Presentation of a double myelomeningocele in the upper thoracic and thoracolumbar spine: A case report

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A B S T R A C T

BACKGROUND: Double myelomeningoceles are rare pathologies, with multiple cases described in developing countries. Causative factors are multifactorial, with folate deficiency as a leading associated factor. We describe a case of double myelomeningocele in the upper thoracic and thoracolumbar spine associated with split cord malformation, with emphasis on imaging appearance of this case.

CASE DESCRIPTION: This is a full-term newborn baby boy delivered through a cesarean section due to an antenatal diagnosis of multiple myelomeningoceles in the upper thoracic and thoracolumbar spine. The baby was operated 24 h after delivery to repair the defects and insert a ventriculoperitoneal (VP) shunt for associated hydrocephalus. The baby tolerated the operation well and is being followed and managed by a multi-disciplinary team.

CONCLUSION: Antenatal screening for myelomeningocele is paramount for prompt management. Multiple myelomeningoceles are uncommon, yet their management remains similar to single myelomeningocele.

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1. Introduction

Neural tube defects are common in developing countries with an incidence of 0.4–1.9 per 1000 live births [1,2]. Multiple neural tube defects are extremely uncommon, with few cases reported in the literature. The recommended operative management time for an un-ruptured cyst is within 2–3 days post-delivery. A multi-disciplinary team’s involvement is integral in managing neural tube defects with its associated urological, orthopedic, and neurological dysfunctions.

2. Presentation of case

The patient is a term baby boy born through a cesarean section on the 37th week of gestation in an otherwise uneventful pregnancy and delivery. He was found to have multi-site myelomeningocele on antenatal ultrasound screening, in addition to ventriculomegaly, bilateral clubfeet, ventricular septal defect, and a solitary kidney. Upon delivery, the Apgar score was 6 in 1 min and 8 in 5 min post-delivery. The head circumference was measured 35 cm (50th percentile), weight was 2170 g (10th percentile), and the length was 41 cm (less than 10th percentile).

Physical examination revealed two swellings on the back, one on the upper thoracic spine and the other on the thoracolumbar spine, no cerebrospinal fluid leakage was seen, and both membranes were intact (Fig. 1). A scoliotic deformity was appreciated on inspection. Stimulating the baby with a gentle touch of the plantar aspects of the feet demonstrated no movements in the entire lower limbs (including thigh flexion and extension). The anal sphincter was patent. Bilateral clubfeet were appreciated.

An x-ray of the entire spinal column was performed to study the scoliosis (Fig. 2). An MRI of the entire spinal column was also performed and showed the inner structures of the 2 myelomeningoceles (Figs. 3 and 4). Laboratory values of the blood count and basic metabolic panel were within normal limits. The Neurosurgery team were involved in the patient’s care immediately after birth. The patient was taken to the operating room 24 h after delivery for VP shunt insertion, followed by a simultaneous repair of the double myelomeningoceles.

The upper thoracic myelomeningocele was managed first. The normal skin was dissected away from the cyst, and a neural tissue was found with an atretic component that was resected after the
Two myelomeningoceles on the upper thoracic spine and thoracolumbar spine can be seen, with intact membranes and no cerebrospinal fluid leakage. A scoliotic deformity can be appreciated.

The patient tolerated the surgery well. The patient was transferred to the neonatal intensive care unit for 3 days, then was taken to the nursery. The orthopedics surgery team initiated the Ponseti-method (manual correction with serial casting) immediately post-operation. The urology team is following the bladder urodynamics for any urinary retention. Physical therapy was involved regarding the paraplegia of the lower limbs. The patient was discharged in good condition. Three months follow up after discharge revealed well-healed skin on the surgical sites. The baby was still unable to move his lower limbs. In addition, bladder urodynamics revealed urinary retention, currently managed by the urology team. The clubfeet were improving, and the family was instructed to continue using the Ponseti-method.
3. Discussion

Multiple neural tube defects (MNTD) is an extremely uncommon pathology with a prevalence of < 1% out of 474 cases by Ahmad et al. [2]. The neural tube starts approximately the first 4 weeks of the embryogenesis period [3]. The theory behind the development of neural tube defects (NTD) have been entertained by multiple authors, with the classic “zipper-like” fashion that occurs in the mid-cervical region and then continues cranially and caudally in a bi-directional pathway [2–4]. However, this theory fails to explain the development of MNTD, as one would expect that most NTDs would occur at the cranial or caudal neuropores rather than halfway in between [5].

Van Allen et al. [6], developed the multi-site closure hypothesis, where they have divided the closure sites into five main components. Firstly, site 4 starts to close, followed by sites 2 and 3, then sites 1 and 5 in an orderly fashion. Sites 1 and 2 are bidirectional, while sites 3, 4, and 5 are unidirectional. Site 1 origin corresponds to the mid-cervical area and meets site 4 rostrally and site 5 caudally. Site 2 origin corresponds to the prosencephalon-mesencephalon junction and meets site 3 rostrally and site 4 caudally. Site 3 origin corresponds to the stomodeum or stomatodeum (precursor to the adenohypophysis and the oral cavity) and meets site 2 caudally. Site 4 origin is the caudal part of the rhombencephalon and meets site 2 rostrally. Site 5 origin is the caudal point of the neural groove and meets site 1 rostrally [6,7].

The multi-site closure theory was challenged by O’Rahilly and Muller [8] as they have concluded that such closure sites do not occur in the human embryos. Therefore, Mahalik et al. [9], theorized that a zipper-like closure re-occurs at multiple sites when interrupted, acting more like a rescue method to complete the neural tube’s formation. This theory may explain the reason why we have multiple site variations of the MTND. In a series of 9 patients with MNTD by Deora et al. [7], 5 of their patients (55%) required a VP shunt. Collectively, of the 57 cases of MNTD found in the literature, only 12 patients (26%) required a VP shunt [7]. In general, it is recommended to repair myelomeningocele within 2–3 days of delivery if un-ruptured. Ruptured myelomeningocele should be considered for an emergency repair.

NTDs are generally induced by multiple environmental and genetic factors, with a prenatal supplementation of folic acid is used to protect against any erroneous formation of the neural tube [7,10]. Interestingly, many women who had conceived children with NTDs were found to have a normal folic acid level, similar to other women who delivered normal babies. Therefore, an innate error of folic acid metabolism is the cause, and supplementation of folic acid may overcome this blockade in the folic acid’s metabolic pathway [10].

4. Conclusion

Multiple myelomeningoceles are rare entities that can be prevented with folic acid supplement before conception can reduce the risk. Early recognition and surgical repair reduce mortality and morbidity. The multi-disciplinary team approach in managing neural tube defects is essential. The case report is reported in line with the SCARE 2018 criteria [10].

Declaration of Competing Interest

Nothing to disclose.

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Ethical approval

The case report was not submitted for ethical approval.

Consent

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Author contribution

T.E: study design, writing the paper, and revision.
S.K: Data collection, revision.
S.A: Supervision, and data analysis.

Registration of research studies

N/A.

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