

Case report

Antenatally diagnosed intraspinal-posterior mediastinal neurenteric cyst - what is the optimal management?

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ABSTRACT

Neurenteric cyst is a rare congenital anomaly that belongs to the spinal dysraphism spectrum. It is classically a disease of late childhood, with some rare reports of intrauterine and adult presentation. The increase in incidental antenatal diagnosis raises new management questions. We present a case of an asymptomatic combined intraspinal-posterior mediastinal neurenteric cyst. The cyst was diagnosed on antenatal ultrasound and was initially treated with image-guided aspiration in early infancy. However, the cyst recurred and the patient developed recurrent episodes of bacterial meningitis. Two-stage surgical resection was performed, although the infection prohibited complete excision. We caution against image guided aspiration of neurenteric cysts, and suggest a management algorithm based on the available literature.

1. Introduction

Neurenteric cyst, also known as endodermal or bronchial cyst, is a rare congenital space-occupying lesion which results from the persistence of the embryologic neurenteric canal that normally closes around the 3rd week of embryogenesis, allowing proper segregation of the foregut from the notochord [1]. Neurenteric cysts are classified as part of the occult spinal dysraphism spectrum [2] since the split notochord often leads to ventral spinal dysraphism. This explains the occasional continuation of a neurenteric cyst between the spinal canal and the posterior mediastinum or the abdominal cavity [2].

Traditionally, neurenteric cysts are diagnosed when they became symptomatic: posterior mediastinal neurenteric cysts can manifest with respiratory compromise in early childhood [3,4], or even during the intrauterine life [5], whereas intraspinal neurenteric cysts typically present with progressive myelopathy during childhood [6].

The management of asymptomatic neurenteric cysts, however, is a more controversial issue. With the advances of intrauterine imaging, antenatal diagnosis of neurenteric cysts is reported more frequently [7], adding to the controversy on how to manage an asymptomatic, incidentally diagnosed neurenteric cyst.

Here we present a case of an asymptomatic, antenatally diagnosed epidural and posterior mediastinal neurenteric cyst that was poorly managed with image-guided aspiration, only to complicate its clinical

course. In addition, we propose a management algorithm based on the available literature.

1.1. Illustrative case

A female fetus of a 30 year-old woman, G1P0, was found during a routine prenatal ultrasound (that was performed in another institution) to have posterior mediastinal and intraspinal cysts. The child was born at term via a vaginal delivery without perinatal complications. The neonate did not have any respiratory distress and had a normal neurological examination. She had a normal cardiac evaluation. The patient did not have any cutaneous stigmata. There was no focal tenderness or limitation in the cervical spine movement.

MRI with and without contrast was performed to evaluate the cyst, showing a ventral, intradural extramedullary cystic lesion with faintly enhancing wall, spanning C7 to T4 levels in the spinal canal, measuring 1.4 × 0.7 × 1 cm; and another posterior mediastinal cyst measuring 3.5 × 2 × 2 cm, extending from the esophageal hiatus all the way cranially behind the carina. The two cysts had a communication through the mentioned vertebral bodies defect (Figs. 1 and 2). CT scan was performed to evaluate the cyst, showing dysmorphic vertebrae with ventral clefting of the C7 and T1 vertebral bodies, thoracic hemivertebra, along with scoliosis of the cervicothoracic junction (Fig. 3).

