

SECTION EDITOR: BEVERLY P. WOOD, MD

Radiological Case of the Month

Anne Paterson, MRCP, FRCR; Louise E. Sweeney, FFRRCS(Ire), FRCR

A FEMALE INFANT was born at 37.5 weeks' gestation, after an uneventful antenatal period and delivery. The Apgar scores were 6 and 8 at 1 and 5 minutes, respectively. Her birth weight was 3.3 kg. Thirty minutes after delivery she was noted to be tachypneic and

hypothermic. Auscultation of her chest revealed decreased breath sounds on the right side. A chest radiograph was performed after transfer to the neonatal intensive care unit (**Figure 1**). An echocardiogram revealed a patent ductus arteriosus and a patent foramen ovale. A 4 × 4-cm cystic lesion was noted in the right hemithorax and extending posteriorly and inferiorly. A magnetic resonance imaging scan was performed on day 6 (**Figure 2**, **Figure 3**, and **Figure 4**).

From the Royal Belfast Hospital for Sick Children, Belfast, Northern Ireland.



Figure 1.



Figure 2.

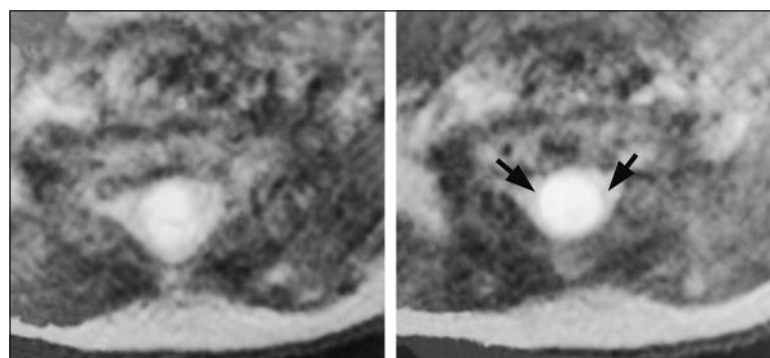


Figure 3.

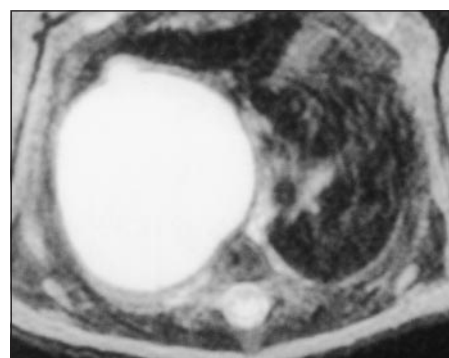


Figure 4.

Denouement and Discussion

Neurenteric Cyst

Figure 1. The chest radiograph shows partially opaque right hemithorax and displacement of the heart and mediastinal structures to the left side. Segmentation anomalies are present in the upper thoracic spine.

Figure 2. Sagittal T₂-weighted magnetic resonance image shows a high-signal mass in the right hemithorax communicating with an extradural cystic lesion via the anomalous upper dorsal vertebra (arrow). An intramedullary cystic component is also present and extends cranially and caudally. The thecal sac is indented anteriorly by the extradural mass. The subdiaphragmatic extent of the cyst is not visualized on this image.

Figure 3. Axial T₂-weighted magnetic resonance images demonstrate cord expansion (arrows) caused by the intramedullary component of the lesion.

Figure 4. Axial T₂-weighted magnetic resonance images demonstrate mediastinal shift caused by the lesion and filling of the right hemithorax by a homogenous high-signal mass.

The imaging features are indicative of a large neurenteric cyst with extradural and intraspinal components.

Surgery was performed on the sixth day after the magnetic resonance scan. The mediastinal component of the cyst was aspirated and dissected. The inferior extent of the cyst was a blind tube on the antimesenteric border of the proximal jejunum. A short section of jejunum was resected.

Postoperatively, repeated chest drainage for a right chylothorax was necessary. At age 12 months the patient is well and her neurological status is being followed up as she is awaiting neurosurgical excision of the intraspinal component of the lesion.

Neurenteric cysts are rare intraspinal masses that arise during notochord development and represent persistence of the embryonic communication between the primitive foregut and the spinal canal.¹ The cysts are well delineated, thin-walled, fluid-filled masses containing elements derived from endodermal cells.² The fluid within them is nearly identical to cerebrospinal fluid or is milky or mucinous.¹ Neuroenteric cysts lie ventral to the spinal cord and associated anomalies of vertebral segmentation are common. Patients presenting later in life may exhibit no bony abnormality other than pressure erosion of adjacent vertebra. The expected location of cysts is at the cervicothoracic junction or the region of the conus.^{1,3} Only 10% to 15% of the cysts occur intracranially.⁴

The clinical presentation depends on the location and size of the cyst. The lesions are sometimes diagnosed antenatally,^{5,6} but most patients present in the first

decade of life with signs of cardiorespiratory embarrassment. Some present in adolescence, with pain or signs of myelopathy secondary to an intraspinal mass effect.⁷

Accurate anatomical information, particularly regarding the spinal component, is a prerequisite to surgery and magnetic resonance imaging is the modality of choice.

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Reprints: Louise E. Sweeney, FFRRCS(Ire), FRCR, Royal Belfast Hospital for Sick Children, 180 Falls Rd, Belfast BT12 6BE, Northern Ireland.

REFERENCES

1. Brooks BS, Durall ER, El Gammel T, Garcia JH, Gupta KL, Kapila A. Neuroimaging features of neurenteric cysts: analysis of nine cases and review of the literature. *AJNR Am J Neuroradiol.* 1993;14:735-746.
2. Malcom GP, Symon L, Kendall B, Pires M. Intracranial neurenteric cysts. *J Neurosurg.* 1991;75:115-120.
3. Hoffman CH, Dietrich RB, Pais MJ, Demos DS, Pribram HFW. The split notochord syndrome with dorsal enteric fistula. *AJNR Am J Neuroradiol.* 1993;14:622-627.
4. Osborn AG. Miscellaneous tumors, cysts, and metastases. In: *Diagnostic Neuroradiology.* St Louis, Mo: Mosby-Year Book Inc; 1994:648.
5. Gulrajani M, David K, Sy W, Braithwaite A. Prenatal diagnosis of a neurenteric cyst by magnetic resonance imaging. *Am J Perinatol.* 1993;10:304-306.
6. Fernandes ET, Custer MD, Burton EM, et al. Neurenteric cyst: surgery and diagnostic imaging. *J Pediatr Surg.* 1991;26:108-110.
7. LeDoux MS, Faye-Peterson OM, Aronin PA, Vaid YN, Marshall Pitts R. Lumbosacral neurenteric cyst in an infant. *J Neurosurg.* 1993;78:821-825.

Submissions

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