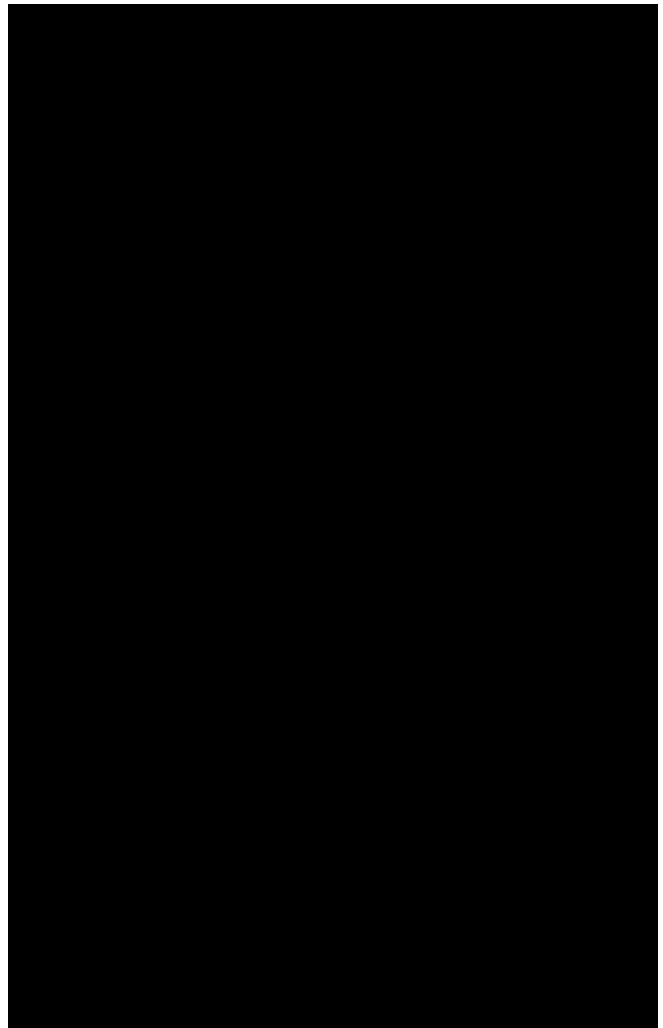


FIGURE 3B. Myelographic block and severe spinal anomalies in middle and upper thoracic region.

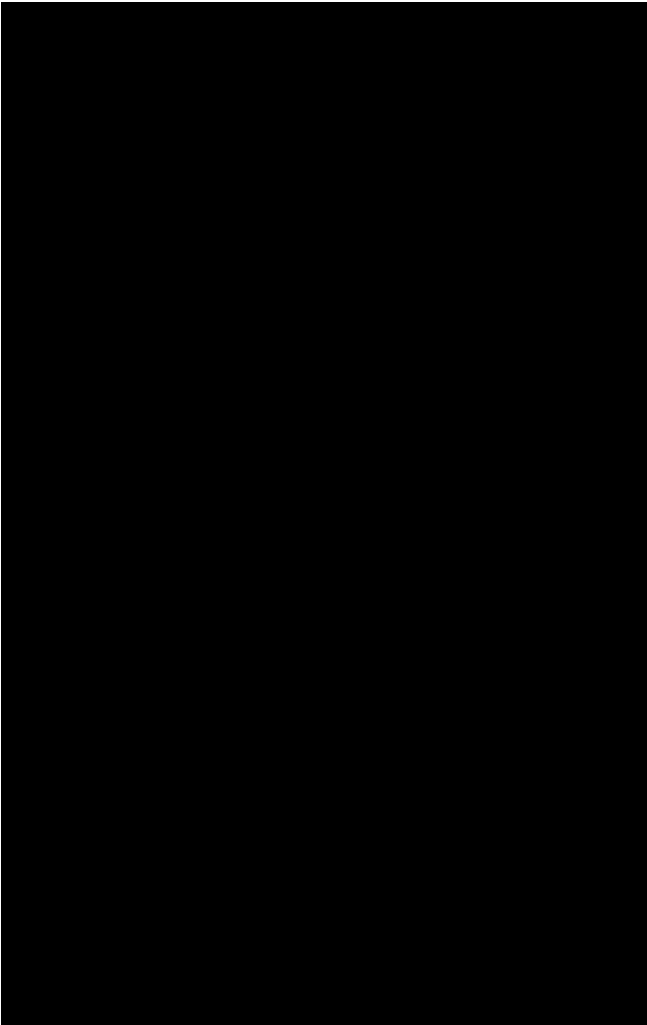


FIGURE 4. Photomicrograph showing the cyst wall lining by mucin secreting columnar epithelium. Note the glands in the mucosa and the underlying muscle coat in two layers (H&E, 80x).

hemivertebra and vertebral fusion.^{1,5,10}

Enterogenous cysts have been classified into three groups, according to histological features.¹² Group A is the simplest type, lined by a single layer of cuboidal or columnar epithelial cells with or without cilia. Group B cysts include more complex elements of the gastrointestinal tract or tracheobronchial tree, including mucus glands and smooth muscle in their wall. Group C cysts have ependymal or glial tissue in addition to the elements seen in group B cysts. Most enterogenous cysts belong to group A. However, all our cases had features of group B.