Due to the parents' concerns, the outside treating physician decided

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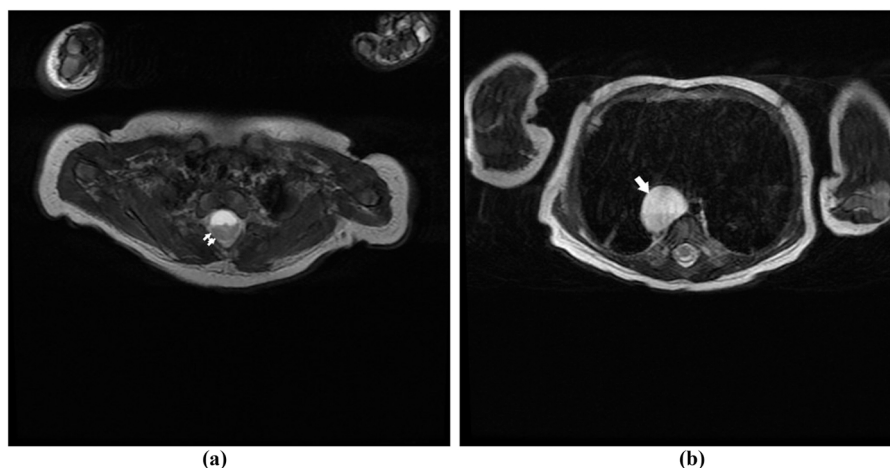


Fig. 1. MRI axial cervical (C7 level) (Fig. 1a) and axial thoracic (T8 level) (Fig. 1b) T2W sequence images. Both cysts are hyperintense on T2. The intraspinal component is intradural, extramedullary. Notice its ventral location in relation to the spinal cord (double arrows on the cord, single arrow on the mediastinal cyst).

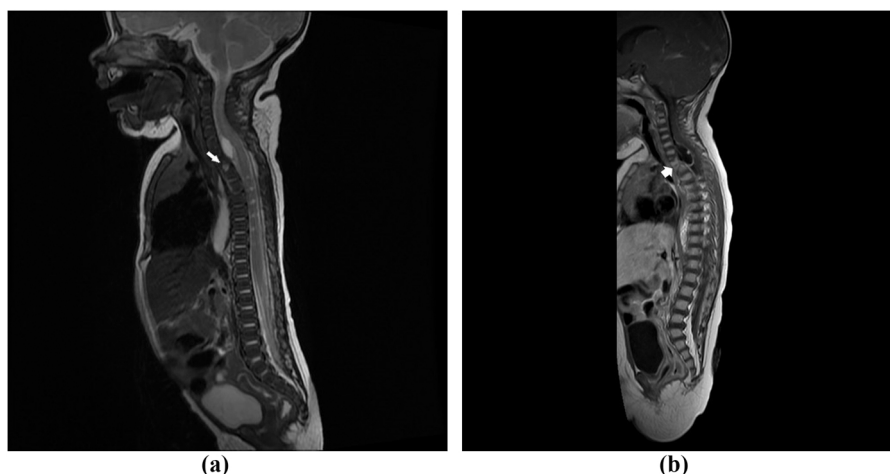


Fig. 2. 2a: Mid-sagittal T2W MRI prior to surgery, notice the trans vertebral communication channel (white arrow). 2b: Mid-sagittal T1W MRI with gadolinium performed between the two surgeries. Notice the ventral location of the cyst in the intradural compartment. There is no clear plane between the cyst and the spinal cord. The vertebral body defect anterior to the cyst is indicated by a wide arrow.

to perform CT-guided aspiration of the asymptomatic posterior mediastinal cyst at the age of 2 months. The cyst was totally aspirated. Interestingly, contrast injected into the empty posterior mediastinal cyst cavity filled the trans vertebral communication channel but did not fill the intraspinal cyst, raising the possibility of a one-way valve through the communication channel (Fig. 3).

Following this seemingly simple and minimally invasive procedure, the patient presented a week later with clinical signs of bacterial meningitis. CSF culture was positive for *Pseudomonas aeruginosa*. After finishing the first course of intravenous antibiotics, the meningitis relapsed again due to the same pathogen. Repeated CT scan showed that the aspirated cyst has filled and increased in size again.

Beside the relapsing meningitis, the patient's pulmonary and neurological status remained stable. However, we decided that surgical resection was necessary to prevent further infectious complications. At the age of 5 months, the patient underwent right posterolateral thoracotomy for excision of the posterior mediastinal cystic mass. The cyst content was drained, and its adherence to the upper thoracic spine was carefully coagulated. The patient had a smooth short post-operative stay, however, she presented 3 weeks later with another episode of meningitis. MRI imaging showed an increase in size of the intradural cyst with progression of cord compression. After proper antibiotics treatment, she underwent C6-T2 laminoplasty exposing the cyst which

was ventral to the spinal cord. It contained clear mucinous material, but its wall was intimately adherent to the ventral surface of the spinal cord, prohibiting total excision. The cyst was fenestrated through the windows between the spinal roots and resected in a piecemeal fashion, leaving a rim of its wall adherent to the spinal cord.

