

Spinal dysraphism and cavovarus foot deformity: a case report

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Neurological impairment secondary to spinal dysraphism most commonly presents as unilateral cavovarus foot in children. The deformity usually develops in the growing child around the age of five or six. The presence of a cavovarus foot of unknown origin in a child should lead to a complete neurological examination, including an assessment of the spine for spinal dysraphism. The early recognition of pathology may prevent severe neurological sequelae. A case of lipomyelomeningocele is presented to illustrate that cord damage in children with spinal dysraphism can present initially as a cavovarus foot. (JCCA 1992; 36(2):91-95)

KEY WORDS: spinal dysraphism, lipomyelomeningocele, cavovarus foot.

L'altération neurologique la plus fréquente des suites d'une dysraphie vertébrale se présente sous la forme d'un pied cavovarus chez l'enfant. La difformité se développe généralement chez un enfant de 5 ou 6 ans, autrement en santé. Chez les enfants, la présence d'un pied cavovarus d'origine inconnue devrait toujours conduire à un examen neurologique complet, incluant la recherche d'une dysraphie au niveau de la colonne vertébrale. Une identification rapide de cette pathologie peut prévenir des séquelles neurologiques sévères. Pour illustrer ceci, un cas de lipomyélingocèle est présenté. Vous y verrez comment un simple cas d'enfant avec un pied cavovarus peut vous mettre sur la piste d'une dysraphie vertébrale, avec endommagement de la moelle épinière. (JCCA 1992; 36(2):91-95)

MOTS CLÉS : dysraphie vertébrale (fermeture incomplète du tube neural), lipomyélingocèle.

Introduction

Young patients presenting with new or progressive foot deformities should be thoroughly investigated. Such investigation will often show a congenital anomaly of the spinal cord or cauda equina. In fact, spinal dysraphism is considered the most common cause of cavovarus foot after Charcot-Marie Tooth disease.¹

Spinal dysraphism is the name given to a group of spinal anomalies occurring secondary to abnormal closure of the developing neural tube in the embryo. It most commonly refers to anomalies like spina bifida, with or without meningocele or myelomeningocele, and diastematomyelia, but also includes

the rarer intra- or extra-dural lipomas and dermoid cyst.² If left untreated, these disorders can cause neurological damage in the growing child secondary to traction, compression or tethering of the spinal cord. The sequelae may range from minor back pain to paraplegia, but the speed and amount of deterioration is variable. The first symptoms usually appear around four to six years of age, but they may be present in the newborn or appear in the adult. While the first symptom is ordinarily urinary incontinence in the adult, the child will present with a unilateral cavovarus foot.

Pes cavus refers to the pathologic elevation of the longitudinal arch in an otherwise normal foot. When this cavus foot assumes a varus position during weight bearing, it is referred to as cavovarus¹ (Figure 1). The deformity can be idiopathic or seen in infancy as a component or residual of clubfoot. However, it usually develops insidiously, secondary to occult neuromuscular disorders.³ Brewerton studied 77 patients with pes cavus and found that 75% had underlying neurologic disease based on the examination, electromyography or nerve conduction studies.⁴ With neuromuscular disorders such as spinal dysraphism, the foot deformity is believed to develop secondary to muscular imbalance, but its exact pathogenesis remains a mystery.⁵