A variety of hypotheses have been suggested regarding the pathogenesis of intraspinal enterogenous cysts, but none are firmly established.¹³⁻¹⁶ They are believed to originate from embryonal dysgenesis.¹⁷⁻¹⁹ During normal development, the neuroenteric canal closes and the notochord separates from the primitive gut in the third

week of embryonic life. It is proposed that during the same period, a transient adhesion occurs between the neural ectoderm and endoderm, or a communication develops along the neuroenteric canal. When such a developmental abnormality persists because of the incomplete separation at this adherence or remnant canal, the cyst forms.^{3,13-16}

MacKenzie and Gilbert²⁰ have demonstrated morphological and immunohistochemical similarities between colloid cysts of the third ventricle and spinal enterogenous cysts, suggesting that these lesions are all derived from primitive gut endoderm.

Our cases illustrate three possible variants of location of these lesions. The first type, the intradural one, had the typical clinical course and radiological presentation of a benign intradural extramedullary mass lesion. The methods of treatment with corresponding surgical techniques and the expected outcome are similar for this type of lesion. The second type of lesion, extradural and intraforaminal, had more subtle initial onset and rapid notorious progression in the end. The excision, however, rendered some recovery despite the presence of apparently irreversible damage. And the last type—the hourglass lesion—presented with severe affections of the neural and mediastinal structures. The spinal cord can be seriously malformed, and complete resection can be difficult or impossible to accomplish. According to the area involved, the teamwork with the thoracic and abdominal surgeons is essential.

The choice of surgical technique varies according to the type of cyst and its exact anatomical location. MRI appears to be the best modality for definite imaging of spinal enterogenous cysts at present. However, for better visualization of the associated bony dysraphic defects, and certain anatomical details, some patients may well benefit from the plain x-ray images (indispensable), myelography or CT scan.²¹

Spinal enterogenous cysts are benign lesions with insidious progression, and their early preplanned surgical removal should be the goal of treatment, as the very advanced stage of manifestation can be critical for neural recovery and clinical outcome. In a population area with a high incidence of congenital malformations, cystic inter- and paraspinal mass lesions without significant surrounding inflammatory reaction should arouse the suspicion of an enterogenous cyst.

References

1. Russell DS, Rubinstein LJ. Pathology of tumors of the nervous system, 5th ed. London: Edward Arnold, 1989;704-6,721-25.
2. Umezy H, Aiba T, Unakami M. Enterogenous cyst of the cerebellopontine angle cistern. Case report. *Neurosurg* 1991;28:462-6.
3. Ito S, Fujiwara S, Mizoi K, Namiki T, Yoshimoto T. Enterogenous cyst at the cerebellopontine angle. Case report. *Surg Neurol* 1992;37:366-70.

4. Mehta VS, Chaudhury C, Bhatia R. Neuroenteric cyst of the cerebellum. *Postgrad Med J* 1984;60:287-9.
5. Leech RW, Olafson RA. Epithelial cysts of the neuraxis. *Arch Pathol Lab Med* 1977;101:196-202.
6. Holcomb GW Jr, Matson DD. Thoracic neuroenteric cyst. *Surgery* 1954;35:115-21.
7. Knight G, Griffiths T, Williams I. Gastrocytoma of the spinal cord. *Br J Surg* 1955;42:635-8.
8. Pussep M. Variété rare de teratome sous-dural de la région cervicale (intestinoma). Quadruplegie. Extirpation. Guérison complète. *Rev Neurol* 1934;2:879-86.
9. Itakura T, Kusamoto S, Uematsu Y, Imai H, Nishiguchi T, Hayashi S, Komai N. Enterogenous cyst of cervical spinal cord in a child—case report. *Neurol Med Chir* 1986;26:49-53.
10. Agnoli AL, Laun A, Shanmayer R. Enterogenous intraspinal cysts. *J Neurosurg* 1984;61:834-40.
11. Silvernail WI Jr, Brown R. Intramedullary enterogenous cyst. *J Neurosurg* 1972;36:235-8.
12. Wilkins RH, Odem GL. Spinal intradural cysts. In: Vinken PJ, Bruyn GN (Eds). *Handbook of Clinical Neurology* Publishing Co. 1976;Vol 20:55-102.
13. Bremer JL. Dorsal intestinal fistula: accessory neuroenteric canal; diastematomyelia. *Arch Pathol* 1952;54:132-8.
14. McLetchie NGB, Purves KJ, Saunders RL de Ch. The genesis of gastric and certain intestinal diverticula and enterogenous cyst. *Surg Gynecol Obstet* 1954;99:135-41.
15. Prop N, Frensdorf EL, Van De Stadt FR. A postvertebral endodermal cyst associated with axial deformities; a case showing the "endodermal-ectodermal adhesion syndrome". *Paediatrics* 1967;39:555-62.
16. Small JM. Pre-axial enterogenous cyst (abstract). *J Neurol Neurosurg Psych* 1962;25:184.
17. Dorsey JF, Tabrisky J. Intraspinal and mediastinal foregut cyst compressing the spinal cord. Report of a case. *J Neurosurg* 1966;24:562-7.
18. Harriman DGT. An intraspinal enterogenous cyst. *J Pathol Bacteriol* 1958;75:413-9.
19. Rhaney K, Barclay GPT. Enterogenous cyst and congenital diverticula of the alimentary canal with abnormalities of the vertebral column and spinal cord. *J Pathol Bacteriol* 1959;77:457-71.
20. Mackenzie IRA, Gilbert JJ. Cysts of the neuraxis of endodermal origin. *J Neurol Neurosurg Psych* 1991;54:572-5.
21. Patrick D, Barnes PD. Developmental abnormalities of the spine and spinal neuroaxis. In: *MRI in Pediatric Neuroradiology*. Wolpert SM, Barnes PD, Eds. St. Louis: Mosby Year Book Publ., 1992:331-77.