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Myelocystocele in Focus: MRI Insights into a Rare Neurological Condition

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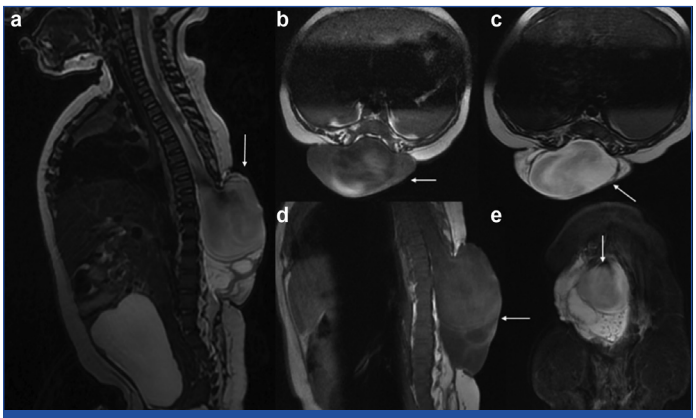
A three-month-old female child initially presented with a palpable back bulge with intact overlying skin, noticed by the mother since, birth. Clinically, there was lower extremity paralysis and sensory impairment since, birth. No ultrasound was performed, and the patient was directly advised to undergo an Magnetic Resonance Imaging (MRI) of the dorsal spine with WSS. MRI revealed a large defect in the posterior vertebral column (posterior spina bifida) at the level of D8-D12 vertebrae, through which the spinal cord, Cerebrospinal Fluid (CSF), and meninges herniated into the overlying soft tissue, forming a large, well-defined, smooth-walled cystic lesion. The lesion appeared hyperintense on T2-weighted sagittal and axial images [Table/Fig-1a,b], hypointense on T1-weighted axial and sagittal images [Table/Fig-1c,d], and demonstrated mixed signal on STIR images [Table/Fig-1e].



[Table/Fig-1]: MRI of the dorsal spine (plain) with Whole Spine Screening (WSS) reveals a large defect in the posterior vertebral column (posterior spina bifida) at the level of D8–D12 vertebrae, forming a well-defined, smooth-walled cystic lesion. The lesion appears hyperintense on T2-weighted sagittal (a) and axial images (b), hypointense on T1-weighted axial (c) and sagittal images (d), and shows mixed signal intensity on Short Tau Inversion Recovery (STIR) images (e).

There was associated widening of the spinal canal involving the adjacent paraspinal muscles and interspinous spaces from D8–D12, reaching up to the subcutaneous plane. No tethering of the cord was observed. Based on these findings, a diagnosis of dorsal myelocystocele was made. Features supporting this diagnosis included: skin-covered dorsal spina bifida, an arachnoid-lined meningocele directly continuous with the spinal subarachnoid space, and the spinal cord traversing the meningocele, expanding into a large terminal cyst. Surgery was performed three days later without complications. Follow-up at six months of age revealed normal neurological examination and appropriate developmental milestones.

Terminal myelocystocele is a rare form of concealed or occult spinal dysraphism. It is characterised by hydromyelic dilatation of the distal central canal (syringocele) bulging through a posterior spina bifida, accompanied by an enlarged dural sheath (meningocele). Although rare, it is an important differential diagnosis for a newborn presenting with a spinal mass. Ultrasonography is usually the initial screening tool; however, MRI provides superior anatomical delineation and confirms the diagnosis [1].

Neurulation is a complex process. Primary neurulation occurs when the notochord and surrounding ectoderm form a neural plate, which then folds to form the neural tube; the ends of the tube close on days 25 and 28, respectively. Secondary neurulation follows, culminating in the formation of the conus medullaris and filum terminale [2]. Aberrations in neurulation result in spinal dysraphism, which can be either open or closed. In open types, tissues are exposed and may protrude; if neural tissue is exposed, it is called a myelomeningocele. Closed types occur when neural tissue is enclosed or covered by skin or fat (e.g., lipomyelomeningocele). Abnormal notochordal formation can lead to complex dysraphism [2]. Tethering bands are released during postnatal surgical repair to minimise neurological degeneration [3].

Differential Diagnosis

1. Lipomyelomeningocele: Both myelomeningocele and lipomyelomeningocele are forms of closed spinal dysraphism. The difference is that lipomyelomeningocele contains a placode-lipoma (fat) interface that extends outside the spinal canal.
2. Myelomeningocele: Both myelomeningocele and myelocystocele cause expansion of the subarachnoid space. However, myelomeningocele is an open spinal dysraphism, with the neural placode protruding above the skin surface, whereas myelocystocele is a closed dysraphism with a dilated central canal herniating through posterior spina bifida.
3. Meningocele: Both myelomeningocele and meningocele are forms of closed spinal dysraphism. Meningocele differs in that it does not contain any part of the spinal canal within the sac.

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