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# SPINAL INTRADURAL EXTRAMEDULLARY ENTEROGENOUS CYSTS

## Report of two cases and review of literature

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RÉSUMÉ

**Kystes entérogènes intra-duraux et extra-médullaires. Deux cas et revue de la littérature.**

Les auteurs décrivent le cas de deux patients porteurs de kystes intra-duraux et extra-médullaires situés dans les régions cervicale et dorsale supérieure du canal rachidien. Tous deux souffraient de myélo-radiculopathie lentement progressive due à des kystes producteurs de mycine et tapissés d'épithélium. A l'examen histologique, les deux lésions ont été diagnostiquées comme étant des kystes neurentériques d'origine endodermique. L'examen radiologique a fait appel à la myélographie simple puis associée à la tomodensitométrie (TDM) et à l'imagerie par résonance magnétique (IRM). Chez ces deux patients, la guérison a été obtenue par exérèse chirurgicale totale. Les signes cliniques, les caractéristiques histologiques, les résultats de l'imagerie et le traitement chirurgical de cette rare lésion sont soulignés.

*Mots clés : Examen radiologique avec myélographie. Imagerie par résonance magnétique. Kyste entérogène. Kyste neurentérique.*

SUMMARY

**Spinal intradural extramedullary enterogenous cysts. Report of two cases and review of literature.**

Two patients with intradural extramedullary cysts of the spinal canal are described. Both presented with slowly progressive mylo-radiculopathy caused by mucin-producing epithelial-lined cysts in cervical and upper thoracic region. Histologically, both lesions were considered to be neurenteric cysts with an endodermal origin. Radiographic diagnosis was made by a combination of myelography, computed tomographic scan with mylography (CTM), and magnetic resonance imaging (MRI). Complete surgical resection was curative in both cases. Clinical presentation, histological characteristics, imaging findings and surgical management of this rare lesion are highlighted.

*Key Words :* Computed tomographic scan with myelography. Magnetic resonance imaging. Spinal intradural cyst. Neuroepithelial cyst. Enterogenous Cyst. Neurenteric cyst.

## INTRODUCTION

Intradural extramedullary (IDEM) cysts within the spinal canal rank among the most unusual causes of symptomatic spinal cord compression [11]. Thus, Lombardi and Morello [9] reported only one intraspinal cyst among 290 symptomatic spinal lesions. Benign cystic lesions in the spinal IDEM space are exceedingly rare [17] and include : arachnoid cysts, ependymal cysts, enterogenous or neurenteric cysts and teratogenous cysts [11]. The rarest of the lesions is the enterogenous cyst. It is defined as a "cyst lined by mucin-secreting epithelium resembling that of the gastrointestinal tract" [18]. The review of world literature by Agnoli *et al.* [1] lists 33 reported cases, of which only 25 were in IDEM position; and few included CT-MRI findings. Since then, Chavda *et al.* [4] have reviewed seven intraspinal enterogenous cysts. Six further cases were reported by Russel and Rubinstein [14]; of these, four were intramedullary and only two in IDEM location.

Spinal enterogenous cysts have a congenital origin [11], and are presumed to be derived from the endoderm of the developing gastrointestinal tract [3] or, rarely, the respiratory system [17]. These cysts are mostly found in the IDEM compartment of the cervical and upper thoracic spine [3, 8, 11]. Because this is a benign and potentially curable condition, it is important to recognize this entity and make an early diagnosis before irreversible neurological deficits and myelomalacia by chronic pressure on the spinal cord have occurred.

