

Neuroenteric cyst of the cauda equina without associated malformation

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Abstract: We report a case of 47-year-old women who presented with progressive lumbalgia since 14 years, exacerbated a year previously by bilateral lumbar radicular pain, gait disturbance and urinary retention for one month.

Neurological examination found spastic paraparesia and cauda equina syndrome. X-rays were normal but lumbar spinal cord magnetic resonance images showed an intradural extramedullary space occupying lesion.

The patient underwent L2-L5 laminectomy with total removal of the lesion (cystic, solid components and wall).

Postoperatively neurological deficits recovered totally within 2 weeks.

Histological examination showed a Type A neuroenteric cyst. (p96-99)

Key words: Neuroenteric cyst and cauda equina syndrome

Introduction

The first description of neuroenteric cysts was described by PUUSEPP in 1934.¹ They are rare and quoted under several names in the medical literature; intestinoma, gastrocytoma, enteric cyst or teratoma. This latter is a wrong diagnosis as teratoma arise from the initial three embryonic layers (ectoderm, mesoderm, endoderm) while neuroenteric cyst only arise from the endoderm. The term enterogenous cyst was first used in 1958, whereas in recent years the name neuroenteric cyst has gained increasing support.⁸

These cysts are congenital malformations tied to precocious abnormalities of the embryonic development, corresponding to an ectopic presence of an endodermic tissue inside the spine.

Clinical findings are spinal cord compression syndrome depending on the level of the compression. In our case, clinical finding was cauda equina compression syndrome.

The encephalic locations are exceptional; they are located in the posterior fossa. Through this observation and a review of the literature, we expose the clinical, diagnosis, therapeutic, histological, and evolutive particularities of this disease.

Case Report

MHS, a 47-year-old with hypertension since 4 years. Presenting 14 years before her hospitalisation, with lumbar pain which was managed successfully with analgesic and non-steroid anti-inflammatory, and bilateral lumbar radicular pain experienced a year before her hospitalisation which worsened a month prior to her hospitalisation with paraparesis, urinary retention and constipation; without fever.

Clinical examination found paraparesis and perianal anaesthesia.

X-rays were normal but magnetic resonance images (MRI) showed an intradural extramedullary space occupying lesion, extending from L3-L4 heterogenous (with cystic and solid component), hypointense on T1-weighted images, and hyperintense on T2-weighted images. The solid component enhanced following contrast injection (Figs. 1 and 2).

The patient underwent laminectomy from L2-L5, with total removal of an intradural extramedullary lesion (cystic, solid component and the wall of the lesion).

Histological examination found neuroenteric cyst of Type A (Figs. 3 and 4).

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Figure 1 ← Sagittal T1 MRI view: Space occupying lesion of the cauda equina, intradural-extramedullary, with double component; cystic and solid components.

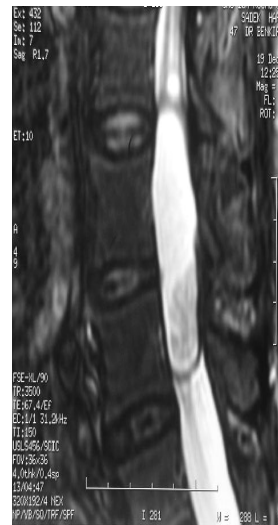


Figure 2 → T2 MRI: showing the importance of the cystic component.

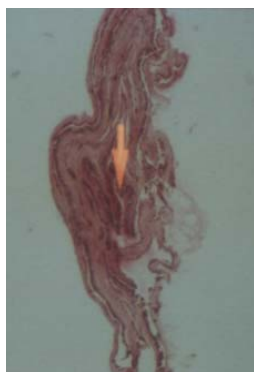


Figure 3 - Microscopic aspect of the cyst wall, pseudostratified aspect (HE × 2).

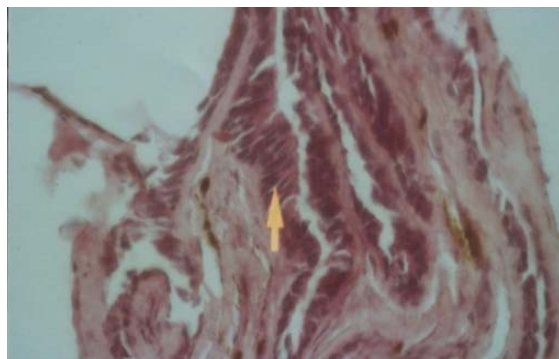


Figure 4 - The pseudostratified aspect of the cyst wall (HE × 4).

The evolution is noted by total recovery of her motor deficit within 2 weeks with sphincter disturbances recovering a short time later.

Discussion

Physiopathogenesis: The presence of gut tissue inside the

spine can be explained by an embryogenesis disorder. This occurs at the moment of chordoendoblastic raphia in the third week of embryonic life, characterised by a transitory adhesion of the chordal duct to endoderm and by it opening inside the primitive gut.^{7,9} At this fugitive stage, there are normal presence of neuroenteric canal cells which connects ectoderm to endoderm by the chordal tissue. This canal connects directly the yolk sac to the intestinal cavity. Later, the endoblastic wall is closed again, the inside of the cord is filled and the cord cut off again.

