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Case Series

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Double meningomyelocele - a rare variant of neural tube defect: A case series

Md Mokarram Ali,* **Yogender Singh Kadian**

Department of Paediatric Surgery, Pt B D S PGIMS, Rohtak, Haryana.

Correspondence*: Md Mokarram Ali, MS, MCh, Assistant Professor, Department of Paediatric Surgery, Pt B D S PGIMS, Rohtak, Haryana-124001. **E-mail**: mohdmokarramali1990@gmail.com

KEYWORDS

Congenital, Embryogenesis, Meningomyelocele, Spine

ABSTRACT

Background: A neural tube defect is a common congenital malformation and it commonly presents as an isolated cystic swelling on the back, anywhere from occiput to sacrum. However, multiple neural tube defects are rare with less than 100 cases reported worldwide.

Case Presentation: We are reporting two cases of multiple neural tube defects that presented in the newborn period with a history of concurrent swellings along the midline on the back. They were evaluated with ultrasonography and the diagnosis of double meningomyelocele was confirmed. Subsequently, they were operated on, both the lesions were excised, and the defects were repaired simultaneously.

Conclusion: Double meningomyelocele is a rare form of neural tube defect. Due to its rarity, exact embryogenesis is not known and thus, there is a need for further study on its embryology.

INTRODUCTION

Neural tube defects (NTDs) are common congenital malformations having an incidence of 1 to 5 per 1000 live births. [1] The prevalence is high in developing countries with wide geographical variations. [2] They usually present as an isolated swelling on the back anywhere along the spinal column from the occiput to the lumbosacral region. However, multiple neural tube defects (MNTDs) characterized by defects at two or more sites with normal intervening tissue are extremely rare. The embryology of NTDs is complex and multiple theories have been postulated to explain the same, of which zipper-like closure theory and Allen's multisite closure theory are widely accepted. However, multisite closure theory is the most appropriate explanation for the development of multiple neural tube defects. [3] In this article, we present two cases of double meningomyelocele [MMC].

CASE SERIES

Case 1: A 10-day-old female baby was brought to the outpatient department with a history of two swellings over the back along the midline noted at birth. The baby was born at term, via normal vaginal delivery to a mother of age 22 years. All the antenatal scans were normal. On examination, there was a small cystic globular swelling in the cervical region of size 2.5 × 2 cm and another similar swelling measuring 4 × 3.5 cm in the lower thoracic region (Fig. 1.) Both the swellings were intact with no evidence of any leak. The anterior fontanelle was flat and the head circumference was within normal range. Bilateral lower limbs were normal with normal movement and muscle bulk. The bladder was not palpable and the anal opening was normal. Ultrasonography (USG) of the lesion was done which confirmed our clinical diagnosis of meningomyelocele at both locations. USG of the cranium was also normal with no evidence of ventriculomegaly. Subsequently, meningomyelocele at both sites was excised and the defects were repaired using standard technique. The patient recovered well post-surgery and at one-month follow-up, there was no evidence of neurological deficit and hydrocephalus.

Case 2: A 26-day-old female baby presented with a complaint of minor swelling over the back of the head since birth. The baby was full term, born by normal vaginal delivery. There was no history of antenatal detection of any congenital malformations. On examination- there was a small cystic swelling of size 2 × 1 cm in the occipital region with a narrow stalk. There was another small swelling of size 1 × 1 cm in the thoracic region with blackish discoloration of the skin around the swelling (Fig. 2). Ultrasonography of the

lesions was suggestive of the occipital encephalocele and thoracic meningomyelocele respectively. USG cranium was also normal. The patient was planned for surgery and both the lesions were excised and the defects repaired. The patient had an uneventful recovery and at 1-year follow-up, no neurological deficit or hydrocephalus was noted.

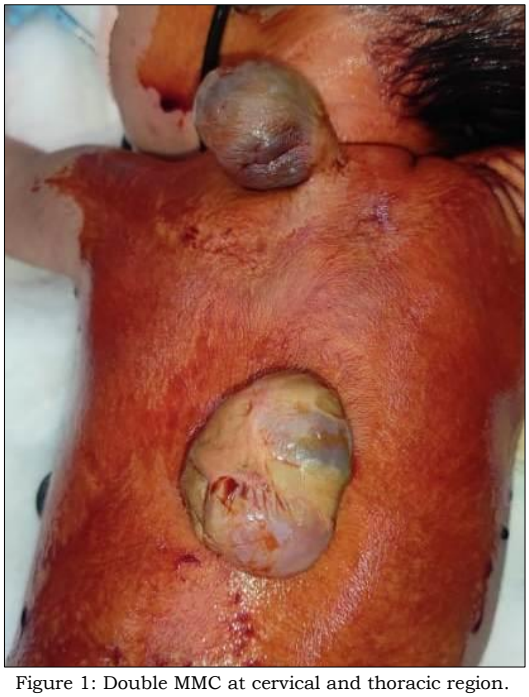

Figure 1: Double MMC at cervical and thoracic region.

