Lipomeningomyelocele with tethered cord syndrome: an unusual cause of paraparesis in adults

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DESCRIPTION

A 28-year-old lady presented with weakness in both lower limbs for 4 months, urinary and fecal incontinence for 3 months along with sensory loss in the perianal region and below both knees for 3 months. Clinical examination revealed intact higher mental functions and normal cranial nerves. A small paramedian swelling, 10×12 cm in size, soft in consistency was noted in lumbosacral region (figure 1). She had hypotonia in both lower limbs with power being Medical Research Council grade 3 in both hips, grade 2 in both knees and ankles. Tone and power were normal in both upper limbs. Both knee and ankle jerks were absent. Planters were non-elicitable. She had loss of sensations in perianal and saddle distribution along with sensory loss below the knees. Thus, she had features of a cauda-conus syndrome. MRI of lumbosacral spine was done which revealed spina bifida with defect in posterior elements of L3, L4, L5 and S1 vertebral bodies and defect in lumbar fascia. The cord was low lying and tethered anteriorly at L2 level. Syrinx was noted in the distal cord (extending below L2 level) proximal to the tethered site. Herniation of fat, meninges and neural elements was seen through the defect in posterior elements of L3–S1 vertebra forming a cystic swelling in the lumbosacral region (figures 2A–D and 3A,B). No evidence of any cerebrospinal fluid herniation was noted. Skin overlying the swelling was intact. No evidence of any herniation of cerebellar tonsils or descent of brainstem was seen. Thus, lipoma extending from subcutaneous tissue into spinal canal through defects in posterior elements of L1, L2, L3 and S1 vertebrae. The fat is seen as hyperintense on T1-weighted and T2-weighted images (B and C) with signal suppression on Short tau Inversion Recovery sequences (D). Spinal cord is seen tethered at L2 level.

Figure 1 A paramedian soft tissue swelling is seen in the lumbosacral region.

Figure 2 Sagittal section MRI of lumbosacral spine (A–D) showing subcutaneous fatty tissue (lipoma) extending from subcutaneous tissue into spinal canal through defects in posterior elements of L1, L2, L3 and S1 vertebrae. The fat is seen as hyperintense on T1-weighted and T2-weighted images (B and C) with signal suppression on Short tau Inversion Recovery sequences (D). Spinal cord is seen tethered at L2 level.

Figure 3 (A and B) Axial-section MRI at L3 level showing defect in posterior element of the vertebra (spina bifida) with extension of subcutaneous fatty tissue into spinal canal with entrapment of nerve roots.
on the basis of typical imaging features, diagnosis of a lipomeningomyelocele with a tethered cord was made. Patient was subjected to neurosurgery for resection of the lipoma and detethering of spinal cord. Unfortunately, the neurodeficit was persistent at 3 months of follow-up.

Tethered cord syndrome (TCS) is characterised by neurodeficit due to restraint of spinal cord as a result of anatomical, physiological or developmental causes. The developmental causes include spinal cord lipoma, lipomeningomyelocele or sacral agenesis. The median age of the presentation of lipomyelomeningocele has been observed to be 5.5 years. Further, Kang et al observed that most of the patients with TCS presented at age less than 6 years. The presentation of lipomeningomyelocele with paraparesis in adulthood in our case is quite unusual. The cutaneous markers of spinal dysraphism include a tuft of hair, subcutaneous lipoma, swelling, dermal sinus or a dimple. As this condition is surgically curable, such patients should be evaluated with neuroimaging to decide further course of management.

Learning points

▸ Lipomeningomyelocele is an important cause of tethered cord syndrome.
▸ Though unusual, presentation of lipomeningomyelocele with neurodeficit in adults is possible.
▸ All young patients with paraparesis or quadriparesis should be clinically examined for cutaneous markers of spinal dysraphism.

Competing interests None.
Patient consent Obtained.

REFERENCES