Journal of Taibah University Medical Sciences (2019) 14(5), 472–476

Taibah University

Journal of Taibah University Medical Sciences

www.sciencedirect.com

Case Report

Meningomyelocele with unusual feet deformity combination: A report of a rare case

Ammar K. Alomran, SB ORTHO [a], Munirah A. Abahussain, MBBS [b,*], Asma A. Aldossary, MBBS [b] and Isra B. Alshammari, MBBS [b]

[a] *Department of Orthopaedic Surgery, College of Medicine, Imam Abdulrahman Bin Faisal University, Dammam, KSA*
[b] *College of Medicine, King Fahd Hospital of the University, Al Khobar, KSA*

Received 21 April 2019; revised 29 July 2019; accepted 31 July 2019; Available online 27 September 2019

الملخص

راجع طفل عمره ١٩ شهرا عيادة جراحة العظام للأطفال عندما كان عمره ٦ أشهر يعاني من قيلة نخاعية سحائية وتشوه في القدمين. تم تنويم المريض في وحدة العناية المركزة لحديثي الولادة بسبب التشوهات الخلقية المتعددة. هذه التشوهات تضمنت قيلة نخاعية سحائية، وقدم فحجاء حنفاء يمنى، وكاحل عمودي أيسر، وشرج أرتق وفتق لا مباشر. أكدت الأشعة تشخيص القدم الفحجاء الحنفاء اليمنى والكاحل العمودي الأيسر. تأخر العلاج بسبب تأخير التشخيص. في عمر ٩ أشهر، بدأ المريض على معالجة متسلسلة باستخدام الجبس بطريقة بونستي للقدم اليمنى وطريقة بونستي العكسية للقدم اليسرى. تم عمل الجبائر أسبوعيا لمدة ١٢ أسبوعا. اكتسبت القدم اليمنى الوضع الطبيعي قبل اليسرى، ولكن قررنا إبقاءها في الجبيرة لحين عمل التصحيح الجراحي للقدمين. خضع المريض لبضع وتر العرقوب الأيمن والجبيرة، وبضع وتر العرقوب الأيسر والمعالجة ورد للمفصل الكاحلي الزورقي وتثبيت بأسلاك كيرشنر مع الجبيرة. استمرت الجبائر لمدة ٣و٦ أسابيع (القدم الفحجاء الحنفاء والكاحل العمودي على التوالي). وكان الهدف النهائي للعلاج هو الحصول على قدم قابلة لاستعمال الدعامة باستخدام جبيرة مبطنة جيدا للركبة والكاحل والقدم لتحسين جودة الحياة.

الكلمات المفتاحية: القدم الفحجاء الحنفاء؛ قيلة نخاعية سحائية؛ حنف قفدي فحجي؛ كاحل عمودي

Abstract

A 19-month-old male infant had presented to a paediatric orthopaedic clinic at the age of 6 months with meningomyelocele and bilateral feet deformity. The patient was admitted to the neonatal intensive care unit because of multiple congenital anomalies. These anomalies included meningomyelocele, right clubfoot, left vertical talus, imperforated anus, and an indirect hernia. Radiographs confirmed the diagnosis of right clubfoot and left-sided vertical talus. The course of management was delayed because of late diagnosis. At the age of 9 months, the patient underwent serial casting using the Ponseti and reverse Ponseti techniques for right clubfoot and left-sided vertical talus, respectively. The casting was performed weekly for 12 weeks. The right foot gained normal position before the left, but we decided to keep it in the cast until surgical correction was performed for both feet. The patient underwent right Achilles tendon tenotomy and casting and left Achilles tendon tenotomy, manipulation, and talonavicular reduction and k-wire fixation with casting. The casts remained for 3 and 6 weeks (clubfoot and vertical talus, respectively). The ultimate goal of the treatment was to produce braceable, plantigrade feet with the use of a well-padded knee ankle–foot orthosis to improve the quality of life.

Keywords: Clubfoot; Meningomyelocele; Talipes equinovarus; Vertical talus

* Corresponding address: Imam Abdulrahman Bin Faisal University, Dammam 31441, P.O. Box: 1982, KSA.
E-mail: muniraabahussain@gmail.com (M.A. Abahussain)
Peer review under responsibility of Taibah University.

