Intradural neurenteric cysts
Case report

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Abstract
Spinal neurenteric cysts are rare lesions—reports are just on isolated cases. According to different bibliographic quotes, they account for 0.3% to 1.3% of all spinal tumors. They usually affect the spinal canal, especially at the levels of the low cervical and the high thoracic spine. Approximately 90% of these cysts are located in the intradural/intramedullary compartment, whereas the remaining 10% occupies the intradural/intramedullary and the extradural compartments in different proportions. Intraneural neurenteric cysts should undergo surgical treatment by as radical a resection as possible. Results are favorable and recurrence rates after partial resection are low.

Key words: Neurenteric cyst; spinal tumor.
Level of evidence: IV

Quiste neuroentérico intradural Reporte de un caso

Resumen
El quiste neuroentérico espinal es una lesión de rara presentación, sólo existen reportes de casos aislados. Representa del 0,3% al 1,3% de los tumores espinales según las distintas citas bibliográficas. Suele afectar el canal espinal, especialmente a nivel cervical bajo y torácico superior. Aproximadamente el 90% de estos quistes se localiza en el espacio intradural/extramedular, mientras que el 10% restante se divide entre la localización intradural/intramedular o extradural. El tratamiento de los quistes neuroentéricos intraespinales es quirúrgico, mediante la resección lo más radical posible. Los resultados son favorables y las tasas de recurrencia después de la resección parcial son bajas.

Palabras clave: Quiste neuroentérico; tumor espinal.
Nivel de Evidencia: IV

Introduction

Spine neurenterics cysts are rare injuries and only isolated cases have been reported. Intraspinal cysts result from the incomplete division between the endoderm and notochordal tissues during early embryological development. These lesions were first described by Kubie and Fulton, in 1928, as teratoma cysts, and then, in 1934, by Puusepp, as intestinomas,[1,2] but finally Holcomb and Matson defined them definitely as neurenteric cysts in 1954.[3]

According to different bibliographic quotes, neurenteric cysts account for 0.3% to 1.3% of all spinal tumors. [4,5]

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They usually affect the spinal canal, especially low cervical and high thoracic levels.[6] Approximately 90% of them are located in the intradural/extramedullary compartment, whereas the remaining 10% occupies the intradural/intramedullary and the extradural compartments in different proportions, [4,5] although there are also reports on other locations in the central nervous system.[7-10]

Intraspinal neurenteric cysts should undergo surgical treatment by radical resection. [4.11.12] Results are favorable and recurrence after partial resection can take years to cause symptoms.

We present the scarcely frequent case of an intradural/extramedullary lumbar neurenteric cyst with a mucinous component in a teenager who showed no other disorders associated.

Medical case

Eighteen year-old male with no specific medical history showing an intradural tumor in lumbar spine detected by means of MRI. The patient reports two-month history of abrupt-onset unspecific-lumbar pain during sports practice, followed by left L4 dermatome paresthesia.

General physical examination does not reveal further data; the neurologic examination of the lower limbs does not give relevant information either; muscular tone and trophism are normal and so are reflexes, with no sphincter alterations.

Anterior-posterior and lateral lumbar spine X-rays are normal. MRI shows a cyst-like, oval, intradural, extramedullary, homogeneous, unilobar T1 and T2 hyperintense mass in the L2 vertebral body (Figure 1), which occupies the whole diameter of the spinal canal with no obstruction to cerebrospinal fluid. There are not other lumbar lesions. Blood and urine tests are normal.

We decided to carry out the resection of the tumor with definite histological assessment; we used a posterior approach with laminectomy. Upon incising the dura mater, we identified the lesion. It was necessary to carry out thorough dissection of two roots intimately adhered to the capsular plane of the lesion. Once isolated the tumor, we incised the capsule getting a component of mucinous aspect. Then, we removed one of its walls and took frozen intra-operative biopsy (Figure 2). Since the histological report was negative for bacteria, parasites and cells with malign criteria, we decided to carry out as ample a resection as possible with no injury of the roots adhered to the walls of the lesion. Moreover, we asked for general and specific germs cultures. The histological studies revealed as final diagnosis: neurenteric cyst whose microscopic slides showed fragments of the cyst wall lined by only ciliated columnar epithelium with glands lined by simple cubic cells and some mucinous cells. There were neither neoplastic cells nor microorganisms in the cultures.

