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Retained medullary cord

Retained medullary cord (RMC) is a newly defined entity of closed spinal dysraphism that originates from the late arrest of secondary neurulation.

Case reports

A 6-year-old boy presented with myoclonus of the lower limbs, who had subpial lipoma at the lumbar cord, just rostral to the low-lying conus, which was tethered by a cord-like structure (C-LS) continuous from the conus and extending to the dural cul-de-sac. Following cord untethering from C-LS and minimal debulking of the lipoma, the myoclonus was improved. Histological examination of C-LS revealed a large central canal-like structure in the neuroglial core and the diagnosis of RMC was made. Subpial lipomas can be incidentally coexistent with spinal dysraphism resulting from the failed secondary neurulation, such as RMC ¹⁾

Murakami et al. report on three cases of closed spinal dysraphism, in which a spinal cord-like tethering structure extended out from the dural cul-de-sac and terminated at a skin-covered meningocele sac in the sacrococcygeal region, which was well delineated in curvilinear coronal reconstructed images of 3D-heavily T2-weighted images (3D-hT2 weighted image). Intraoperative neurophysiology revealed the spinal cord-like tethering structure was nonfunctional, and histopathology showed that it consisted of central nervous system tissue, consistent with RMC. The tethering structure histologically contained a glioneuronal core with an ependymal-like lumen and smooth muscle, which may indicate developmental failure during secondary neurulation.

When the RMC extending to a meningocele is demonstrated with the detailed magnet resonance imaging including 3D-hT2 weighted image, decision to cut the cord-like structure for untethering of the nervous tissue should be made under careful intraoperative neurophysiological monitoring ²⁾.

A retained medullary cord (RMC) is a rare spinal dysraphism, described as a late arrest of secondary neurulation. RMC is also a severely tethering lesion. The critical role of intraoperative neurophysiological monitoring to safely manage a RMC has been only anecdotally reported.

Sala et al describe the case of a RMC in a 1.5-year-old child with Currarino syndrome. At surgery, an apparently normal-looking spinal cord, stretched and tethered by a lipoma to the level of S2-S3, was observed. The border between the functional conus and the non functional RMC was defined through neurophysiological mapping. The cord was sharply interrupted at this level and untethered. A specimen was sent for pathology, which confirmed the presence of glial and neural elements. The post-operative neurological exam was normal.

Neurosurgical procedure for RMC should only be rendered with intraoperative neurophysiological mapping, as the anatomical judgment would not suffice to allow a safe cutting of these "normal-looking" neural structures ³⁾.

1)

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PMID: 34012749; PMCID: PMC8116927.

2)

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