Egyptian Pediatric Association Gazette (2014) **62**, 54–57

HOSTED BY

ELSEVIER

Contents lists available at ScienceDirect

Egyptian Pediatric Association Gazette

journal homepage: http://www.elsevier.com/locate/epag

Terminal hemimyelocystocele associated with Chiari II malformation

Umamaheswara Reddy V. [a], **Amit Agrawal** [b,*], **Kishor V. Hegde** [a], **Venugopal Sharma** [c], **Priya Malpani** [a], **Venkatesh Manchikanti** [a]

[a] *Department of Radiology, Narayana Medical College Hospital, Chintharedypalem, Nellore, Andhra Pradesh, India*
[b] *Department of Neurosurgery, Narayana Medical College Hospital, Chintharedypalem, Nellore, Andhra Pradesh, India*
[c] *Department of Pediatrics, Narayana Medical College Hospital, Chintharedypalem, Nellore, Andhra Pradesh, India*

Received 24 May 2014; accepted 30 May 2014
Available online 4 July 2014

KEYWORDS

Chiari malformation;
Diastematomyelia;
Myelocystocele;
Spinal dysraphism

Abstract Terminal myelocystocele (TMC) results from failure of embryonic CSF to drain outside the neural tube creating CSF reservoir within a dorsal meningocele. Association of Chiari II malformation with diastematomyelia and myelocystocele is extremely rare. Myelocystoceles do not have neural tissue so they have good prognosis after treatment, however when associated with hydromelia and Chiari malformation they present with neurological deficits. We present details of a 2 year old female who presented to us with this rare anomaly.

Introduction

Terminal myelocystocele (TMC) results from failure of embryonic CSF to drain outside the neural tube creating CSF reservoir within a dorsal meningocele.[1–5] TMC is the closed dysraphism as dilated terminal ventricle disturbs the growth of dorsal mesenchyme but not the surface ectoderm.[1,2] Myelocystoceles are associated with OEIS complex (omphalocele, exstrophy of bladder, imperforate anus and Spinal defects).[1,2,5,6] Association of Chiari II malformation with diastematomyelia and TMC is extremely rare. We describe a case of a 2 year old female who presented to us with this rare anomaly.

Case report

2 month old female baby was brought to hospital with complaint of swelling in lower back. Baby was born through an uneventful full-term normal delivery and baby cried spontaneously after birth. Subcutaneous swelling of child was noticed with overlying skin since birth and it was not increasing in size. Bladder and bowel were normal. On examination child was conscious and active. Soft tissue swelling in the dorso-lumbar junction was noted measuring 7.1 × 5.2 cm covered by skin surface with areas of scarring and discontinuity (Fig. 1). Swelling was fluctuant, increasing in size when baby was crying and was showing positive transillumation test. Head circumference and spine curvature were normal, plantar reflex was extensor, and tone was decreased on right side. Patient also had congenital talipes equino varus of the right foot. Plain radiographs obtained showed spina bifida at multiple lumbar vertebrae. MRI brain and spine were performed. MRI brain showed crowding of posterior fossa structures, tonsilar herniation,

* Corresponding author. Address: Department of Neurosurgery, Narayana Medical College Hospital, Chintharedypalem, Nellore 524003, Andhra Pradesh, India. Mobile: +91 8096410032.
E-mail addresses: dramit_in@yahoo.com, dramitagrawal@gmail.com (A. Agrawal).

Peer review under responsibility of Egyptian Pediatric Association Gazette.

1110-6638 © 2014 Production and hosting by Elsevier B.V. on behalf of The Egyptian Pediatric Association.
http://dx.doi.org/10.1016/j.epag.2014.05.002

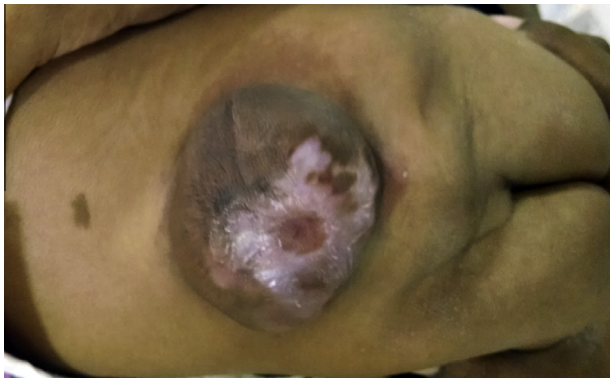

Figure 1 Large subcutaneous swelling in the lumbar region with overlying skin excoriation.

tectal beaking and concave clivus suggestive of Chiari malformation (Fig. 2). MRI spine revealed hydromelic and terminal spinal cord split by bony spur in the thoraco-lumbar region, split cords were also hydromelic and one of the hemicords was forming syringocele and was embodied into the meningocele (Figs. 3 and 4). Parents were counseled and child was operated for cosmetic reasons and nursing care.