## CASE REPORTS

### CASE 1

A 63-year-old man had numbness around upper chest punctuated by intermittent right-sided pain in upper intercostal distribution for many years. Intermittent clumsiness, weakness and stiffness of lower extremities with complaints of sexual and urinary dys-

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function dated back 13 years. Six weeks prior to admission, leg weakness prevented him from walking without help. Progressively increasing paraparesis followed. Four days before admission he developed pins and needles in both legs and trunk. Inability to stand and pass urine led to hospitalization four days later. Neurological examination on admission revealed spastic paraparesis with hyperreflexia and bilaterally upgoing plantars. Power was 1/5 in the right and 2/5 in the left leg. Hypoaesthesia with a sensory level at about nipples was noticed. Position and vibration sense were lost in the right leg. Plain x-ray films of dorsal spine showed an anterior bony defect in the body of D3-D4. Myelography demonstrated a complete block at D2-D3. It also outlined a large intradural mass severely deforming and compressing the cord to the left *(fig. 1)*. Post-myelogram CT cuts at the level of block showed a large mass almost totally filling up the spinal canal and pushing the thinned out cord to the left side *(fig. 2a* and *b)*. Pre-operative diagnosis of a schwannoma was made and patient underwent C7-T3 laminectomy. When the dura was opened, a tense, large oval cyst with whitish opaque wall was found in the subarachnoid space. It filled the whole spinal canal and compressed the flattened and indented cord into left lateral recess. There were minimal adhesions between the cyst wall and surrounding structures, these were dissected and cyst totally excised. Cyst contents were turbid and milky-mucinous. Postoperatively the neurological recovery was rapid and complete. One year later, the patient is ambulatory and asymptomatic.

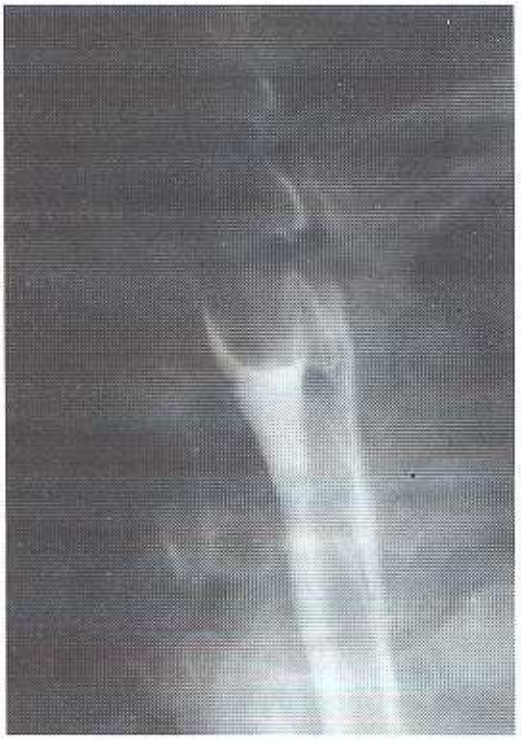
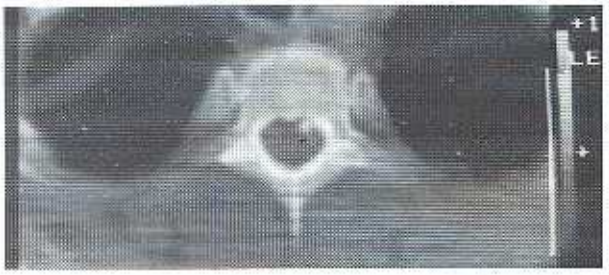


FIG. 1 (Case 1). – Myelogram revealed complete block to the flow of contrast at D2-D3 with lateral deviation of the spinal cord shadows as well as the rounded inferior margin of the lesion with widened subarachnoid cap.

FIG. 1. (Cas 1). *Ce myélogramme révèle un blocage complet du moyen de contraste en D2-D3 avec déviation latérale de la moelle épinière, ainsi qu'un aspect arrondi de la limite inférieure de la lésion avec élargissement de l'espace subarachnoïdien.*



a

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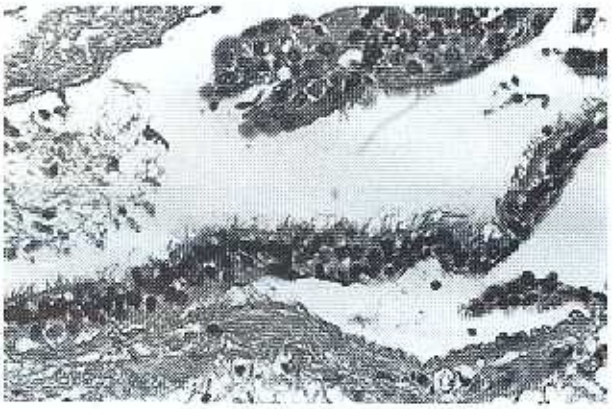
FIG. 2 (Case 1). – (a) CT scan following myelography (CTM) at the level of block demonstrates the lesion and the laterally compressed spinal cord (arrows); (b) CTM just below the lesion showing the filling defect of the laterally displaced spinal cord (arrow) and contrast in the widened subarachnoid cap at the lower pole of lesion.