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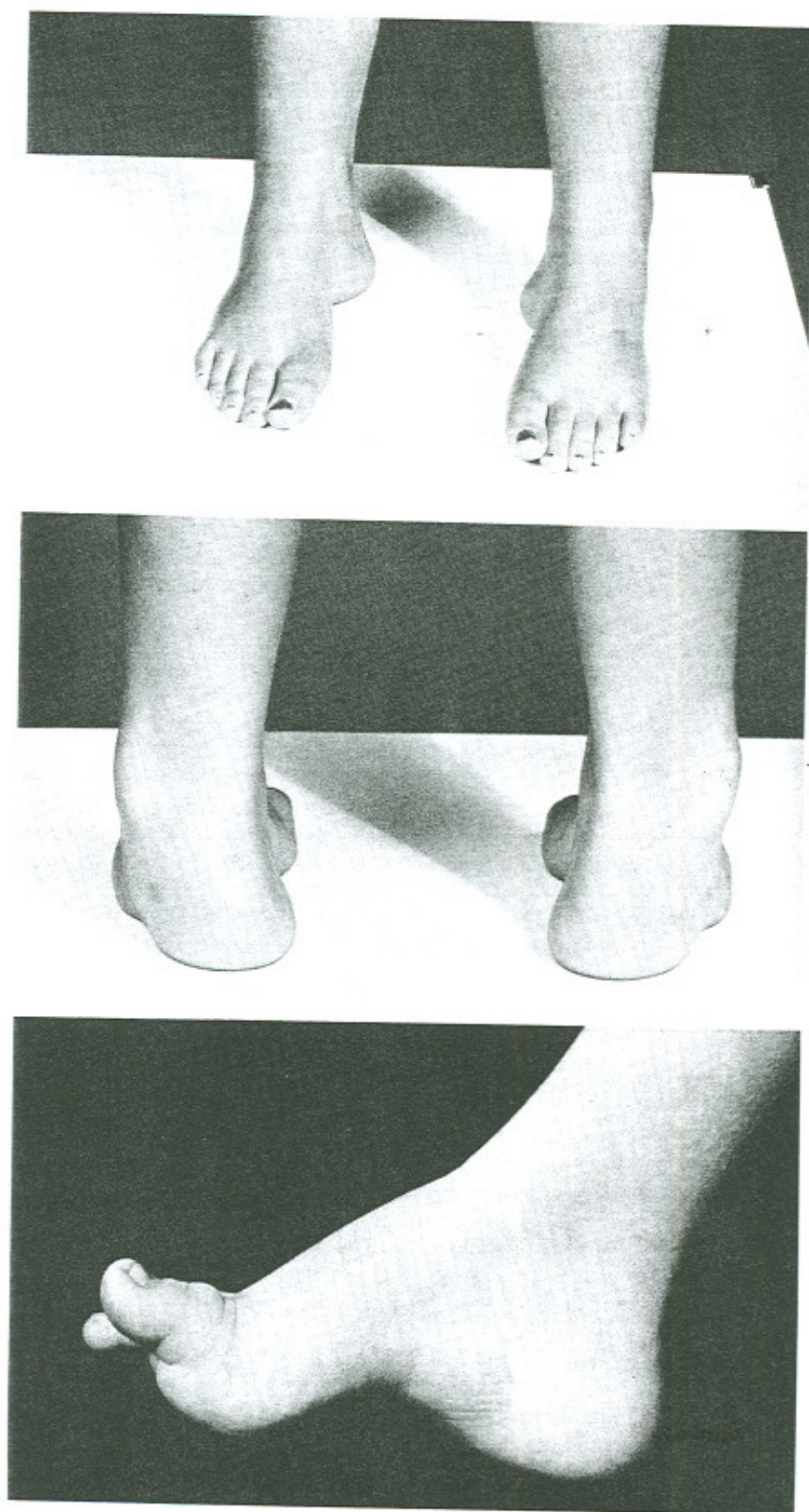


Figure 1 (a, b and c) Anterior, posterior and lateral photographs of cavovarus feet showing the elevated medial longitudinal arch and the varus position of the hind foot.