Production and hosting by Elsevier

Introduction

Talipes equinovarus or clubfoot is a complex developmental deformity characterized by four main features: cavus,

1658-3612 https://doi.org/10.1016/j.jtumed.2019.07.004

adduction, varus, and equinus.[1] A recent systemic review examining the birth prevalence of clubfoot in low-income countries, such as India, found that the prevalence was 1.19 per 1000 births.[2] Clubfoot associated with myelomeningocele (MMC) varies, including idiopathic clubfoot wherein the foot is severely rigid and less responsive to treatment.[3]

Eighty percent of patients with MMC present with orthopaedic problems.[4] Most children with spina bifida develop foot and ankle deformities, the most common being clubfoot and vertical talus.[5] About 32% of patients with MMC present with Talipes equinovarus, and surgical correction is required in most cases.[3]

Vertical talus is severe flatfoot with inflexible malalignment of the hindfoot and midfoot in which the talus is nearly vertical with the hindfoot demonstrating equinovalgus, and the navicular is dislocated on the head of the talus.[3] Vertical talus has an incidence of 1 in 10,000 births with an equal distribution among males and females.[6] In half of the cases, vertical talus presents as an idiopathic deformity, while in the other half, it is accompanied by a neuromuscular or genetic disorder, such as MMC.[6]

Although 80–95% of patients with MMC develop foot problems, a case of MMC with both clubfoot and vertical talus has not yet been reported.[7] Considering the rarity of this presentation, we report this case to improve our understanding of the multiple foot defects in patients with MMC.

Figure 1: Clinical photograph showing right clubfoot and left rocker bottom deformity due to vertical talus.

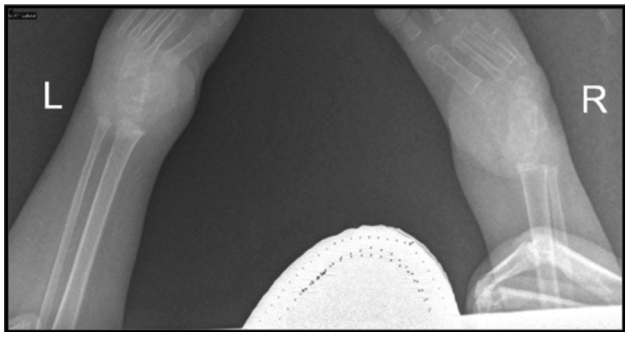

Figure 2: AP view of both feet showing reduced talocalcaneal (TC) angle on the right side with negative talus: first metatarsal angle and increased TC angle on the left side.

Case summary

A 19-month-old male infant was referred by a paediatrician to a paediatric orthopaedic clinic at the age of 6 months as a case of MMC with bilateral feet deformity. The baby was a product of consanguineous marriage and was born by an elective Caesarean section at 38 weeks of gestation with birthweight of 2.4 kg. The mother underwent regular antenatal follow up and began receiving folic acid supplements in the 1st trimester. In the 3rd trimester, the mother was diagnosed with polyhydramnios. The patient was admitted to the neonatal intensive care unit (NICU) because of multiple congenital anomalies. These anomalies included MMC Level L1, right clubfoot, left vertical talus, imperforated anus, and an indirect hernia.

Furthermore, due to the co-occurrence of these multiple congenital anomalies, the patient was diagnosed with VACTERL. Two of the patient's elder siblings had cleft palates. There was no other family history of congenital anomalies. Examination showed a right rigid clubfoot and a left foot with rigid calcaneovalgus and rocker bottom deformities (Figure 1). There was no movement or sensation in both feet. A scar from the MMC surgical repair was noted on the back.

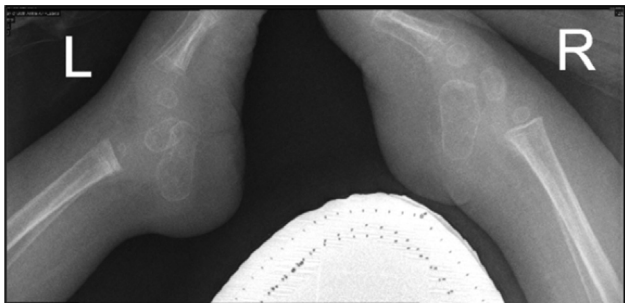

Figure 3: Lateral X-rays of both ankles showing the difference in talocalcaneal (TC) angle with right-sided parallel TC angle and left-sided increased TC angle. There is left vertical talus and dorsal dislocation of the navicular and right-sided cavus.