FIG. 2. – (Cas 1). *(a) Tomodensitométrie (TDM) après myélographie au niveau du blocage, mettant en évidence la lésion et la moelle comprimée latéralement (flèches). (b) TDM juste au-dessous de la lésion, montrant le défaut de remplissage dû à la moelle épinière déplacée latéralement (flèche) et le moyen de contraste dans l'espace subarachnoïdien, au pôle inférieur de la lésion.*

## PATHOLOGICAL EXAMINATION

Gross specimen showed a cystic structure that measured 2 x 1 cm with irregular folding. When opened whitish wall was 2 mm in thickness. Histological examination of the cyst wall revealed a lining of ciliated and non-ciliated columnar epithelium, at places

showing stratification *(fig. 3a* and *b)*. This epithelial layer showed goblet cell formation and presence of mucous. The material was positive to alcian blue and periodic acid-schiff (PAS) staining. The epithelium rested on fibrous tissue in which unstriped muscle fibres were seen.



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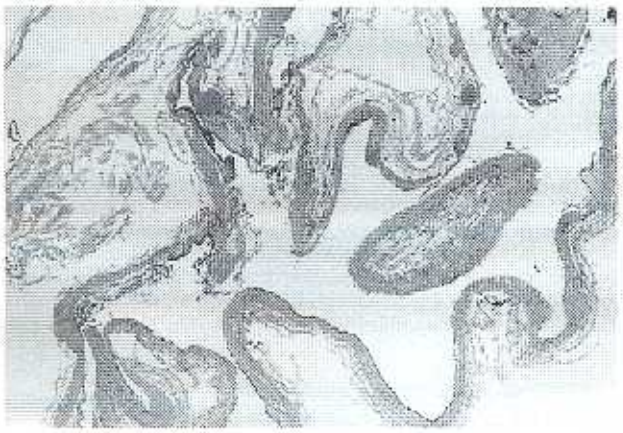

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FIG. 3 (Case 1). - (a) Photomicrograph demonstrating ciliated and non-ciliated columnar epithelium, at places showing stratification, resting on vascular collagenous capsule. The epithelial cells show goblet cell formation and the presence of mucus (H&E x 300); (b) cyst wall : epithelial layer resting upon vascular connective tissue (H&E x 50).

FIG. 3. - (Cas 1). *(a) Photomicrographie montrant un épithélium cylindrique cilié ou non cilié et parfois stratifié, reposant sur une capsule vasculaire collagénique. Les cellules épithéliales montrent des cellules caliciformes en formation et la présence de mucus (H&E x 300); (b) Paroi du kyste : revêtement épithélial reposant sur un tissu conjonctif vascularisé (H&E x 50).*

Case 2

A 38-year-old man presented with an 18-month history of increasing left sided neck pain. Prior to this he had intermittent symptoms of clumsiness of upper and lower extremities and pins and needles in left arm and leg for many years. He was known to have an asymptomatic congenital scoliosis due to hemivertebrae in the cervicothoracic spine. On examination he had spasticity, hyperreflexia and minimal weakness in upper and lower limbs but no objective sensory loss. Radiographs of the cervical spine showed fusion of the bodies of C2 and C3 with hemivertebra at C6 and widening of the spinal canal. CT-myelography demonstrated a partial block at C2 with an IDEM mass. MRI *(fig. 4)* revealed a cystic lesion, with signal characteristics similar to CSF, in front of and to the left of the cervical cord at C2.