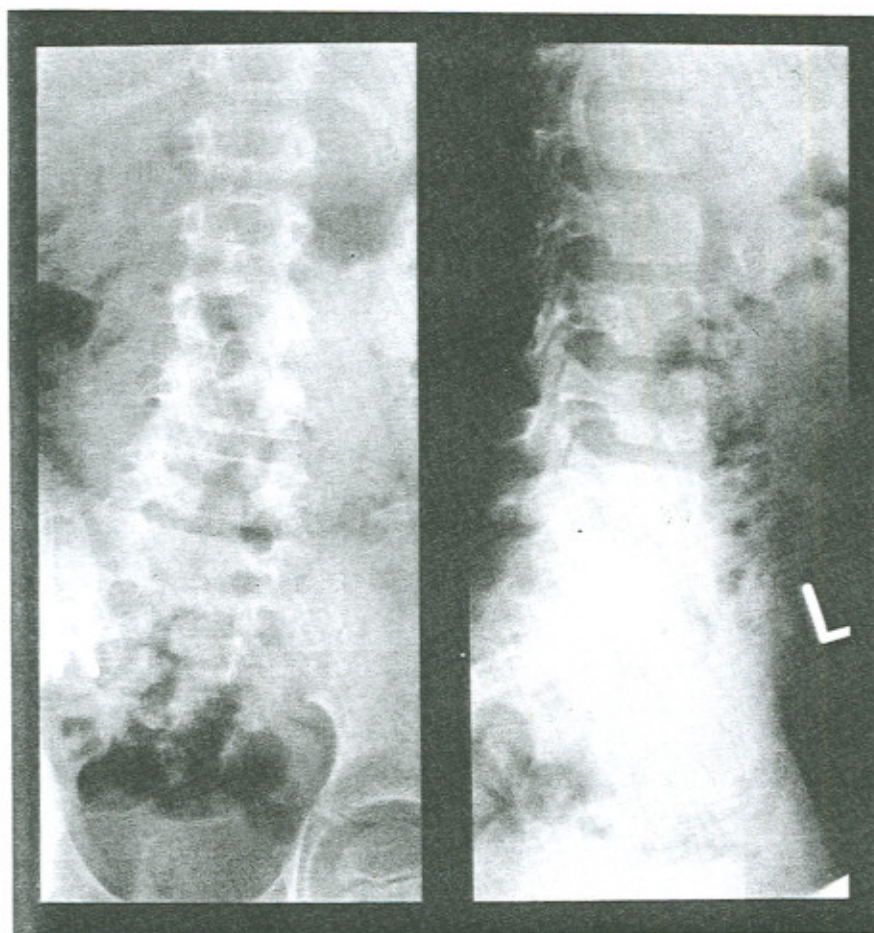


Figure 2 The anteroposterior and lateral lumbar spine radiographs show a large spina bifida from the fifth lumbar to the last sacral vertebrae. There is associated widening of the interpedicular distance.

Case report

A 6-year-old boy, was seen in Orthopaedic Outpatients in early 1988 because his mother noticed a deformity of his left foot that had developed over the last year. She felt that the child's foot was smaller, kept turning in, and that he was tripping frequently while walking and running.

He was the product of a normal pregnancy and delivery. The only anomaly recognized at birth was a hairy spot at the lumbosacral level associated with a spina bifida. Still, this was not investigated further because of the absence of any associated anomaly. He met the normal developmental milestones. A thorough neurological history revealed occasional faecal and urinary soiling, as well as urinary urgency over the last year. No family history of neurological disease was present.

Examination revealed a fit looking young boy, well developed for his age. The deformity of the lower limbs was limited to his left foot. The foot had a normal size, but a cavovarus deformity was present. The deformity was flexible and could be corrected passively. The neurological examination showed an

absent ankle jerk with very weak ankle evertors, but no sensory abnormality was detected. The muscle tone was normal and the upper limbs were neurologically intact. Examination of the spine revealed a hairy spot associated with a dimple at the first sacral level and a palpable defect in the bony arch.

A radiographic study of the lumbar spine confirmed the presence of a large defect in the posterior elements, from the fourth lumbar segment to the distal sacrum, associated with widening of the interpedicular distance (Figure 2).

The foot deformity and neurological problems were suspected to be associated with the spina bifida. An ankle/foot orthosis was then prescribed to prevent progression of the cavovarus foot and the child was referred for a neurosurgical opinion.

Neurosurgical consultation disclosed the additional examination findings of left gastrocnemius atrophy and saddle anaesthesia. A myelogram revealed the presence of a widened thecal sac associated with a tethered cord and a posterior lipoma located at the fifth lumbar level (Figure 3). These findings were confirmed by a CT scan done the same day (Figure 4). A spinal ultrasound

