

The effect of size of the sac on neurological state in patients of Myelomeningocele

Urooj Fatima, Hussain Mehdi, Abdullah Muttaqi, Meesam Iftikhar, Surriyya Sarwat, Mohammad Farooq Umer

Abstract

Objective: To determine the effect of the size of the sac on neurological state in patients of Myelomeningocele.

Material and Methods: 50 diagnosed cases of myelomeningocele were collected from Jinnah Post-graduate Medical Center from January 2104 till December 2017. The age of the patient ranges from 0-10 years. Patients were included regardless of sex, socio-economic and ethnic back-ground. Patients were examined for the size of the sac through MRI scans.

Results: The study shows that as the sac size increases, the disease got worsen. Whereas the complications would be lessen with decrease size of the sac.

Conclusion: Myelo-meningocele is the most frequent neural tube defect which occurs due to improper closure of neural tube. The dimension of cyst has greater influence on the neurological state of patients.

Keywords: Myelomeningocele, Sac, MRI scans, neural tube defect

Introduction:

The prime source of mortality of newborn is the inborn abnormality of central nervous system. The most frequent disorder of the nervous system is myelomeningocele (MMC). It is characterized by protrusion of meninges and cerebrospinal fluid alongwith neural elements through a sac.¹ The principal causative agent of the disease has not been known but it is an established fact that MMC is caused by multiple factors including both environmental as well as genetic. The disease can only be restricted by continuous intake of folic acid before pregnancy.² The cyst of MMC can be evident on any part of the spinal column but the most consistent location is lumbo-sacral region.³ The patients having such type of illness have long lasting disorder such as lack of mobility of lower limbs and improper functioning of bowel and bladder.⁴ It involves atonic bladder and bowel and partial or complete paresis of lower limbs. The individuals usually depend upon crutches, wheel-chairs or braces for movement. The magnitude of the sac con-

taining neural elements reveals the worsening of the disease.⁵

In this study we want to share our experience that the diameter of the sac of MMC has great impact on the neurological state of the patients.

Material and Methods:

A total number of 50 known cases of myelomeningocele (MMC) were included in the study. The study was conducted in Dow Diagnostic and Research Laboratory (D.D.R.L.). The study design is cross sectional. The severity of the disease is evident by the fact that majority of patients were neonates i.e. less than one year of age. The attendant of patients was informed by the study and the consent form was signed by them. The size of the cyst is different in every individual. The scale present on the MRI film is taken as standard.

Two anatomical parameters were undertaken.

- Anterio-posterior diameter of cyst = 'a'
- Craniocaudal diameter of cyst = 'b'

Received:

17th February, 2018

Accepted:

2nd December, 2018

Jinnah Sindh Medical University, Karachi

U Fatima
S Sarwat

Jinnah Medical and Dental College, Karachi

H Mehdi
A Muttaqi
M Iftikhar
MF Umer

Correspondence:

Dr. Urooj Fatima
Department of Anatomy
Jinnah Sindh Medical University, Karachi
Cell:
Email: urooj.fatima@jsmu.edu.pk

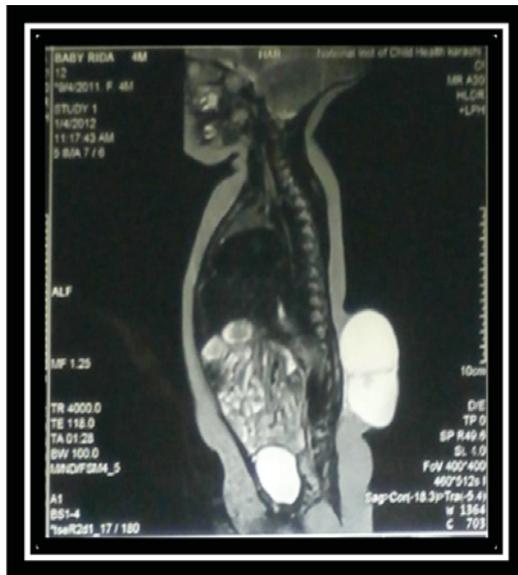
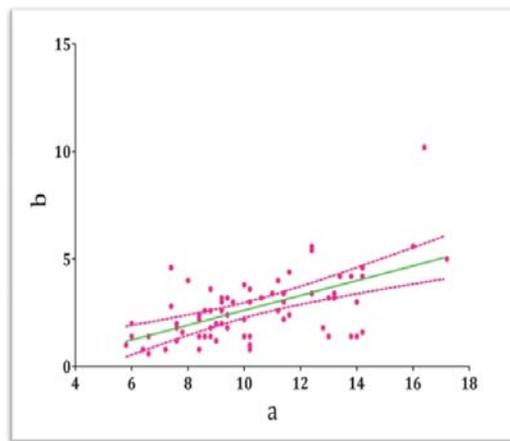


