Split Cord Malformation (SCM): Case Report of a Rare Disease and Its Operative Repair

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Introduction

Split cord malformation (SCM) [2], is a rare congenital condition where there is splitting of the spinal cord. Each semicord is surrounded by a complete thecal sac and meningeal covering. In type I SCM, the two semicords are separated by a bony spur, and they usually join below the bony spur level forming a normal spinal cord segment. In type II SSCM, the two semicords are contained in a single dura sac. The overall incidence of type I SSCM is low, and it only represents 3.8 – 5% of all spinal cord anomalies. We report a baby with type I SCM. He underwent untethering operation at age 3 months.

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Case Report

A full-term Chinese female baby born with unremarkable antenatal scan and tests by uncomplicated spontaneous vaginal delivery was found to have thoracolumbar hypertrichosis **(fig 1),** as well as a sacral dimple with no deviation of gluteal cleft.

Subsequent ultrasound **(fig 2),** MRI and CT spine **(fig 3-7)** revealed type I SCM at T10 to T12 levels, and mild hydromyelia below this level. The conus was at L4 and there was a fatty thicken filum and a small lipoma at posterior element at S2 level **(fig 8)**.

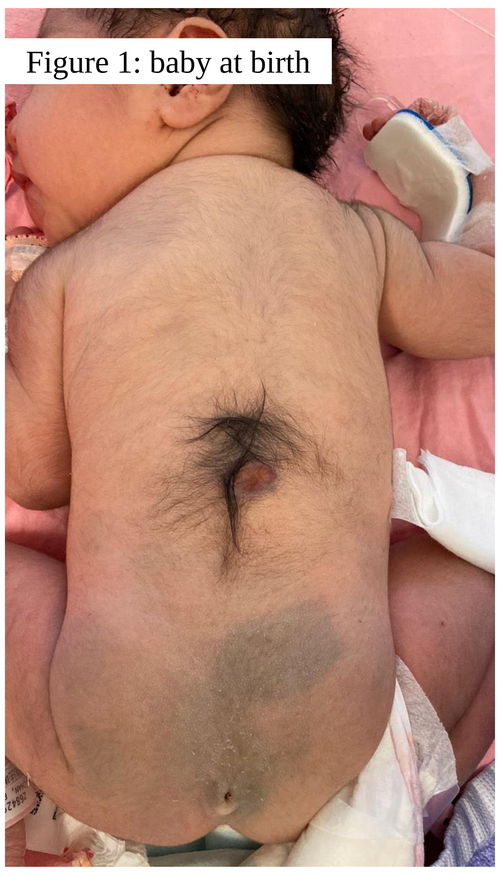

Figure 1: baby at birth

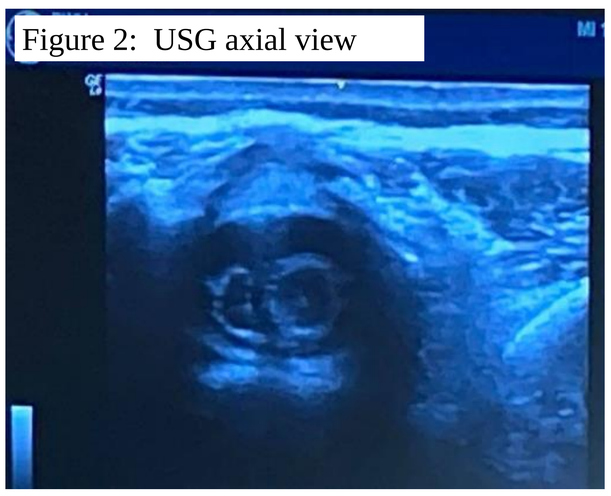

Figure 2: USG axial view

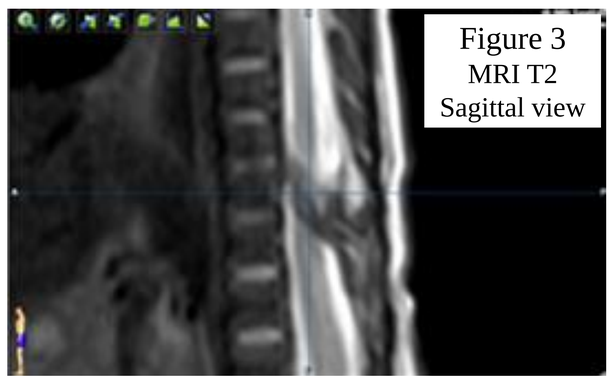

Figure 3
MRI T2
Sagittal view

Physical examination revealed normal limb tone, movement and tendon reflex. There was mild claw toes and equinovarus of the left foot.
MRI brain, ultrasound urinary system and axial skeleton was unremarkable.