The patient did post-operatively well, with neither lumbar pain nor paresthesias. Two months after the surgery, the patient was perfectly well, he did daily activities normally, and the symptoms that he had suffered before the surgery were completely gone.

At 12-month follow-up, he did not suffer pain and physical examination was normal; he did daily activities and sports unhampered.

Discussion

Neurenteric cysts in the central nervous system are developmental anomalies which cause a defective division between the neuroectoderm and the endoderm during the third week of embryogenesis. They are defined as cyst lesions lined by ciliated or simple columnar epithelium which can present mucous-secretor characteristics similar to those of the respiratory or gastrointestinal tracts. This is because, during the early stages of development, the notochordal or neurenteric canal connects the vitelline sac with the amniotic chamber and gives place to a temporary direct connection between the proto- spine, vertebral column and intestine; therefore, failures in this developmental stage can cause combined anomalies in these three regions.[1,2,4, 13]

They have been named different names: neurenteric cysts, enterogenous cysts, teratoma cysts, etc.[14,15] They are benign low-prevalence neoplastic lesions; their malignant transformation is very rare.[16,17] Neurenteric cysts account for 0.3% to 1.3% of all spinal tumors. The male: female ratio is 2:1. They are most frequently located at thoracic and cervical levels. Chavda et al. reported eight cases of neurenteric cysts and half of them were located at high thoracic level, two, at cervical level, one was located at the level of the medullary cone and another one at that of the brain stem.[6] About 95% of the cysts occupied the intradural/extramedullary compartment, whereas 5% were intramedullary.[13,15,18]. There are reports on rare cases of other locations in the central nervous system.[7-9,19-21]

They preferably affect children and young adults (the diagnosis of neurenteric cyst is made during the first decade of life in 34% of the cases and during the second decade of life in 23% of the cases), following years of slow growth.

Spinal neurenteric cysts cause the neurological symptoms and signs usually associated with spinal slow-growth masses. Pain in the affected area of the vertebral column is frequent and prominent, and it could be accompanied by root pain or meningeal disorders with subsequent sensitive or motor deficit. The location of the lesion determines the early symptoms. Different reports in specialized literature affirm that the most frequent early symptoms are quadriparesis or paraparesis (83.8%), sensorial dysfunction (74.2%), decreased reflexes (78.1%), pain (72.2%)