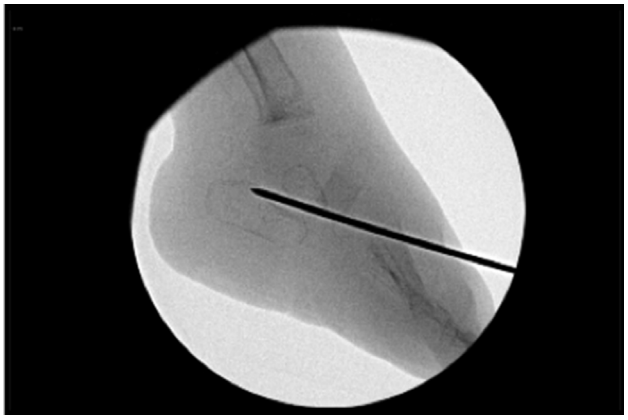

Figure 4: Intraoperative lateral X-ray after reduction of talonavicular joint and pinning.

Radiographs confirmed the diagnosis of right clubfoot and left-sided vertical talus (Figures 2 and 3). The course of management was delayed because of the delay in diagnosis. At 9 months, the patient underwent serial casting using the Ponseti and reverse Ponseti techniques for right clubfoot and left-sided vertical talus, respectively. The casting was performed weekly for 12 weeks. The right foot was corrected before the left, but we elected to keep it in cast until surgery was performed for both feet.

The patient underwent right tendon Achilles tenotomy and casting and left tendon Achilles tenotomy, manipulation, and talonavicular reduction and k-wire fixation with casting (Figure 4). The casts remained for 3 and 6 weeks (clubfoot and vertical talus, respectively). The ultimate goal of the treatment was to produce braceable plantigrade feet (Figures 5 and 6) with the use of well-padded knee–ankle–foot orthosis with neutral ankle and plantigrade feet to improve his quality of life (Figure 7). Table 1 summarises the Pirani scores of the patient before and after management.

Discussion

MMC results from failure of neural tube or mesenchymal closure in the early embryonic period, which leads to exposure of the spinal cord and meninges.[8] A study conducted at Asir Central Hospital (ACH) between 1995 and 1998 showed that 6.6% of all admissions to the NICU were for neural tube defects; 70% of these neural defects were MMC. The most commonly affected site was the thoracolumbar region (44.4%), followed by the lumbosacral region (40%).[9]

According to the study conducted at ACH, the most common associated orthopaedic anomaly was talipes equinovarus, followed by dislocation of the hip.[9] The aetiologies of these orthopaedic deformities were classified as congenital, such as hip dislocation, clubfoot, and vertical talus, or acquired due to paralysis or muscle imbalance.[10] Most patients with spina bifida will present with other foot and ankle deformities; these include clubfoot, equine, vertical talus, and ankle valgus.[7] The cosmetic appearance caused by these deformities is not the only problem; patients may develop skin irritation and experience difficulties in ambulation.[3]

A study conducted between 2010 and 2014 showed that most patients with spina bifida and sacral injury have normal feet, patients with high-level lumbar injury (L1–L2) commonly have isolated equinus and clubfoot deformities, and Pes cavus deformities mostly present in patients with low-level lumbar injury (L3–L5).[5]