Figure. 1: This is the picture of MRI showing (a) antero-posterior and (b) craniocaudal diameters in sagittal section imaging.



Graph. 1: Graph showing the correlation between a&b. (a) cyst and the vertebrae diameter (b) length of the cyst.

Results were statistically analyzed by Fisher Exact test and One way Anova (ANOVA) using SPSS version 21.

Results:

To measure different diameters of cyst of myelomeningocele, 2 variables i.e., "a" and "b" were selected. The transverse line taken between cyst and the vertebrae is shown as (a). The vertical line from one point of cyst to the other is shown as (b).

The graphical representation shows that 'a' and 'b' are positively correlated (Graph 1).

Mean±SD of size was 17.12 ± 9.75 with C.I (14.34-19.89) cm as shown in table 1. Out of 50 myelo-meningocele patients, 39(78%) have sac size less than 5×5 cm. These patients have atonic bladder and decrease bowel movements. Whereas 11(22%) patients have sac size greater than 5×5 cm. These patients in addition to atonic bladder and bowel also show lower limb paralysis (table 2). The data analysis by Fisher Exact test revealed significant differences ($p < 0.05$) between size of cyst of patients with respect to neurological deficit (table 3). The data analysis by One way Anova (ANOVA) revealed insignificant differences ($F = 1.126$, $p > 0.33$) among size of myelo-meningocele cyst with respect to location (table 4).

Discussion:

Neural tube defects (NTD) are the most frequent type among all birth anomalies.⁶ Myelomeningocele (insufficient closure beneath the zone of cranium) is the significant disorder of NTD. Apart from deficiency of folic acid⁷ there are various other factors which play a significant role in the causation of disease. It includes raised maternal body temperature,⁸ usage of anti-seizure drugs⁹ like carbamazepine¹⁰ and valproic acid.¹¹ In addition to it, elevated blood glucose levels¹² and over-weight¹³ mothers are more prone to have the disease. There are a variety of genes which have been involved in the disorder.¹⁴ Experimental analysis reveals about 250 genes to be associated with NTD.¹⁵ Globally the frequency of NTD is 18.6 cases/1,000 live births.¹⁶ In United States the occurrence of Spina bifida is 3.5/10,000 live births.¹⁷ The incidence of MMC in Poland is 6/10,000 live births.¹⁸ In Iran the frequency of Spina bifida is 11/10,000 live births.¹⁹ The occurrence of NTD in India ranges from 0.5-11/1,000 live births.²⁰ The incident of Spin bifida in Pakistan is 3-5 per 1,000 live births and the prevalence of MMC among different types of Spina Bifida is 95% and that of meningocele is 5%.²¹ On clinical examination, a cyst is evident on any part of the vertebral column. The deformity in spinal cord would lead to impairment of leg, bladder and bowel function.²² Patients present with various

Table-1: Descriptive statistics of size of Myelomeningocele(in cm)

Descriptive		Statistic	Std. Error
	Mean	17.1200	1.37933
Size	95% Confidence Interval for Mean	Lower Bound 14.3481	
		Upper Bound 19.8919	
	5% Trimmed Mean	16.6111	
	Median	16.0000	
	Variance	95.128	
	Std. Deviation	9.75337	
	Minimum	4.00	
	Maximum	45.00	
	Range	41.00	
	Interquartile Range	16.00	
	Skewness	.642	.337
	Kurtosis	-.089	.662

Table-2: Frequency of Neurological Deficit(lower limb Paralysis)

	Frequency	Percent	Valid Percent	Cumulative Percent
Valid	No	39	78.0	78.0
	Yes	11	22.0	100.0
	Total	50	100.0	100.0