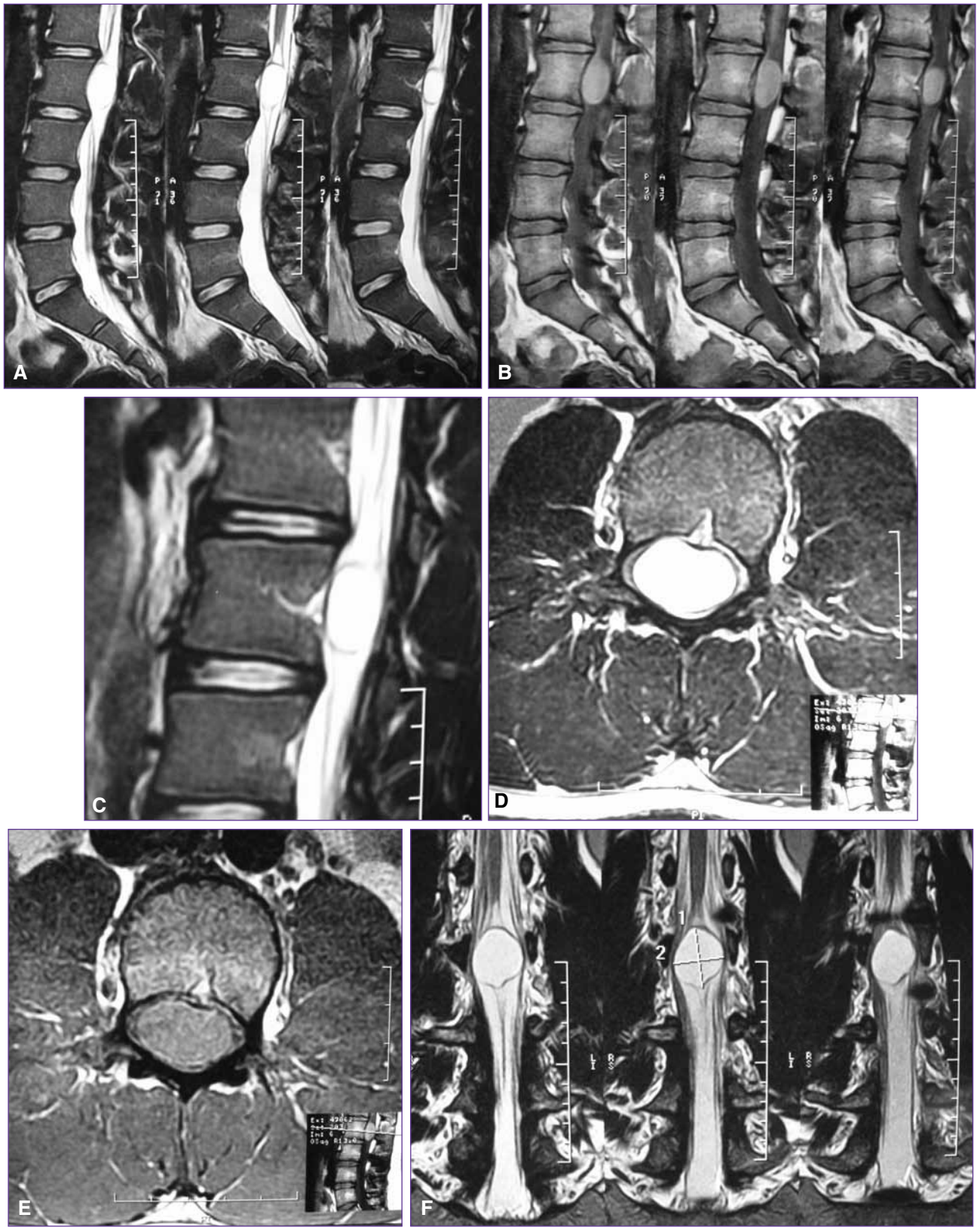

Figure 1. MRI in T1 and T2. **A and B.** Sagittal sections showing the oval cyst behind the body of L2. **C.** Detail of sagittal section showing the neurenteric cyst. **D and E.** Transverse sections: the cyst occupies virtually the entire diameter of the spinal canal. **F.** Coronal section.

