A newborn term male infant was found to have the stigmata of VACTERL syndrome, including cervical and thoracic hemivertebra, partial sacral agenesis, 13 pairs of ribs, high imperforate anus with rectovesical fistula, and tracheoesophageal fistula with proximal esophageal atresia. No open spinal dysraphism or skin dimpling was seen. Ultrasonography of the spinal cord was performed to screen for occult anomalies. The examination revealed a lipoma of the filum terminalis and a low-lying conus medullaris at the L3-L4 level with a large central cystic structure in the terminal cord, which was continuous with the central canal (Fig. 1). This was thought to most likely represent a dilated ventriculus terminalis, and the diagnosis was confirmed on a subsequent MR image (Fig. 2). Fluid in the cyst followed cerebrospinal fluid signal in all sequences. We observed no abnormal signal in the adjacent parenchyma or enhancement associated with the cystic mass. A cranial ultrasonogram did not reveal any intracranial anomalies. The patient demonstrated no neurologic deficits in the lower extremities. Bowel and urinary continence was not assessable in this infant.

DISCUSSION

The origin of the ventriculus terminalis can be traced to the embryonic development of the spinal cord, which can be subdivided into two stages: (1) neurulation and (2) canalization or retrogressive differentiation. Neurulation, starting at the 3 week embryonic age, is responsible for formation of the neural tube by progressive closure of the neural plate and its separation from the overlying ectoderm. The distal cord develops from a caudal cell mass that forms from neural epithelium and notochord caudal to the primary neural tube. This caudal cell mass develops microysts that coalesce to form an ependyma-lined tube that fuses with the central canal of the neural tube. With differentiation, the
Figure 1  A, Longitudinal ultrasonogram of the terminal cord reveals a bulbous anechoic mass (arrows) continuous with the central canal in the region of the conus medullaris. Echogenic signals (arrowheads) are seen caudal to the conus medullaris, consistent with a lipoma of the filum terminale. B, Transverse view reveals a relatively uniform rim of cord tissue expanded by the cystic mass (arrows). The dimensions are 12 mm craniocaudad × 6 mm anteroposterior × 8 mm transverse.

Figure 2  Sagittal T1-weighted and T2-weighted MR images of the spine confirm that the cystic mass (arrow) is continuous with the central canal. The content of the mass followed cerebrospinal fluid signals, and no soft tissue nodules are associated with it. In addition, the cord terminates at the L3-L4 level and the presence of a lipoma (arrowhead) of the filum terminale is confirmed.
caudal end of the cell mass involutes, leaving a focal prominence in the ependyma-lined canal in the conus medullaris and a fibrous tail, the filum terminale.

The normal sonographic appearance of the spinal cord is well documented. The longitudinal linear central echo has been termed the central echo complex, and it was shown by Nelson and coworkers to represent an acoustic interface between the anterior median fissure and white matter fiber tracts of the ventral commissure. The presence of the ventriculus terminalis was then described as the hypoechoic splitting of this central complex (Fig. 3). More ovoid and bulbous appearances have been described by MR imaging. In the case presented here, the ventriculus terminalis is especially prominent and cystic in appearance. The conus medullaris is expanded in the affected segment, but there is no thinning of overlying neural tissue. Pathologic dilation of the ventriculus terminalis has been described by Sigal and colleagues, who postulated that the dilation may represent isolation of the ventriculus terminalis and possibly duplication of the central canal. In our case, the cyst demonstrated apparent communication with the central canal and therefore is not explained by this theory. This appearance may then represent one end of the morphologic spectrum of ventriculus terminalis. Whether or not pathologic dilation of ventriculus terminalis exists at this age is still debatable. The importance lies in distinguishing this entity from cystic neoplasm and syringohydromyelia. The lack of an associated mass or abnormal enhancement can be confirmed on MR imaging. A posttraumatic syrinx should be included in the differential diagnosis in the appropriate clinical setting.

As ultrasonography becomes used more frequently to screen for spinal cord anomalies in newborn infants, awareness of the appearance of the normal and dilated ventriculus terminalis will allow for correct interpretation of terminal cord structures. If the ventriculus terminalis appears as a hypoechoic splitting of the central echo complex and is not associated with neurologic symptoms, ultrasonography alone may be adequate. Infants with a dilated ventriculus terminalis, as in this case, should undergo MR imaging to confirm the absence of coexisting pathologic lesions of the cord.

REFERENCES

Figure 3 Axial and longitudinal sonographic views of a normal ventriculus terminalis reveal hypoechoic splitting of the central echo complex (arrowheads).