Terminal myelocystocele

Terminal myelocystocele (TMC) is thought to be caused by a misstep during secondary neurulation. However, due to the paucity of data on secondary neurulation and the rarity of TMC, proofs of this pathogenetic mechanism are unavailable. Based on a previous observation that TMC resembles a step of secondary neurulation in chick, a closer look was taken at secondary neurulation of chick embryos focusing on the cerebrospinal fluid-filled distal neural tube (terminal balloon).

A dilated balloon doubtlessly exists in the terminal secondary neural tube in chick embryos, and its subsequent disappearance occurs in a variable time course and sequence. Arrest of apoptosis resulting in failure of detachment of the terminal balloon from the surface ectoderm may well be the basis for human TMC¹⁾.

Terminal myelocystocele (TMC) can be deconstructed into essential and nonessential features. Essential features are present in all TMCs and constitute the core malformation, comprising an elongated spinal cord extending extraspinally into a cerebrospinal fluid-filled cyst that is broadly adherent to the subcutaneous fat. The functional conus resides in the proximal cyst or within the intraspinal cord, and the caudal myelocystocele wall is nonfunctional fibroneural tissue. Nonessential features include variable measures of hydromyelia, caudal meningocele, and fat, present in only some patients. The core structure of TMC strikingly resembles a transitory stage of late secondary neurulation in chicks in which the cerebrospinal fluid-filled bleblike distal neural tube bulges dorsally to fuse with the surface ectoderm, before focal apoptosis detaches it from the surface and undertakes its final dissolution. We theorize that TMC results from a time-specific paralysis of apoptosis just before the dehiscence of the cystic distal cord from the future skin, thereby preserving the embryonic state.

Besides tethering, the myelocystocele may show early rapid expansion causing precipitous deterioration.

Pang et al recommend early repair with resection of the nonfunctional caudal cyst wall, reconstruction of the proximal neural placode, and duraplasty².

1)

Lee JY, Kim SP, Kim SW, Park SH, Choi JW, Phi JH, Kim SK, Pang D, Wang KC. Pathoembryogenesis of terminal myelocystocele: terminal balloon in secondary neurulation of the chick embryo. Childs Nerv Syst. 2013 Sep;29(9):1683-8. doi: 10.1007/s00381-013-2196-3. Epub 2013 Jun 19. PubMed PMID: 23780405.

Pang D, Zovickian J, Lee JY, Moes GS, Wang KC. Terminal myelocystocele: surgical observations and theory of embryogenesis. Neurosurgery. 2012 Jun;70(6):1383-404; discussion 1404-5. doi: 10.1227/NEU.0b013e31824c02c0. PubMed PMID: 22270234.

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