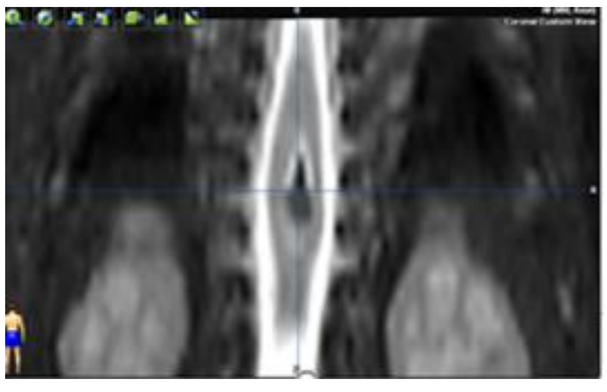

Figure 4: MRI T2 coronal view

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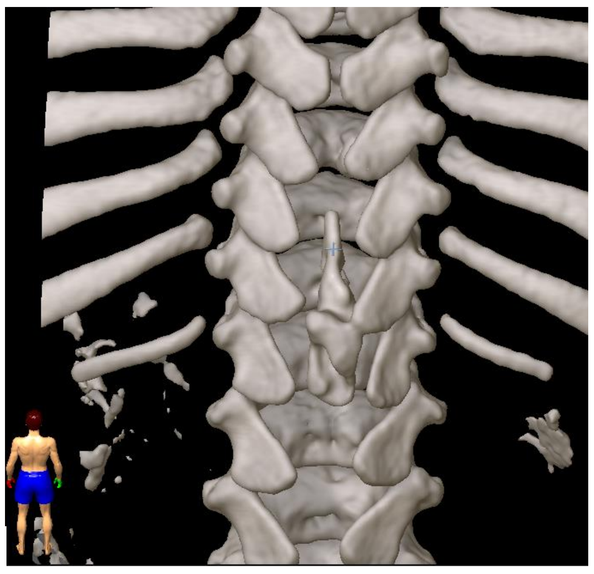