Histological examination of the intraspinal and mediastinal specimens revealed the same cyst wall, composed of pseudostratified columnar respiratory ciliated epithelium. At the time of writing this manuscript the patient has been neurologically intact in a good condition for the past six months. She has been advised about the risk of cyst relapse and the importance of long term neurological follow up.

2. Discussion

The natural history of neurenteric cysts is variable [8], with a wide range of clinical presentations, depending on the cyst size, extent, and its mass effect on the adjacent structures. Traditionally, neurenteric cysts have been diagnosed and treated when they became symptomatic. However, incidental diagnosis is increasingly common, challenging the treating physicians about the optimal management decision.

Neurenteric cyst is usually a disease of late childhood, with few reported adult-onset cases [8]. In a 23-cases single center series, the median age at presentation was 15 years [6]. Complete surgical resection is

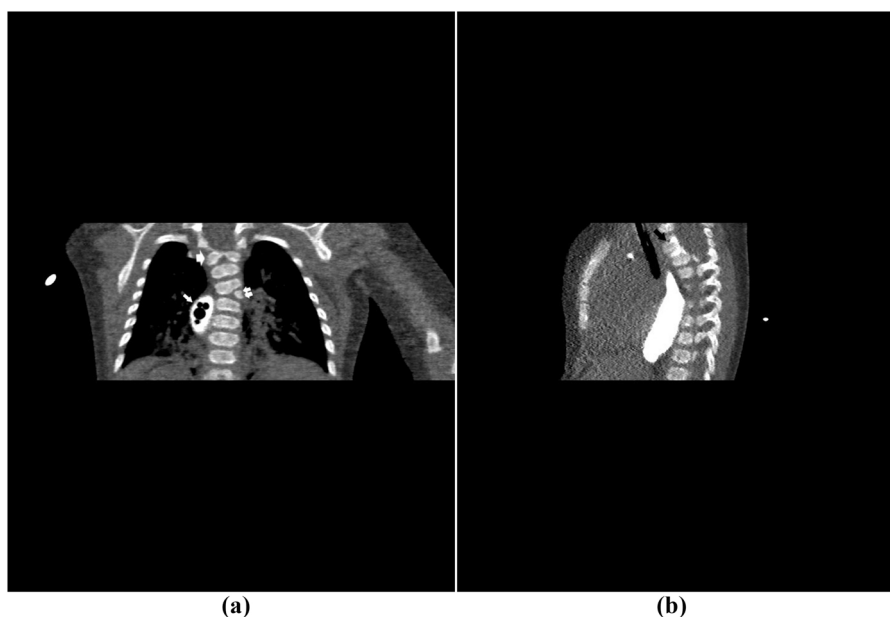


Fig. 3. **3a:** Coronal CT scan during the guided aspiration. The right posterior mediastinal cyst (arrow) has been emptied and filled with contrast. Notice the thoracic hemivertebra (double arrows) and the upper thoracic vertebral body cleft (wide arrow). **3b:** Sagittal CT scan. The contrast fills the posterior mediastinal cyst, and spreads through the communication channel (black arrow) through vertebral bodies, till it reaches the intraspinal compartment.