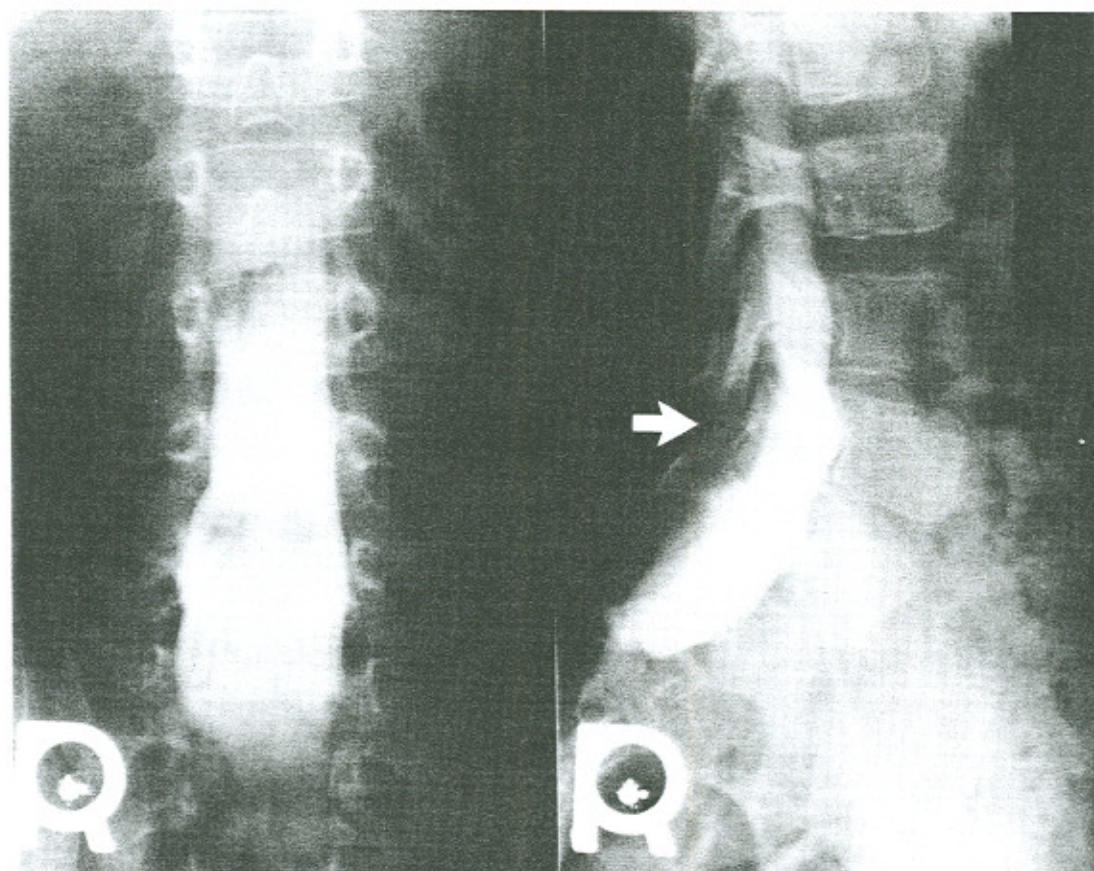


Figure 3 An anteroposterior and lateral myelographic view of the lower cord shows a widened thecal sac with a posteriorly located lipoma and a tethered cord (arrow).

showed a diffusely ectatic spinal canal and the cord tethering. A voiding cystourethrogram showed a normal bladder, anatomically and functionally.

The patient underwent surgery during the summer of 1988. The procedure released his tethered cord and excised his lipomyelomeningocele. At the two-month follow-up, his incontinence was very much improved, but the foot deformity was still evident. In early 1989, seven months after his spinal surgery, he underwent a second operation to correct the cavovarus deformity. The cavovarus deformity had become more rigid and the foot appeared smaller. This surgery consisted of a posteromedial soft-tissue release and a split tibialis anterior transfer in his left foot. He was also instructed to continue wearing his foot splint for the year following the surgery.

Four months after his foot surgery, eleven months after his spinal surgery, he was continent and could walk and run without difficulty.

Discussion

Traditionally, spinal dysraphism is suspected in patients with external skin manifestations such as subcutaneous lipomas, hemangiomas, dermoid cysts and hair growth. However, one study of 24 subjects with spinal dysraphism found that only 13 (54%) presented with an external skin manifestation. The others (46%) were diagnosed by observation and examination of the

legs and feet.⁶

Most commonly, a child with a progressive spinal cord lesion secondary to spinal dysraphism will present with an established, but slight, cavovarus deformity of one foot.⁶ The affected leg may be shorter than the other. The parents will often notice a change in gait. Frequently, no other abnormality will be present. Still, one should do a complete neurological examination of the lower limb in these patients. In severe cases, the cord

Table 1
Common neurological conditions associated with cavovarus foot deformity

Charcot-Marie tooth disease*
Spinal dysraphism**
Freidreich's ataxia
Cerebral palsy
Poliomyelitis
Spinal muscular atrophy
Syringomyelia
Hereditary peripheral neuropathy
Traumatic peripheral nerve injury

* Most common. **Second most common.