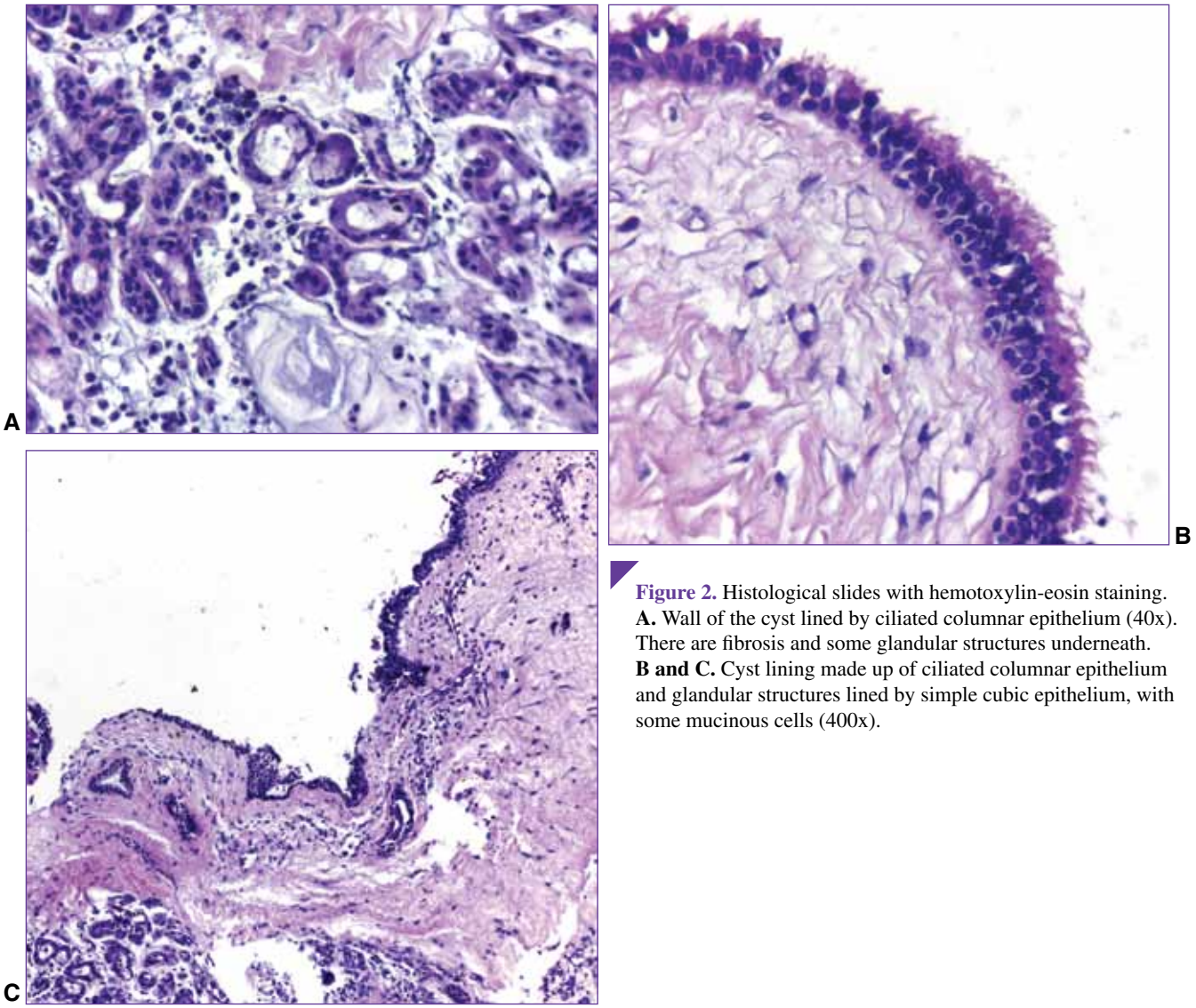

Figure 2. Histological slides with hemotoxylin-eosin staining. **A.** Wall of the cyst lined by ciliated columnar epithelium (40x). There are fibrosis and some glandular structures underneath. **B and C.** Cyst lining made up of ciliated columnar epithelium and glandular structures lined by simple cubic epithelium, with some mucinous cells (400x).

and sphincter dysfunction (50.1%).[1,3,4,14,17-26] Other symptoms less frequently found are incontinence, urinary tract infection and acute paraplegia, which can be considered either traumatic or caused by the increase in the cyst size.

Unless early diagnosis is made, the condition generally remains for many years and it is often characterized by remissions and recurrences.

MRI is the study of choice for neurenteric cysts diagnosis, [17,22,26] whereas CT scan myelography is more useful to determine the communication between the cyst and the subarachnoid space. In MRI, neurenteric cysts show intensity similar to or higher than that of the cerebrospinal fluid, both in T1 and T2. Due to the high protein contents, hyper-intensity of the cyst contents is evident both in T1 and T2. Gadolinium does not enhance the wall of the cyst. In general, these lesions show as space-occupying masses with medullary and extramedullary components, as multilobar but also as unilobar lesions.[26]

Neurenteric cysts are associated with vertebral body anomalies and spina bifida; therefore, simple X-rays and CT scan can contribute to the identification and description of vertebral anomalies such as anterior or posterior spina bifida, vertebral body widening, fused vertebras, hemi-vertebras and diastematolmyelia. [14,27,28]

With respect to the histological characteristics of these cysts, nomenclature has been widely discussed; the best one is the histological classification modified by Wilkins and Odom[13] which considers three types of cysts:

Type A: Cysts lined by a simple epithelium of cubic or columnar pseudostratified cells, with or without cilium, which imitate the gastrointestinal or respiratory epithelium and cover the basal membrane lying on vascular connective tissue.

Type B: Epithelial cysts with glandular structures that can show in complex invaginations; they usually produce serous or mucinous fluid. The cyst wall can have elements

of smooth muscle, striated muscle, fat, cartilage, bone, elastic fibers, lymphoid tissue, nerve fibers, ganglion cells or Vater-Pacini corpuscles.

Type C: They are the most complex cysts, containing ependymal or glial tissues.

The case we present here is a Wilkins A-type neurenteric cyst.