Discussion

TMC is a closed spinal dysraphism constituting 4–8% of all spinal dysraphisms resulting from disturbed secondary neurulation.[1–3,5] TMC is a dorsal meningocele which lies posterior to a bony spina bifida and contains a terminal syringocele (Hydromelic cord).[7] The meningocele is the larger component lined with arachnoid cells which is contiguous with the subarachnoid space and the syringocele embodied within in the meningocele.[1,2,7,8] Caudal dilated central canal is lined by ependyma and consists of dysplastic glial tissue.[1,2] Generally there will be no communication between the outer meningocele and inner syringocele giving cyst within cyst appearance. Myelocystoceles are classified into non-terminal and terminal, depending on whether the malformation involves the conus medullaris or proximal segment of the spinal cord.[1–5,8] When there is associated diastematomyelia (abnormal notochord sequestration defect resulting in two split cords) either of hydromelic hemicord can enter cyst and it is called hemimyelocystocele similar to hemimyelomeningocele.[8–10] Association of myelocystocele with Chiari II malformation is rare and the combination of Chiari II with hemimyelocystocele is extremely rare and as for our knowledge this is the first case report where we are mentioning this rare association.[3] Advances and expertise in antenatal ultrasound have helped in early detection and characterization of neural tube defects (NTDs) however it is very difficult to diagnose myelocystocele from other conditions like meningocele, myelomeningocele and sacrococcygeal teratoma on ultrasound unless until we see classical cyst within cyst appearance.[11] Fetal MRI is an adjunctive tool that can provide better details for specific diagnosis of NTDs.[12–14] In neonatal life MRI is most important tool in diagnosing the various NTDs. As protocol one should image the whole spine and brain for specific diagnosis of NTDs. TMCs are potentially treatable spinal dysraphisms with least chances of neurological deficits if intervened in early neonatal period.[1–4]

Conclusion

Terminal hemimyelocystocele associated with Chiari II malformation is extremely rare. Antenatal ultrasound showing cyst within cyst appearance is very specific for myelocystocele. MRI is the imaging modality of choice for characterizing spinal dysraphisms. It is advisable to image the entire spine and brain to look for associations when such rare anomaly is detected. Myelocystoceles do not have neural tissue so they have good prognosis after treatment, however when associated with hydromelia and Chiari malformation they present with neurological deficits.[15]

Conflict of interest

None declared.

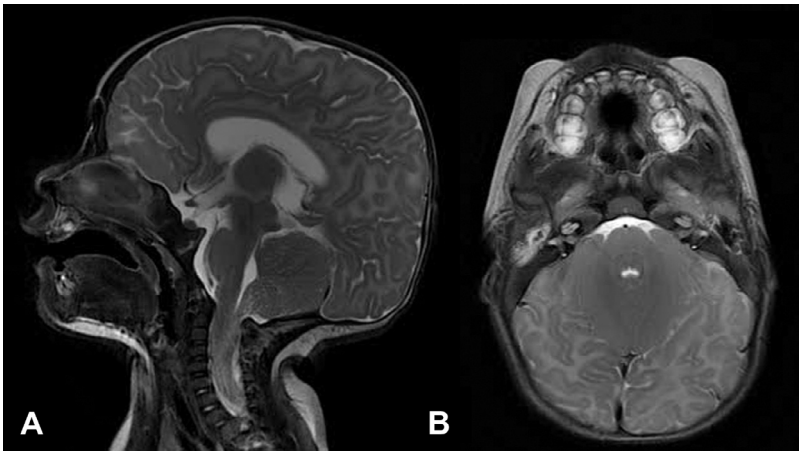

Figure 2 (A) Saggital T2W brain showing large Massa intermedia, tectal beaking, small posterior fossa and concave clivus. (B) Axial T2W image showing cerebral peduncles wrapping pons.