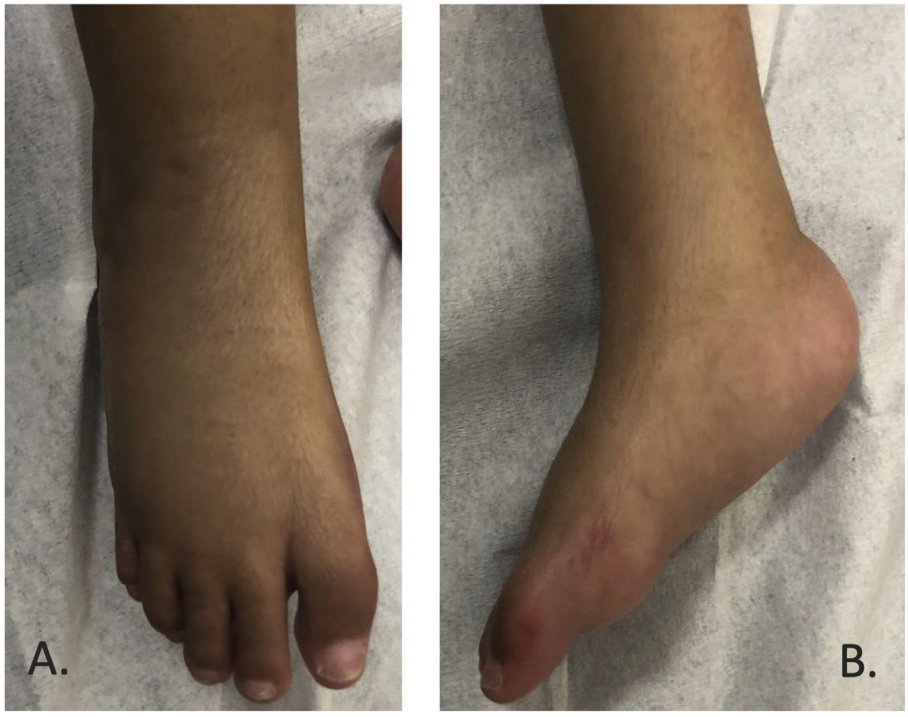

Figure 5: A. Anterior view of a braceable plantigrade right foot. B. Lateral view of a braceable plantigrade right foot.

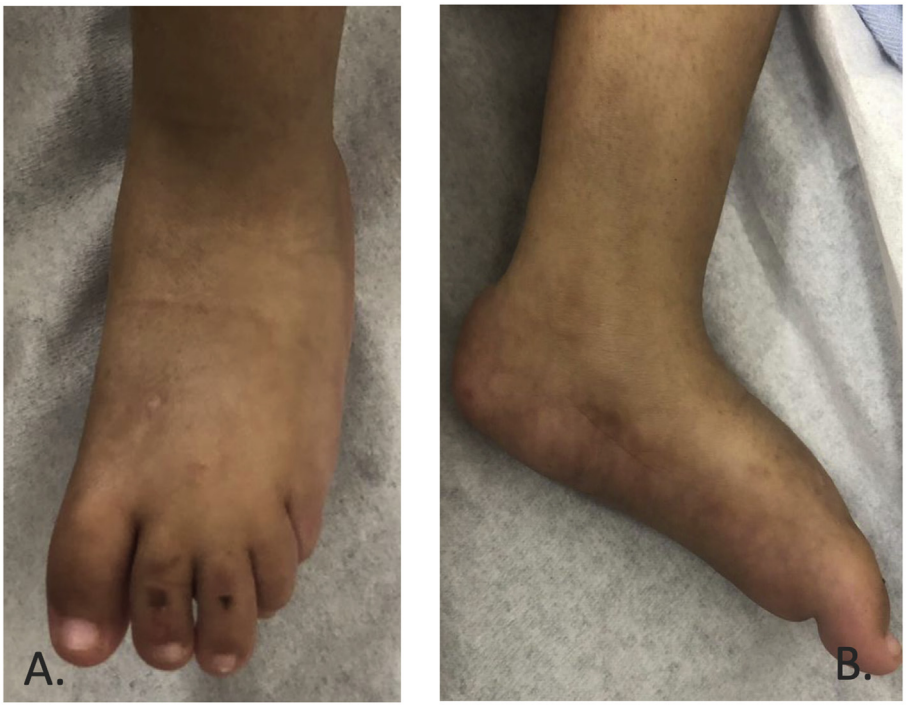

Figure 6: A. Anterior view of a braceable plantigrade left foot. B. Lateral view of a braceable plantigrade left foot.

Talipes equinovarus in patients with MMC is known to be more resistant and rigid; traditionally, extensive soft tissue release is thus performed in these patients. There are reported cases of early results after implementing the Ponseti method of serial manipulation and casting in clubfoot with spina bifida. Some complications arise when using this method, such as skin irritations and a high recurrence rate. For these reasons, it is recommended to perform at least a 1-cm excision in an Achilles tendon to avoid the recurrence of equines. It is also helpful to use ankle–foot orthosis, which helps avoid fractures and skin breakdown.[3]

Table 1: showing Pirani score pre/post management.