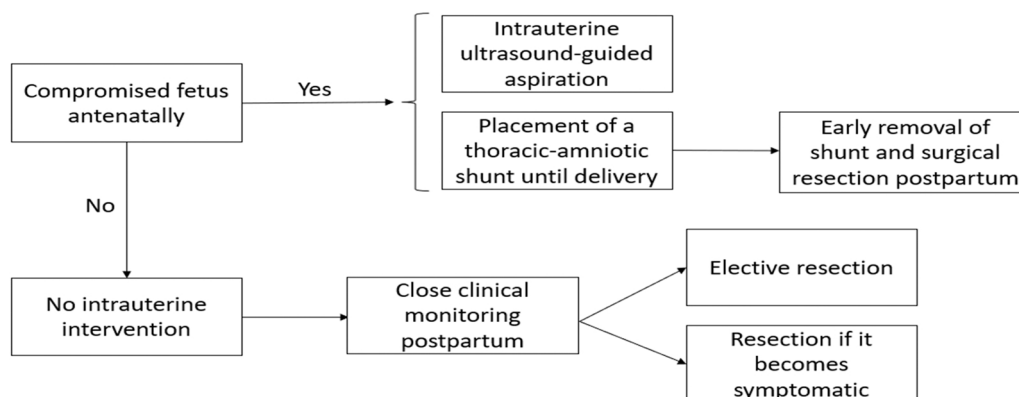


Fig. 4. Suggested algorithm for the treatment of antenatally diagnosed neurenteric cyst.

the advocated modality of treatment, as partial resection is the main risk factor for cyst recurrence [9–11]. Posterior spinal approach and posterolateral thoracotomy are the most reported surgical approaches for intraspinal and intrathoracic cysts, respectively. Many authors advocate the resection of incidental intraspinal [12,13] and thoracic cysts [14,15] prior to the onset of symptoms, since many symptomatic patients will actually remain with neurological deficits even after surgical treatment [10,16].

In our case, the image-guided cyst aspiration did not only fail to eradicate the cyst, it also complicated its clinical course with bacterial meningitis. The infection presumably spread through the trans-vertebral connection that was visualized on the MRI from the posterior mediastinal cyst to the meninges. Had it not been for the infection, we would have better chances in dissecting the intradural cyst off the spinal cord and achieve complete surgical resection. Now that the cyst is incompletely resected, the patient needs long term follow up and, if the cyst grew again, a possible further surgical intervention.

On the other end of the clinical spectrum, neurenteric cyst can become symptomatic as early as intrauterine life. Intrauterine treatment of posterior mediastinal neurenteric cysts (e.g. intrauterine ultrasound-guided aspiration) might be necessary when signs of hydrops and fetal

edema are present [6,17,18]. Even more, simple aspiration is not always successful in preventing the re-accumulation of the cyst, and two authors [6,17] reported the need for placement of a thoracic-amniotic shunt, as a temporizing measure, until delivery. Removing the shunt postpartum in these two cases led to rapid cyst growth that required early surgical resection [6,17]. In fact, a fetus that has a posterior mediastinal neurenteric cyst associated with hydrops and ascites, might die if a timely intrauterine drainage is not performed [7].

While prenatal sonography is still the imaging of choice for detecting intrauterine anomalies, fetal MRI is gaining acceptance as a more accurate diagnostic method, especially in neurological defects assessment [19]. It has been used for intrauterine neurenteric cyst evaluation [14, 20,21], and even more, fetal MRI can detect neurenteric cysts that are not detected by sonography [21].

Based on our experience and review of the literature we suggest this treatment algorithm to help the treating physician in making the optimal decision that suits an individual patient (Fig. 4).

3. Conclusion

Neurenteric cyst is a rare developmental anomaly, with various

clinical presentations. While neurenteric cyst is classically a disease of late childhood, the availability of imaging modalities has increased the frequency of antenatal and incidental diagnosis. Complete surgical resection is treatment of choice, as it has the best chance of cure. Based on the available literature, asymptomatic patients might benefit as well from elective surgery, rather than observation. Image-guided aspiration could be a necessary temporizing measure for symptomatic intrauterine neurenteric cyst. We caution against the use of postnatal image-guided aspiration as it does not prevent cyst recurrence, and it can complicate its clinical course.

Author contribution

Writing the manuscript: Niovi Bejjani, Rebecca Andraos, Khaled Alok. Editing and reviewing the manuscript: all the authors.

Conflict of interest

The authors have no conflict of interest to declare.

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