The lesion was approached via a left C2 hemilaminectomy. On opening the dura, a tough opalescent cystic tumour was seen beneath the ligamentum denticulatum and lying amongst the ventral nerve roots of C2 and C3. The dorsal nerve roots at these levels were found to be stuck together by fibrosis and were dissected free. The cyst was removed piecemeal and did not appear to be attached to cord or dura. There were no problems post-operatively. Five months later, the patient was asymptomatic. Histological examination *(fig. 5)* of the specimen revealed a cyst with a lining of widely different appearance, including areas lined by respiratory type pseudostratified ciliated epithelium, flattened cuboidal cells, and by columnar, ciliated, mucin-producing epithelium *(fig. 5)*. Goblet cells that showed positive staining with alcian blue and PAS were scattered in the epithelial lining. A number of subepithelial mucous glands were also present. Immunocytochemistry showed strong positive staining of the mucous producing and ciliated areas of epithelium with antibody to cytokeratin (CAM 5.2). Diagnosis of a benign epithelial cysts was made. Histological features were suggestive of an endodermal origin.

## DISCUSSION

In origin, spinal intradural cysts can be epithelial, such as endodermal (neurenteric) or neuroepithelial (ependymal) cysts; mesenchymal, such as arachnoid cysts; or mixed, such as teratomatous cysts. In the present cases, the histological appearances ruled out arachnoid cysts because they are formed of connective tissue and possess no epithelial lining [3, 11]. Diagnosis of teratomatous cyst was unlikely because the cysts in our cases do not possess elements of all three germ cell layers. The presence of gastrointestinal/respiratory type epithelium, goblet cells, positive staining of the lining cells with PAS, alcian blue, cytokeratin, vertebral anomalies and classical cervical/upper thoracic location of cysts in our cases supports an endodermal rather than an ependymal origin. Therefore, these two patients with mucin-producing epithelial lined cysts in the spinal IDEM space are most probably neurenteric cysts.

Origin and pathogenesis of enterogenous cysts, still a subject of debate is probably best explained by the

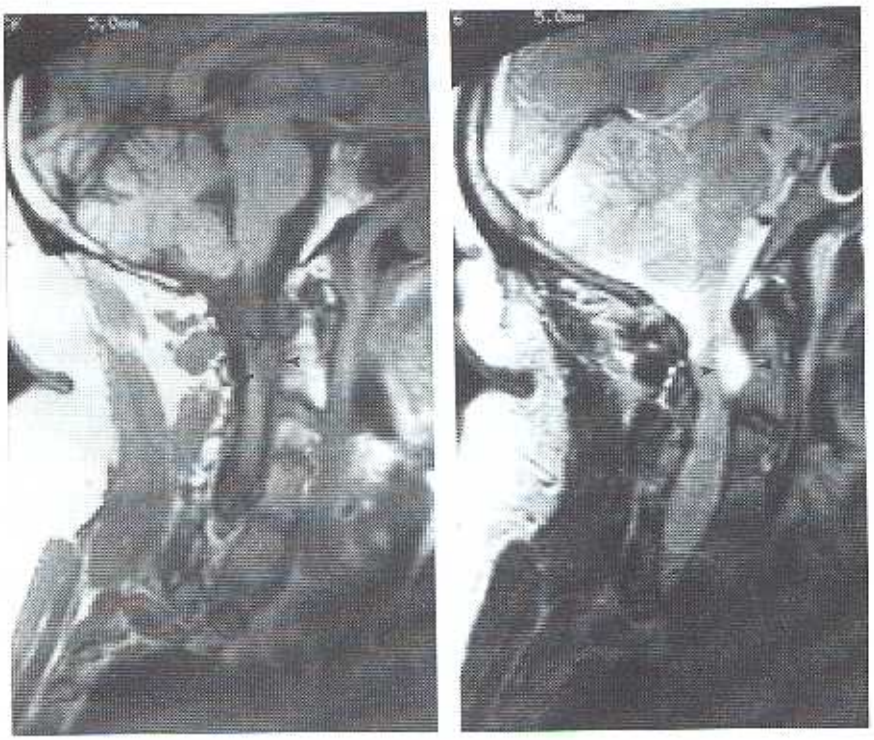

FIG. 4. – (Case 2). T1 and T2 weighted MRI scans (left and right respectively) reveal a discreet cystic lesion (arrows) compressing the cord at C2. Lesion is hypointense on T1 and hyperintense on T2 weighted images.