	Pre-Management	Post-Management
Curved Lateral Border	1.0	0.0
Medial Crease	1.0	0.0
Talar head coverage	0.5	0.0
Posterior Crease	1.0	0.0
Rigid Equines	1.0	0.0
Empty heel	1.0	0.0

Figure 7: Well-padded ankle–foot orthosis with neutral ankle and plantigrade feet.

On the other hand, vertical talus treatment helps provide a plantigrade weight-bearing surface by re-establishing the normal anatomic relationships between the talus, navicular, and calcaneus.[11] The traditional and most common technique used to treat congenital vertical talus is extensive soft-tissue release surgery when the patient is between 10 and 12 months of age. However, a less invasive method of treatment includes open talonavicular pin fixation and percutaneous tenotomy of the Achilles tendon subsequent to serial manipulation and casting. Slow correction and adequate padding are essential, similar to that in clubfoot serial casting.[3]

Source of funding

None.

Conflict of interest

The authors have no conflict of interest to declare.

Ethical approval

The researchers received ethical clearance and approval from the institutional review board of Imam Abdulrahman Bin Faisal University (number IR3 2018- 01–332).

Consent

An approved waiver of consent for photography and publication was obtained from the legal guardian.

Authors contributions

AKA was involved in gaining ethical approval, providing research materials, patient recruitment and management, obtaining photographs, and revising and editing the manuscript. MAA and AAA obtained consent, reviewed the literature, and wrote the first draft of manuscript. IBA obtained patient data and case details and wrote the case summary. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

References

1. Besselaar A, Sakkers R, Schuppers H, Witbreuk M, Zeegers E, Visser J, et al. Guideline on the diagnosis and treatment of primary idiopathic clubfoot. **Acta Orthop 2017**; 88(3): 305–309.
2. Smythe T, Kuper H, Macleod D, Foster A, Lavy C. Birth prevalence of congenital talipes equinovarus in low- and middle-income countries: a systematic review and meta-analysis. **Trop Med Int Health 2017**; 22(3): 269–285.
3. Swaroop V, Dias L. Orthopaedic management of spina bifida—part II: foot and ankle deformities. **J Child's Orthop 2011**; 5(6): 403–414.
4. Beuriat P, Poirot I, Hameury F, Szathmari A, Rousselle C, Sabatier I, et al. Postnatal management of myelomeningocele: outcome with a multidisciplinary team experience. **World Neurosurg 2018**; 110: e24–e31.
5. Gunay H, Sozbilen M, Gurbuz Y, Altinisik M, Buyukata B. Incidence and type of foot deformities in patients with spina bifida according to level of lesion. **Child's Nerv Syst 2015**; 32(2): 315–319.
6. Mckie J, Radomisli T. Congenital vertical talus: a review. **Clin Podiatr Med Surg 2010**; 27(1): 145–156.
7. Westcott M, Dynes M, Remer E, Donaldson J, Dias L. Congenital and acquired orthopedic abnormalities in patients with myelomeningocele. **RadioGraphics 1992**; 12(6): 1155–1173.
8. Adzick N. Fetal myelomeningocele: natural history, pathophysiology, and in-utero intervention. **Semin Fetal Neonatal Med 2010**; 15(1): 9–14.
9. Asindi A, Al-Shehri A. Neural tube defects in the Asir region of Saudi Arabia. **Ann Saudi Med 2001**; 21(1–2): 26–29.
10. Guille J, Sarwark J, Sherk H, Kumar J. Congenital and developmental deformities of the spine in children with myelomeningocele. **J Am Acad Orthop Surg 2006**; 14(5): 294–302.
11. Yang J, Dobbs M. Treatment of congenital vertical talus: comparison of minimally invasive and extensive soft-tissue release procedures at minimum five-year follow-up. **J Bone Jt Surg-Am Vol 2015**; 97(16): 1354–1365.

How to cite this article: Alomran AK, Abahussain MA, Aldossary AA, Alshammari IB. Meningomyelocele with unusual feet deformity combination: A report of a rare case. J Taibah Univ Med Sc 2019;14(5):472–476.