Split notochord syndrome

Split notochord syndrome is an exceedingly rare type of spinal dysraphism. SNS is sometimes associated with other congenital dysraphic defects but, as in our case, the association with spinal cord lipoma, tethered cord, and spinal deformity in the form of spinal column duplication would be exceedingly rare. Herein, the authors report a three-year-old child presented with SNS associated with complex spinal deformity and other associated congenital anomalies. The patient underwent microsurgical release of the tethering element with excellent short- and long-term outcomes.

Clinical presentation: A male newborn with healthy nonconsanguineous parents was born with multiple gastrointestinal and genitourinary anomalies, and duplicated vertebral columns at the lumbosacral area consistent with split notochord syndrome. The patient was initially managed for the gastrointestinal and genitourinary anomalies. As there was no obvious neurological deficit initially, the neurosurgical intervention was postponed till the child reached 30 months of age, when he underwent uneventful release of both spinal cords at their spit point.

Conclusions: SNS is an exceedingly rare developmental anomaly that is usually associated with varying degrees of complex congenital dysraphic defects. Early clinical diagnosis, understanding of the pathophysiology of spinal cord tethering, and microsurgical cord untethering are the important steps in optimal management [1)]

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Alelyani F, Aronyk K, Alghamdi H, Alnaami I. Split Notochord Syndrome with Spinal Column Duplication and Spinal Cord Lipoma: A Case Report. Children (Basel). 2022 Jul 29;9(8):1138. doi: 10.3390/children9081138. PMID: 36010029; PMCID: PMC9406422.