Limited dorsal myeloschisis’ association with dermoids

Sebastian Eibach M.D., FEBNS ¹; Greg Moes M.D. ²; John Zovickian M.D., FACS ²; Dachling Pang M.D., FRCS (C), FRCS (Eng), FACS ²

¹ Paediatric Neurosurgery, Altona Children’s Hospital, Hamburg, Germany
² Paediatric Neurosurgery, Kaiser Foundation Hospitals of Northern California, Oakland, California, USA
Disclosure

Sebastian Eibach M.D., FEBNS
Pediatric Neurosurgery, Altona Children’s Hospital, Hamburg, Germany

• Does not intend to discuss commercial products or services

• Does not intend to discuss non-FDA approved uses of products/providers of services

• No financial disclosure
Normal Embryology:
Neural Tube Formation

Primary Neurulation:
Brain to $S_1$ Cord

Secondary Neurulation:
$S_2$ cord down including Filum

1°Neurulation Malformations must be ABOVE CONUS
Absence of Complete Disjunction

Persistent Cutaneo-Neuroectodermal Tract
- Fibroneural Stalk-joining skin crater to dorsal spinal cord

Incomplete Fusion

Most Important Surface Clue For LDMs

Embryogenesis of LDM

Bridging Squamous Epithelium

Formation of Limited Dorsal Myeloschisis
Primary Neurulation Failure:

Limited Dorsal Myeloschisis (LDM)

Due to Persistent “Limited” Non-disjunction Between Cutaneous Ectoderm & Neuroectoderm
Limited Dorsal Myeloschisis

Non-Saccular

Crater

Pit

Thick Squamous Top

Thin Squamous Top

Dome Pit

Membranous Sac

Saccular

Neural Stalk

Basal Nodule

Stalk to Dome

Segmental Myelocystocele
Typical Lumbar LDM. 11 y/o with motor delay and recurrent UTIs
LDM Stalk: Contiguous Cutaneous and Neuroectoderm

Dermoid cyst

Squamous epithelium (Cutaneous ectoderm)

Glia (Neuroectoderm)
46 y/o status post incomplete LDM surgery at 3 days of age, now presenting with radicular pain and recurrent UTIs.
Large Dermoid Cyst compressing the Spinal Cord at the LDM Merge Point
Histopathology with Glioneuronal Core and Squamous Epithelium
Normal Newborn with Mid-thoracic Skin Bubble:
Saccular LDM. No Pre-Op MRI

Squamous Epithelium Apex

Full thickness Skin
Fibro-glial LDM Stalk with Embedded Hair Follicles

Hair Follicles

Glial Stalk
8-Weeks Post-Op with Dorsal “Scar”
9 Months Later with Intramedullary Lesion & Weak Legs
Intraoperative View 2\textsuperscript{nd} operation

- Engorged Spinal Cord
- Intramedullary Dermoid with Hair
- Exuding Dermoid Content
- Clean Resection Cavity
- Neurulation with 8-0 Nylon
Histopathology
1 Year Later Normal Leg Function
Lessons learned …

- 5/75 LDM cases had associated Dermoid Elements within or contiguous with LDM stalk (6.6 %)

- In 2 patients, incomplete resection of LDM Stalk led to late development of harmful Dermoid Cysts
LDM with Concomitant Dermoid

Always remove entire fibroneural LDM stalk in toto

Always submit entire LDM stalk to pathology
LDM Stalk

Tuft on Cord

Intradural

Extradural