Fig 5: CT 3T reconstruction

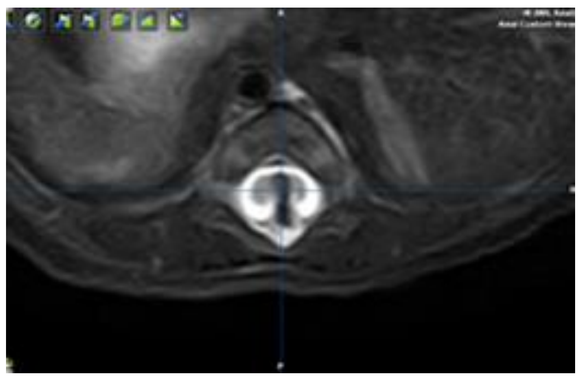

Fig 6: MRI T2 axial view

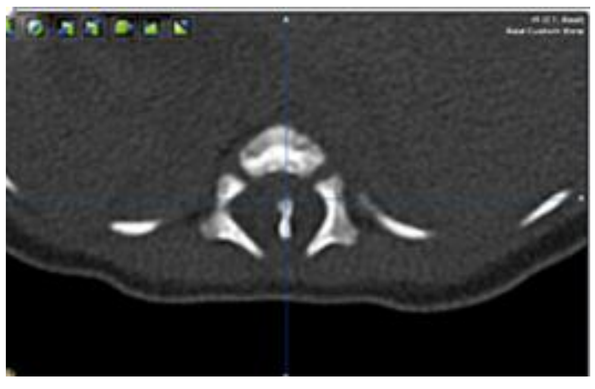

Fig 7: CT axial view

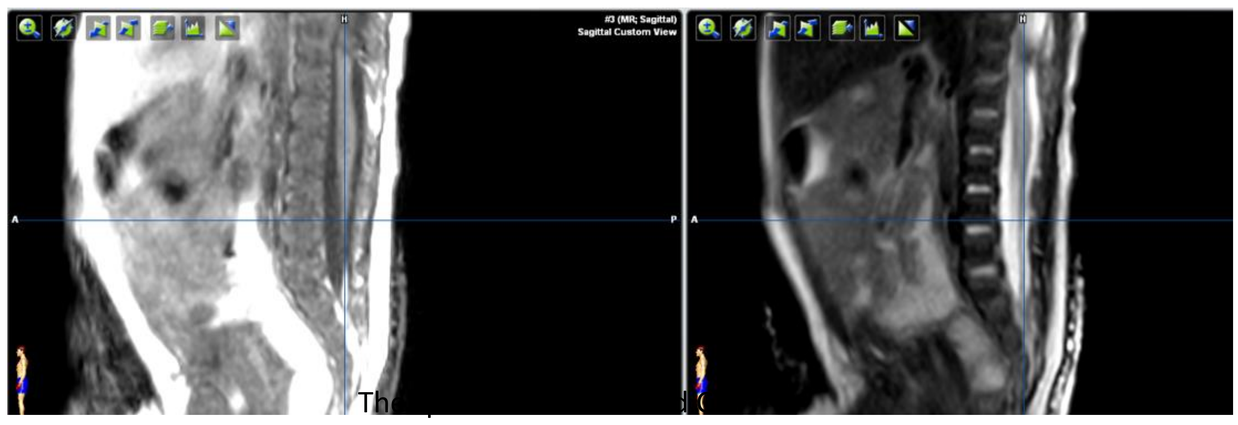

Fig 8: MRI T1 (left) and T2 right) sagittal view showing the fatty thicken filume.

Operation

T10-L1 laminectomy, excision of bony spur and the medial dural of the split cord for untethering of the cord was performed at 75 days of age under intra-operative monitoring with somatosensory evoked potential (SSEP) and triggered EMG for thelower limbs and anal sphincter.
During the same session, untethering of cord and incise of the fatty hilum was also performed at a lower level (L5-S1) through a separate incision.

SSEP based line showed the left lower limb was 1/3 lower than the right side. This was consistent with the subtle claw toes and equinovarus of left foot. Also consistent with that the left cord was the small one. SEEP remained stable and no deterioration through out the operation. Post-operatively the extent of equinovarus appeared similar to pre-operative status.

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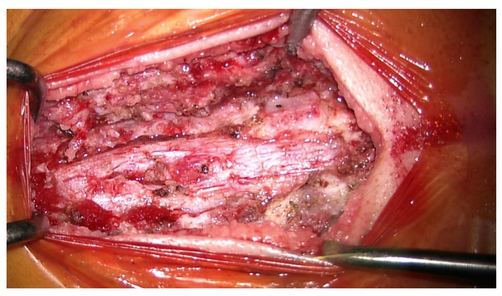

Fig 9: on exposure of the dura, the spit cords were seen separated by a bone spur.

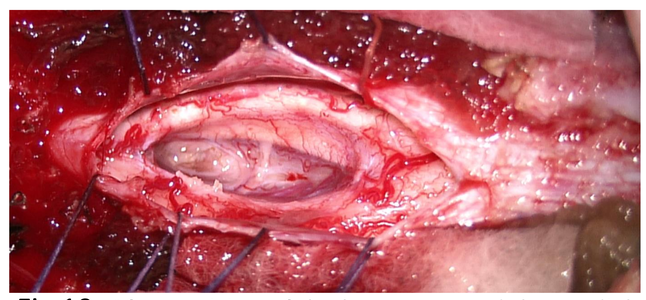

Fig 10: After excision of the bone spur and the medial dura, before dural closure.

Discussion

Type I SCM is a cord tethering lesion and likely to cause neurological damage from its natural history. Patient tend to present with sensorimotor deficits and pain later on in life and surgical treatment is often required. For those receiving surgery, the outcome was excellent and symptoms are greatly improved. Although some may require multiple re-operation, symptoms showed improvement consistently after operation. Untethering operation is recommended in children even asymptomatic[2]. Furthermore, strong correlation was observed in low-lying conus and other cord tethering lesion including thickened filum and lipoma, was both present in our patient. Untethering of both lesions were recommended at during the same operation[2]. The indication of untethering operation for SCM remains an active topic in the International Society of Pediatric Neurosurgery[3]

In our patient, surgery was offered early (less than 3 months of age) for the possibility of neurological deficit. In small children, the symptoms can be subtle and the cord tethering can be important for the developing body thus early surgery should be considered. Operation in paediatric patients lead to less re-tethering compared to adults[1]. Yet, the decision to offer early untethering operation needs to consider both the presence of possible neurological deficit and surgical safety in such a small baby. It was reported that overall surgical morbidity was slightly higher with type I lesion compared with type II[2].Surgical technique, intra-operative neurophysiological monitoring and the roll of an experienced paediatric neuroanaesthetist cannot be over emphasized.

Conclusion

The timing of operation and a unified management plan for SCM is difficult to formulate due to its rarity. Decision should be made individually considering both the symptoms, the age and the capacity of the institutes. Our experience is consistent with the world literature.

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References

1. Andrew J. Kobets et.al, Split cord malformation and tethered cord syndrome: case series with long-term follow-up and literature review, Child's Nervous System (2021) 37:1301–1306

2. Pang, Dachling et.al, Split Cord Malformation, Neurosurgery 31; 481-500, 1992

3. ISPN Clash of the Titans – Prophylactic surgery is indicated for split cord malformation 2021Oct22

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