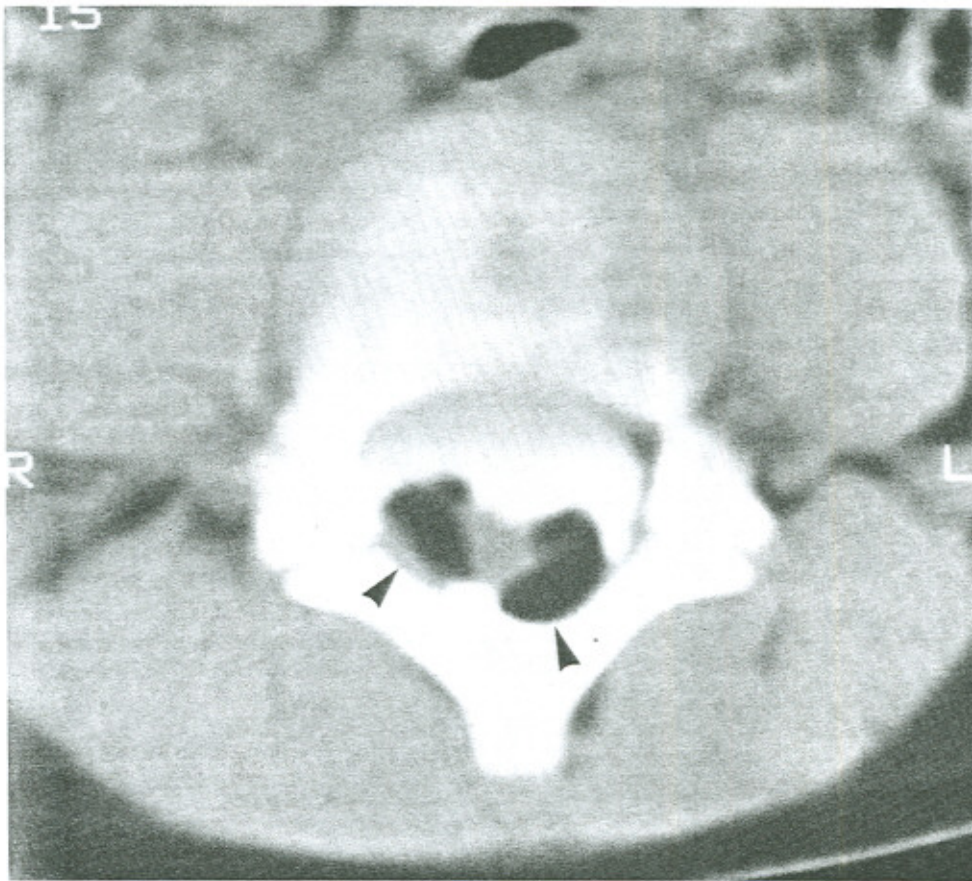


Figure 4 An axial CT scan through the fifth lumbar level clearly shows the posteriorly located lipoma (arrows).

injury will cause sensory, motor and reflex changes. It can even lead to trophic skin changes, incontinence and signs of paraplegia. The examination should help exclude other neurological diseases capable of causing a cavovarus foot (Table 1).

Radiographic examination of the lumbar spine is essential in these patients. Spina bifida occulta is very common and usually clinically insignificant when a simple split in the spinous process is present. However, a large defect is often associated with abnormal development of the spinal cord. Further investigations should include more sophisticated spinal and neurological studies such as a myelogram, CT scan, MRI, electromyograph and nerve conduction studies.

If recognized early, surgical intervention will often minimize the neurological sequelae. Once the cause of the foot deformity has been addressed, it then becomes possible to treat the cavovarus deformity. This treatment will usually include splinting and/or operative correction.

Conclusions

This case illustrates that progressive neurological damage secondary to spinal dysraphism can present as a cavovarus foot deformity. Some of these patients can present with low back pain. It is important to rule out associated spinal and neurological anomalies to prevent irreversible spinal cord damage and disability.

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