At the time of diagnosis, neurenteric cysts have to be differentiated from schwanommas, neurofibromas and meningiomas, which together represent 55% of all primary spinal neoplasias and 80% of the neoplasias of the intradural/extramedullary compartment.[29] Other less frequent lesions are lipomas, dermoid cysts, epidermoid lesions, and subarachnoid metastasis.[29,30]

Neurofibromas usually occur in patients with type I neurofibromatosis; meningiomas are the most frequent ones at thoracic level (80%). Epidermoid and dermoid cysts represent less than 1%.[17,29] Malignant transformation is exceptionally infrequent.[16-18]

Intramedullary spinal neurenteric cysts should undergo surgical treatment. They frequently show firm adherences to the spine or the cauda equina; therefore, full resection is not feasible, but anyway the evacuation and partial resection of these lesions accompanied by cyst drainage get the same good results as full extirpation. [4,6,11,12,23,24] Neurological function improves and recurrence after partial resection can take years to cause symptoms. The strategy for the resection of these cysts is different from that used in solid tumors, because the cyst can contain irritating substances that should not make contact with the subarachnoid space.

Kumar et al. carried out a two-year follow-up in 13 patients with spinal neurenteric cysts, out of which four were intramedullary. Six patients were subject to surgery, and full resection was only got in one patient; there were five recurrences related to incomplete resection.[25]

Long-term prognosis in the treatment of the neurenteric cyst is, in general, good, especially in children with moderate pre-operative symptoms. However, cysts tend to recur and, in general, full removal is difficult due to the adherences to important structures.[4,11,12,23-25]

The case we present here coincides with the epidemiological and medical characteristics described in bibliography. There were not lumbar spine anomalies associated. During the patient's follow-up, cultures and staining did not show data about neither infection nor malignant transformation.

The patient was treated by posterior approach with radical surgery technique (removing as much as possible), because the cyst showed intimate adherences to nerve roots, so, we aimed at preserving radicular function. Although it was not possible to get full resection of the cyst walls, the lesion size decreased remarkably and, therefore, it was possible to alleviate symptoms with no functional impairment.

In conclusion, it is possible to affirm that this rare lesion should be inferred in young patients who show recurrent lumbar pain or neurological signs; imaging studies lead to presumptive diagnosis, but diagnosis is verified by the histological studies that follow surgical resection, which has to be as radical as possible so as to avoid or delay recurrences and give the patient their previous life-quality back for him or her to do daily activities normally.

Bibliography

1. Kubie LS. A clinical and pathological study of two teratomatous cysts of the spinal cord, containing muscus and ciliated cells. *Surg Gynec Obstet* 1928;47:297-311.
2. Puusepp M. Variete rare de teratome sous-dural de la region cervicale (intestinome). *Rev Neurol Pari* 1934;2:879-886.
3. Holcomb GW Jr, Matson DD. Thoracic neurenteric cyst. *Surgery* 1954;35:115-121.
4. Lippman CR, Arginteanu M, Purohit D, Naidich TP, Camins MB. Intramedullary neurenteric cysts of the spine. Case report and review of the literature. *J Neurosurg* 2001;94:305-309.
5. Tekkok LM. Intramedullary epidermoid cysts of the cervical spinal cord associated with extraspinal neuroenteric cysts: case report. *Neurosurgery* 1992:31(l):121-125.
6. Chavda SV, Davies AM, Cassar-Pullicino VN. Enterogenous cysts of the nervous system: a report of eight cases. *Clin Radiol* 1985;36:245-251.
7. Nicholas ELJ, Neil K, Francesco S, Gerald BB. Neurenteric cyst of the cerebellopontine angle: case report. *Neurosurgery* 1998; 42:655-658.
8. Perry A, Scheithauer BW, Zaias WB, Minassian HV. Aggresive enterogenous cyst with extensive craniospinal spread: case report. *Neurosurgery* 1999;44:401-405.
9. Scaravelli F. Neuroenteric cysts of the optic nerve: case report with immunohisto-chemical study. *J Neurol Neurosurg Psychiatry* 1992;55(2):1197-1199.
10. Schelper RL, Kagan-Hallet KS, Huntington HW. Brainstem subaracnoidal respiratory epithelial cyst: report of two cases and review of the literature. *Hum Pathol* 1986;17:417-422.

