Lumbar Intramedullary Abscess and Thoracic Syrinx Secondary to Congenital Dermal Sinus: A Rare Case of Paraplegia

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ABSTRACT

Congenital dermal sinus (CDS) results from the failure of neuroectoderm separation from the cutaneous ectoderm during the process of neurulation. Although cutaneous markers are frequently associated with CDS, these patients seek medical attention only after an infectious or neurological complication. Common infections secondary to dermal sinus include meningitis and abscess in subcutaneous, extradural, or subdural region. Intramedullary spinal cord abscess (ISCA) in children is an extremely rare infection which may clinically resemble spinal cord neoplasm. We report a rare case of lumbar ISCA and thoracic syrinx secondary to CDS.

Keywords: Abscess, Dermal sinus, Intramedullary, Paraplegia.

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CASE REPORT

A 2-year-old male child presented with history of intermittent low-grade fever since 2 months. He developed bowel and bladder incontinence with inability to move both the lower limbs since 15 days. His perinatal and developmental history was uneventful, with no history of any significant illness.

On examination, a skin dimpling was noted in the lumbosacral region. Neurological examination revealed flaccid paraplegia with complete sensory loss below D12 for touch, pain, temperature, and pressure. Deep tendon reflexes were absent in both the lower limbs; cremasteric reflexes were absent with bilateral extensor plantar response and relaxed anal tone.

Laboratory investigations revealed elevated white blood cell count (23,100/mm³), elevated erythrocyte sedimentation rate (76 mm/hr), and blood culture report showed methicillin-resistant Staphylococcus aureus.

Magnetic resonance imaging of lumbosacral spine showed absent posterior elements at S1 and S2 level with tethered cord (Fig. 1). Enlarged and edematous cord with intramedullary abscess as noted from L1 to S2 level with a sinus tract extending to the skin from thecal sac at S2 level. Syrinx was noted from D3 to D12 level (Fig. 2).

Fig. 1: Magnetic Resonance Imaging of lumbar spine (T1 contrast image) showing peripheral enhancement of intramedullary lumbar abscess

Fig. 2: Magnetic Resonance Imaging of whole spine (T1 image) showing syrinx from D1 to D12 level and intramedullary abscess from L1 to S1 level
The sinus tract was traced through the subcutaneous tissue, fascia, and was found at the level of S2 entering the dura; posterior elements were absent at S1 and S2 level. L4 and L5 laminectomy and excision of dermal sinus was done. Intramedullary abscess was evacuated and tethered cord was released.

Microbiological analysis of pus samples revealed no microorganisms. Serial cultures were also negative. Histopathology revealed inflammation of dermal sinus. He was treated with broad spectrum antibiotics for 8 weeks. Postoperatively, the child developed spastic tone in lower limbs; however, there was no improvement in power. The child regained sensations in both the lower limbs. His bladder and bowel functions remained altered. Magnetic resonance imaging of spine after 1 month showed overall significant reduction in the size of the conus, evacuation of intramedullary abscess, and excision of dermal sinus.

At 3 months follow-up, patient had persistent loss of bowel and bladder control. He regained sensations below D10, but paraplegia persisted.

DISCUSSION

Congenital dermal sinus (CDS) is a form of dysraphism resulting from a failure of normal midline fusion between the 3rd and 5th weeks of gestation and is found anywhere from the nasion to the coccyx in the midline. Congenital dermal sinus is seen mostly in the lumbosacral region and needs detailed clinical examination for its identification. The approximate incidence of lumbosacral dermal sinus tracts ranges from 1 in 2,500 births.1

Infection in the central nervous system due to a dermal sinus may manifest diffusely as meningitis or focally as epidural, subdural, or parenchymal abscess. Meningitis is the most common and intramedullary spinal cord abscess (ISCA) the rarest complication of a dermal sinus.2 The clinical presentations may vary from incontinence (88%) and paraplegia (72%) to paraparesis (11%). Normal motor functions have also been reported (17%).2 As age advances, the chance of developing neurological deficit increases.3 The commonest causative organism of an abscess associated with CDS is S. aureus.4 The ISCA may be confused with transverse myelitis or Guillian–Barré syndrome or intramedullary tumor in a more chronic disease.

Radiological evaluation is indicated in any patient with CDS presenting with neurological symptoms.2 Magnetic resonance imaging of the spinal cord is the diagnostic procedure of choice. Magnetic resonance imaging enables to identify the extent of the vertebral column involvement; the presence of epidural, subdural, or intramedullary infections; concomitant dermal sinus; epidermoid or intramedullary tumors; and the extent of the spinal cord abnormality.

Treatment includes broad spectrum antimicrobial therapy for 6 to 8 weeks, myelotomy and surgical evacuation of abscess with copious irrigation of saline, and correction of any concomitant abnormalities.4

Early diagnosis and prompt surgical intervention give the best chance of functional neurological recovery. A complete neurological recovery is achieved in only 20% of patients with ISCA.2 Neurological recovery is better in patients if treated surgically within 3 days of severe motor loss. Prophylactic surgery is indicated for CDS to prevent dangerous and recurrent infections of the central nervous system.

In our case, ISCA was involved from L1 to S2. With limited laminectomy and irrigation, good evacuation of abscess was achieved. Neurological recovery was not promising as child presented after 15 days of acute neurological deficits. Many cases of ISCA have been reported so far. What makes this case unique is the presence of ISCA with a dorsal syrinx.

CONCLUSION

The development of ISCA by contamination through the dermal sinus indicates the importance of early excision. Early diagnosis is the key for better prognosis. A complete neurological assessment, radiological investigations, and prophylactic surgery are advised in CDS to prevent the severe morbidity and mortality associated with this condition.

REFERENCES