FIG. 4. – (Cas 2). *Sections d'IRM pondérées en T1 et T2 (respectivement à droite et à gauche). Elles révèlent une lésion kystique discrète (flèches) comprimant la moelle en C2. La lésion est hypointense en T1 et hyperintense en T2.*

split notochord theory proposed by Bentley and Smith [2]. An anomalous protrusion of the primitive foregut through a split notochord against the primitive neuroectoderm in the third week of embryonic life is thought to lead to the development of such cysts [6, 16].

The most common clinical presentation in patients with symptomatic spinal intradural cysts is an insidious onset of a slowly progressive myelopathy with or without radicular symptoms [11]. A fluctuating clinical course punctuated with remissions and exacerbations with a slow step-ladder-like progression of neurological deficits is not unusual [1, 11, 13] as happened in our patients. This presentation has sometimes resulted in patients being erroneously diagnosed as having multiple sclerosis or some other demyelinating disease [1, 11, 12]. A review of reported cases in the literature [1] shows that 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 [1]. Vertebral anomalies were present in about half of the reported cases of enterogenous intraspinal cysts [1].

MRI and CT documentation has been lacking in majority of the reported enterogenous intraspinal cysts [1, 17]. More recently, some authors have described MR and CT myelography (CTM) findings of the cystic spinal intradural lesions along with the comparative accuracy and sensitivity of these diagnostic modalities [11, 15]. CTM locates the enterogenous cyst as an intradural filling defect that displaces the spinal cord but fails to demonstrate its cystic nature in a significant number of cases [11, 15], as happened in our Case 1. Unlike the majority of the spinal arachnoid cysts, enterogenous cysts do not communicate with the spinal subarachnoid space, and therefore, the contrast fails to enter the cyst on delayed postmyelogram CT scan. MRI, because of signal characteristics, accurately identifies the lesion pre-operatively as an intradural cyst in all patients [1, 15], as it did in our Case 2.

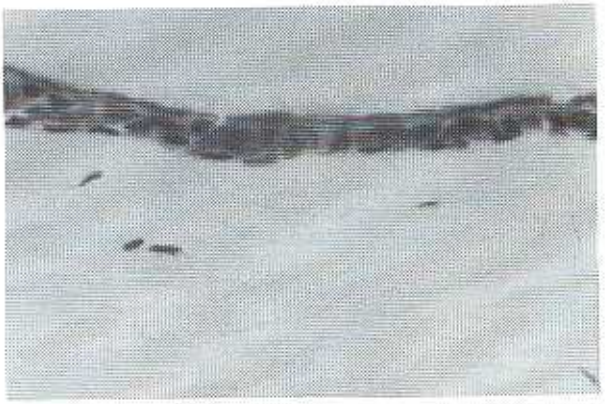

FIG. 5 (Case 2). - Photomicrograph of cyst lining demonstrating ciliated cuboidal epithelium resting over loose connective tissue (H&E x 50).

FIG. 5. – (Cas 2). *Photomicrographie du revêtement du kyste, montrant un épithélium cuboïde cilié reposant sur un tissu conjonctif lâche (H&E x 50).*

Patients with symptomatic spinal intradural cysts should be treated surgically before irreversible neurological deficits occur. Majority of these cysts enlarge and become progressively more symptomatic [11]. Large cysts that significantly deform and compress the cord may place the patient at higher risk of acute neurological deterioration in the event of trauma [11, 10]. The surgical procedure of choice is complete excision of the cyst as was possible in both of our cases. Ventrically located cysts, particularly when they are tenaciously adherent to the cord, are frequently not amenable to complete resection without risking injury to the cord and its blood supply [11]. In the se cases partial resection or wide fenestration is an acceptable alternative and is generally curative [5, 11]. Recovery is usually complete and recurrence is not the rule even after partial resection [5]. However, long-standing neurological deficits, chronic paresis and thinning of the cord are poor prognostic signs for recovery of function [7].

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