This embryogenesis has evoked several theories:

- Bentley and Smith, have described a syndrome called “split notochord syndrome” explaining that the initial phenomenon is a division of the notochord, leading to a duplication of the neural canal; through this opening, the endoderm will intervene and adhere to the ectoderm.¹⁰ According to the grade of the regression of this process, there will be different lesions (spina bifida, dermic sinus, diastematomyelia, neuroenteric cyst).
- According to Bremner, the transitory persistence of communication between the amniotic sac and the yolk sac on the 18th and 19th day, can explain the genesis of neuroenteric cyst; in this case there was an herniation of endoderm through the neuroenteric canal which divided both the vertebra body and the spinal cord “accessory neuroenteric canal theory”.

It is noted that the location of the neuroenteric canal (corresponding to the location of the Hensen knot), is situated on the coccyx; the existence of accessory or neuroenteric canal. The “split notochord syndrome” can be compared to the fact that the dorsal part of the primitive gut attaches

itself to ectoblast by hyper compression in the digestive cavity. The digestive fistula to the skin is the ultimate phenomenon and the main result of these dysraphias, but all the intermedial malformations can exist during the in-utero life and can be completely or incompletely repaired.

Spinal or spinal cord malformation can persist as rachischisis, diastematomyelia, myelomeningocele; or can reduce itself.

Epidemiology: Neuroenteric cyst is rare and is frequently difficult to establish.^{5,8} In a series of 250 spinal cord space occupying lesions we notice only 3 cases of neuroenteric cyst.

There is a male predominance, with a mean age of 29 years (8 days - 72 years), but those associated with spinal malformation occur at a young age.⁸ In our case, the patient was 47 years old.

Location: Neuroenteric cysts can occur at all sections of the spine, with a noted preference for the cervicodorsal region; the cauda equina location is also rare. They are often intradural extramedullary. Those intramedullary occur at the level of the lower thoracic spinal cord and have been constituted probably at the late stage of the embryogenesis because of the delayed closing of the neural tube at the cephalic and caudal extremities.^{7-9,12}

Clinical findings: The clinic state is variable according to the age, the location and associated malformation:

- Fortuitous discovery during myelomeningocele surgery in newborn.
- Aseptic or purulent meningitis caused by cyst rupture. (E. Coli).
- Pseudosyringomyelia by the cyst rupturing into the spinal cord.
- Spinal cord compression syndrome; this syndrome is the most frequent form of presentation.^{1,4,9,12,13,16} Common symptoms are radicular pains.^{8,13,16} In our case, the presentation was cauda equina syndrome preceded by long-time bilateral L5 radicular pain.

Other malformations can be associated with neuroenteric cyst such as rachischisis, myelomeningocele or scoliosis.

Radiologic findings: Standard x-rays are often normal but in certain cases they can show anomalies such as anterior or posterior spina bifida, spondylolisthesis, or vertebra fusions.^{2,3,6,12,14} The best radiologic examination is MRI which shows the T1 hypointense and T2 hyperintense lesion; with double component, cystic and solid one; the solid component is often enhanced by the contrast.^{8,12,14,15}

Magnetic resonance imaging findings in our patient were

T1 hypointense and T2 hyperintense lesion at the level of L3 and L4 located intradural-extramedullary, with cystic and solid component enhanced by contrast.

Treatment: The only efficient treatment of neuroenteric cyst is total surgical removal. This surgery is based on three principles.^{13,15,16}

- Decompression of nervous tissue must be soft in respect to the spinal cord and vascularization.
- Opening and inspection of the cyst wall is sufficient because the neuroenteric cyst never recurs. We must be content with simple biopsy of the cyst wall.
- Assure the stability of the spine.

Our patient underwent L2-L5 laminectomy without spinal stability compromise.

Any associated malformation should be treated at the same time.

Histology: Histologically, three types of neuroenteric cyst are distinguished based on the structure of the cyst wall⁸:

- Type A: Cysts are lined by a single-layer or pseudo-stratified cuboidal or columnar, ciliated or non-ciliated epithelium, resembling gastrointestinal or respiratory epithelium, mounted on a basement membrane.
- Type B: The wall of the cyst, in addition, contains mucous or serous glands, smooth muscle, various connective tissue components, lymphoid tissue and even nerve ganglion.
- Type C: May also have ependymal and other glial elements.

About 50% of cysts are lined by gastrointestinal-type epithelium; 17% by respiratory and 33% mixed.

In our patient, the cyst was Type A.

Electron microscopy and immunohistochemistry⁸: Ultrastructural examination of neuroenteric cysts shows striking similarities with colloid cyst, Rathke's cleft cysts and follicular cysts of the normal pituitary gland. Their immunophenotype is strikingly similar to that of other cysts showing endodermal or ectodermal differentiation or to the respiratory tract mucosa. The epithelial cells of neuroenteric cyst are positive for cytokeratins, EMA, secretory component and vimentin, and bind U. europaeus lectin, but fail to stain with antibodies to prealbumin (transthyretin), S-100, GFAP and NSE.

Evolution: Neuroenteric cysts generally have a good outcome but the prognosis depends on the initial neurologic deficit.^{1,5,9}

Early diagnosis and immediate surgery allow total and rapid recovery.

In our patient the recovery was total.

Conclusion

Neuroenteric cyst of cauda equina is a rare complex spinal dysraphy. It generally occurs in male, young patients.

Clinical state is dominated by the cauda equina syndrome. Spinal MRI is the most important examination for diagnosis assessment. It is often located intradural-extramedullary and frequently associated with spinal malformation.

Histologic examination determines its endodermal nature and type.

Main treatment is the surgical total removal of the cyst and the prognosis is often good.

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