11. Orrego Puelles J, Heinicke Ydfiez H, Arbaka Aldazabal D, Chang Kooseng A, Yépez Rios V. Quiste neuroentérico intraespinal. *Rev Diagnóstico* 2000;39(1):43-46.

12. Ramos-Zúñiga R, Casillas-Espinosa P, Sánchez-Prieto J, Encarnación Ibarra O, López Hernandez M. Quiste neuroentérico y embarazo. Informe de un caso con seguimiento a largo plazo. *Cir Cir* 2011;79:361-365.

13. Wilkins RH, Odom GL. Spinal intradural cysts. En: Vinkin PJ, Bruyn GW (eds.) *Handbook of Clinical Neurology. Tumors of the Spine and Spinal Cord, Part II*, Amsterdam: North-Holland; 1976:55-102.

14. Khosla A, Wippold FJ. CT myelography and MR imaging of extramedullary cysts of the spinal canal in adult and pediatric patients. *Am J Roentgenol* 2002;178:201-207.

15. Rosai J (ed.) *Rosai and Ackerman´s Surgical Pathology,* 9th ed. St. Louis, MO: Mosby; 2004:2468-2471.

16. Ribas Ariño T, González Medina A, Santos Salas Valién J, Izquierdo García F, Fernández Fernández J. Adenocarcinoma en quiste neuroentérico. 7º Congreso Virtual Hispanoamericano de Anatomía Patológica, 2005. Trabajo 316.

17. Kudo N, Hasegawa K, Ogose A, Hotta T, Tojo T, Otsuka H, Endo N. Malignant transformation of a lumbar intradural dermoid cyst. *J Orthop Sci* 2007;12:300-302.

18. Russell DS, Rubinstein LJ. *Pathology of Tumors of the Nervous System,* 5th ed. London: Edward Arnold; 1989:762-775.

19. Gimeno A, López F, Figuera D, Rodrigo L. Neuroenteric cyst. *Neuroradiology* 1972;3:167-172.

20. Kapoor V, Johnson DR, Fukui MB, Rothfus W, Jho H. Neuroradiologic-pathologic correlation in a neurenteric cyst of the clivus. *Am J Neuroradiol* 2002;23:476-479.

21. Kim CY, Wang KC, Choe G, Kim HJ, Jung HW, Kim IO, et al. Neurenteric cyst: its various presentations. *Childs Nerv Syst* 1999; 15:333-341.

22. Brooks BS, Duvall ER, Gammal T, Garcia JH, Gupta KL, Kapila A. Neuroimaging features of neurenteric cysts: analysis of nine cases and review of the literature. *Am J Neuroradiol* 1993;14:735-746.

23. Garg N, Sampath S, Yasha TC, Chandramouli BA, Devi BI, Kovoor JM. Is total excision of spinal neurenteric cysts possible? *Br J Neurosurg* 2008;22(2):241-251.

24. Klekamp J, Samii M. Extramedullary tumors. En: Klekamp J, Samii M (eds.) *Surgery of Spinal Tumors*, Berlin: Springer; 2007: 144-312.

25. Kumar R, Nayak SR. Unusual neurenteric cysts. Diagnosis and management. *Pediatr Neurosurg* 2002;37:321-330.

26. Gebauer G, Farjoodi P, Sciubba D, Gokaslan Z, RileyIII L, Wasserman B, et al. Magnetic resonance imaging of spine tumors: classification, differential diagnosis, and spectrum of disease. *J Bone Joint Surg Am* 2008;90(Suppl 4):146-162.

27. Rhaney K. Enterogenous cysts and congenital diverticula of the alimentary canal with abnormalities of the vertebral column and spinal cord. *J Pathol Bacteriol* 1959;77:457.

28. Santos de Oliveira R, Cinalli G, Sainte-Rose C. Neuroenteric cysts. En: Özek MM, Cinalli G, Maixner WJ (eds.) *Spina Bifida. Management and Outcome*. Milan: Springer; 2008:475-485.

29. Masaryk T. Neoplastic disease of the spine. *Radiol Clin North Am*1991;29:829-845.

30. Zimmerman RA, Bilaniuk LT. Imaging of tumors of the spinal canal and cord. *Radiol Clin North Am* 1988;26:965-1007.