ECRONICON OPEN ACCESS

EC NEUROLOGY
Case Report

Multiple Neural Tube Defects: Do We Really Know the Truth?

Hemanshi Shah*, Jyoti Bothra, Charu Sharma and Gursev Sandlas

Department of Pediatric Surgery, Topiwala National Medical College and BYL Nair Charitable Hospital, India

***Corresponding Author:** Hemanshi Shah, Department of Pediatric Surgery, TNMC Topiwala National Medical College and BYL Nair Charitable Hospital, Mumbai, India.

Received: March 22, 2016; **Published:** May 09, 2016

Abstract

We present a case of 2month old boy with two synchronous meningomyelocele in support of the Multi closure theory. Our patient had two distinctly separate meningomyelocele in lumbar and thoracic regions of spine which concur in the direction of the new embryological theory.

Keywords: *Neural tube defects; multiple; meningomyelocele*

Introduction

Neural tube defects especially open neural tube types are becoming increasingly uncommon in the present century due to better understanding of the patho-physiology and introduction of folic acid prophylaxis. Ante-natal detection of NTD's and subsequent termination of such pregnancies also has a major role to play in the decreased prevalence of the disease. However, multiple neural tube defects which were rarely reported earlier seem to have made an exception to the trend. The basic understanding about the embryology of neural tube formation and its defects has also undergone a paradigm shift from the "Zipper" based theory to the "Multi-site closure" model. It is with this scenario in mind we present a case of a 2month old child with synchronous thoracic and lumbosacral MMC.

Case Summary

A 2month boy, first child of a non-consanguineous marriage, was brought by parents with complaints of two swellings over his back since birth. On examination a thoracic meningomyeloceleof 2x2cms and a large 7x6cms lumbosacral meningomyelocele were observed. Rest of the spine was normal on examination. He was paraplegic, however bladder was not expressible and anal tone was good. Transcranial ultrasound showed a normal ventricular/hemispheric ratio. MRI revealed a type-II Arnold Chiari malformation, thoracic menigomyelocele extending from D4 to D6, a thoracolumbar defect from D11 to L5 along with diastometamyelia between D6 to D7 and syrinx distal to D1. Surgical excision of sac sparing the nerve fibres and closure of both NTD's was done. No other defect or fibrous cord was seen. The patient is on follow up is doing well except for lower limb paralysis.

Discussion

Neural tube defects are a bigger continuum of problems than merely a congenital anomaly consequent to presence of the irreversible disability and need for lifelong monitored care. This also leads to social, economic and emotional cost to the family and the society [1].

Multiple neural defects were rarely reported in earlier literature. However whether this was due to a true rarity of the condition or paucity of published literature remains a grey area. Only 10 such published cases were found in literature till 2000, however recently two case series of 7 [2] and 10 [3] patients spanning over a period of 5 years were published, justifying the fact that this entity might not be as rare as it was thought to be.

Introduction of microsurgical electronic studies has led to better understanding of the embryo-pathology of Neural tube defects. The traditional "Zipper Theory" [4] of neural closure had postulated that closure starts in mid cervical region and proceeds rostrally and caudally with anterior and posterior neuropores closing at the end (24th and 26th embryo day respectively). This theory could explain the co-existence of encephalocele with meningocele as seen in <1% cases [5]. However occurrence of multiple NTD's could not be explained by this simplified theory. Further advanced studies in neural tube embryology in 1993 put forth a second theory of "Multisite Closure Model" by Van Allen., *et al.* [6] in which they hypothesized that there are five sites of initiation of neural tube closure. NTD's occur at the "collision sites" with opposing closure directions. These zippers are presumably under the control of one or more genes, mutations in which would cause neural tube defect in the region of the respective zipper [7,8]. This theory is more inclusive and logical but does not enjoy widespread support. Martinez-Frias., *et al,* conducted an epidemiological study based on the 'Multisite Closure Theory'. They found that isolated closure failure commonly at site 1 followed by site 4 and 2 is the most prevalent anomaly.

Split cord malformation is a common association with Meningomyelocele. However there has been only one reported case in literature till date [5].

Our case report helps in providing another evidence for the new theory. In the present case that we present here our patient had two different neural tube defects which had no connection between them. This can be explained by the new theory where two sites of closure might have failed in same neural tube but in isolation from each other. The old theory though explains a meningocele falls short to explain this anomaly. A better understanding of embryological and molecular mechanisms of NTD's will hopefully help the medical fraternity in detecting, preventing and treating the NTD's in utero and thus decrease the morbidity and disability burden of these patients.

Bibliography

1. Digra and Nasib C. "Primary Prevention of Neural Tube Defects". *Jk Sciences Journal* 2.6 (2004): 437-449.
2. Ahmad FU., *et al*. "Multiple Neural Tube Defects: A Clinical Series of Seven Cases and Their Embryological Basis". *Pediatric Neurosurgery Journal* 44.4 (2008): 280-287.
3. Mahalik SK., *et al*. "Multiple Neural Tube Defects May Not Be Very Rare". *Child Nervous System Journal* 29.4 (2013): 609-619.
4. Moore KL and Persaud TV. "The Developing Human: Clinically Oriented Embryology in the Nervous system". 7th edition. Philadelphia: WB Saunders Co (2004): 431-442.
5. Singh N., *et al*. "Multiple Neural Tube Defects in a Child: A Rare Developmental Anomaly". *Surgical Neurology Journal* 3 (2012): 147.
6. Van Allen MI., *et al*. "Evidence for Multi-Site Closure of the Neural Tube in Humans". *American Journal of Medical Genetics* 47.5 (1993): 723-743.
7. Nasib C. "Digra Primary Prevention of Neural Tube Defects". *Jk Sciences Journal* 6.1 (2004): 1-3.
8. Monika GO and Agnieszka S. "Genetic Basis of Neural Tube Defects, Regulatory Genes for the Neurulation Process". *Journal of Applied Genetics* 43.3 (2002): 343-350.

Volume 3 Issue 2 May 2016