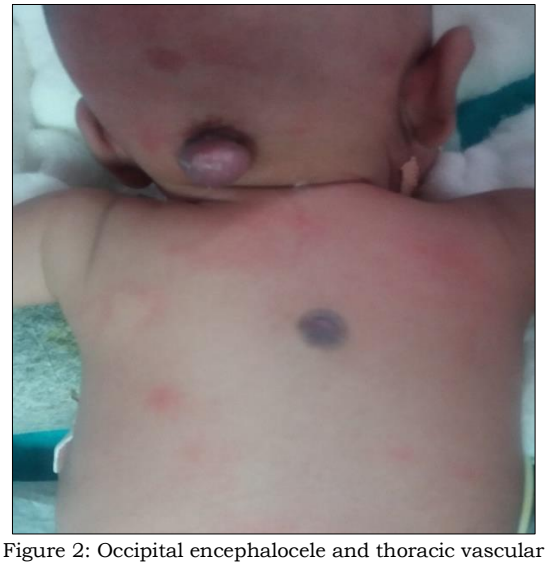

Figure 2: Occipital encephalocele and thoracic vascular lesion with underlying MMC.

DISCUSSION

Multiple neural tube defects are extremely rare and are characterized by defects in neural tube at two or more sites simultaneously with normal intervening tissue. [4] These cases are reported mostly from developing nations owing to poor socioeconomic status, lack of awareness as well as poor compliance with periconceptional folic acid supplementation and early pregnancy. [5] Various theories have been postulated to explain the embryogenesis of neural tube defects but not a single theory explains the development of MNTD completely. The zipper-like theory is a widely accepted theory for the embryogenesis of neural tube defects, (6) according to which the closure starts at a point in the cervical region and progresses in the cranial and caudal direction. The cranial neuropore closes at around 24 days of fetal life and the caudal neuropore closes two days later. According to this theory, neural tube defects result from the failure of these two ends to close. This explains the occurrence of occipital and lumbosacral MMC. However, it fails to explain the embryogenesis of MNTD's. The multisite closure theory proposed by Van Allen et al. is the most plausible explanation of MNTD's which states that there are five closure sites of neural tube and closure occurs at each site independently of one another. [7] Neural tube defects can result from failure of closure at any site and it may occur simultaneously at multiple sites. Mahalik et al. have proposed another explanation for the development of MNTDs according to which the zipper process restarts after being interrupted at any site. [8] Sarda D et al also reported a case of double meningomyelocele and supported the multisite closure theory. [9] Due to the rarity of MNTDs and not a single theory being accepted as a complete explanation, further work needs to be done to explain the embryogenesis.

Table 1: Showing comparison of previous studies with present study

Previous studies	Number of cases	Findings	Closure theory supported
Mahalik et al., 2012	10	Occipital and lumbar – 3 cases Thoracic and lumbar – 4 cases Lumbar and sacral- 1 case Cervical and thoracic- 1 case Cervicothoracic, lumbar and sacral swelling (triple NTD)- 1 caes	Multisite closure theory
Sarda D et al., 2007	1	Thoracic and lumbosacral	Multisite closure theory
Present study	2	Cervical and thoracic- 1 case Occipital and thoracic- 1 case	Multisite closure theory

In our case series [Table 1], we have successfully managed two cases of double meningomyelocele. Though we didn't find any significant antenatal history, this may not be reliable owing to operator dependency of an ultrasound. Both patients underwent surgery in a single stage and had no complications. On follow-up, both are doing well with no signs of neurological deficit and hydrocephalus.

CONCLUSION

Multiple neural tube defects are extremely rare. The exact mechanism of their development is not yet known. Further work needs to be done to understand the exact embryogenesis of this rare entity

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Abbreviations: MMC- meningomyelocele; MNTD- multiple neural tube defect; USG- ultrasonography

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