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Diagnostic Image

Split Notochord Syndrome

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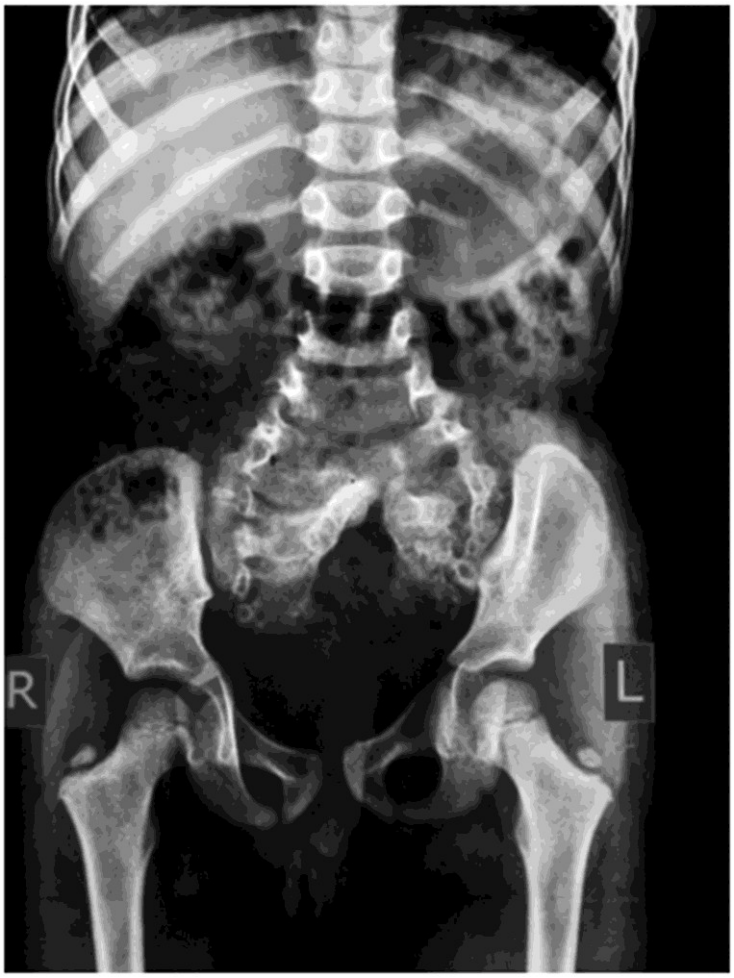

Fig 1. Plain radiograph showing split notochord at the sacrum

Split notochord syndrome is a rare malformation, with only fewer than 35 cases reported in the world literature until 2017.[1] The child depicted herein presented soon after birth with a lesion resembling myelomeningocele in the sacral region. It was a soft, pink, fleshy mass. There was no neurological deficit of lower limbs. During surgical operation, a communication between the dural sac and the rectum was discovered. It was securely closed with 4-0 silk sutures. Dura was closed in routine manner. Patient made uneventful recovery and he continues to remain well.

Split notochord malformation has the following components: (a) A wide defect in the vertebral column; (b) Communication between the neural axis and intestinal tract; and (c) Open neural tube defect externally. Many variations have been described. Neuraxis may communicate with the ileum, cecum, colon or the rectum (as in this patient). There may be associated meningocele, neurenteric cysts or sacral agenesis.

The yolk sac and the amniotic cavity communicate with each other during the third week of gestation. Persistence of this communication can lead to formation of neurenteric canal.

Management of split notochord should be tailored individually according to the nature of anatomical variations. Considering its rarity, wide variations in presentation and heterogenous pathological anatomy, management guidelines do not exist. Surgeon should aim to achieve complete separation of the neural and enteric systems without causing operative neurological damage. Long-term follow-up is essential to detect late sequelae and treat complications.

REFERENCE

[1] Dhawan V, Kapoor K, Singh B, Kochhar S, Sehgal A, Dada R. Split Notochord Syndrome: A Rare Variant. J Pediatr Neurosci. 2017 Apr-Jun; 12(2): 177-179.

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