Table-3: Stratification of Size of cyst with respect to Neurological Deficit

	Size (in cm)	Neurological Deficit		Total	P-Value
		Yes	No		
	4-25	4	38	42	0.001
	>25	7	1	8	

Table-4: Stratification of size with respect to location

	N	Mean	Std. Deviation
Cervical	3	9.0000	3.00000
Lumbar	46	17.6739	9.92204
Thoraco-lumbar	1	16.0000	.
Total	50	17.1200	9.75337

Table-4: Stratification of size with respect to location

	Sum of Squares	Df	Mean Square	F	Sig.
Between Groups	213.171	2	106.586	1.126	.333
Within Groups	4448.109	47	94.641		
Total	4661.280	49			

complications like lower limb weakness and loss of bladder control. Patients may have other associated symptoms like orthopedic deformities,²² hydro-cephalus,²³ Chiari II malformation.²⁴

We have found that the dimension of the sac has a great impact on the neurological state of the

patients. Although the sac size has been shown to be a significant prognostic factor but unfortunately this area has been remain neglected. It has been observed that greater sac contains a large amount of neural elements which would ultimately worsen up the state. Whereas, if the diameter of the sac is small, ir-respective of the age of the patient, fewer amounts of neurological elements would be protruded leading to decrease neurological disabilities. The patient has to stay for a shorter period in hospital which in turn lessens the risk of infection. While reviewing literature, it was found that Eseoğlu et al in 2017 supported the view considering sac size as the most important factor.⁵ In 2012 Musluman et al also favored the significance of sac size.²⁵ Wilson RD et al in 2007 acknowledged the association of the neurological deficits with the dimension of the sac.²⁶ In our study we also come across the fact that the size of the sac has no relation with the location of the cyst. The cyst of myelo-meningocele could be present anywhere throughout the spine irrespective of its diameter.

Conclusion:

Myelo-meningocele is a life threatening state. It can be prevented by folic acid fortification prenatally. The size of the sac and the health care provided to these patients will help to improve the neurological state.

Conflict of interest: None

Funding source: None

Role and contribution of authors:

Dr. Urooj Fatima, did data collection, measurements and interpretation

Dr. Hussain Mehdi, did facilitated the data collection and diagnosis.

Dr. Abdullah Muttaqi, did facilitated the data collection and diagnosis.

Dr Meesam Iftiqhar, did data collection, measurements and interpretation

Dr. Surriyya Sarwat, did data collection, measurements and interpretation

Dr. Farooq Umer, did facilitated the data collection and diagnosis

References:

- Adzick NS. Fetal myelomeningocele: Natural history, pathophysiology, and in-utero intervention. In *Seminars in Fetal and Neonatal Medicine*, WB Saunders. 2010; 15(1): 9-14.
- Shaw GM, Lammer EJ, Zhu H, Baker MW, Neri E and Finnell RH. Maternal periconceptional vitamin use, genetic variation of infant reduced folate carrier (ASOG) and risk of spina bifida. *American journal of medical genetics*. 2002; 108(1): 1-6
- Chand MB AJ, Bista P. Anaesthetic Challenges and Management of Myelomeningocele Repair. *Postgrad Med J NAMS*. 2011; ;11 ((1)):41-6.
- Adzick NS, Thom EA, Spong CY, Brock III JW, Burrows PK, Johnson MP et al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. *New England Journal of Medicine*. 2011; 364(11): 993-1004.
- Eseoğlu M, Eroğlu A, Kemer S, Arslan M. Determination of the Effect of Diameter of the Sac on Prognosis in 64 Cases Operated for Meningomyelocele. *Korean Journal of Spine*. 2017;14(1):7.
- Sotres-Alvarez D, Siega-RizAM., Herring AH, Carmichael SL, Feldkamp ML, Hobbs CA et al. Maternal dietary patterns are associated with risk of neural tube and congenital heart defects. *American journal of epidemiology*. 2013; 12(8):1279-88
- Salomão RM, Cervante TP, Salomão JFM, Leon SVA. The mortality rate after hospital discharge in patients with myelomeningocele decreased after implementation of mandatory flour fortification with folic acid. *Arquivos de neuro-psiquiatria*. 2017;75(1):20-4
- Moretti ME, Bar-OzB, Fried S and Koren G. Maternal hyperthermia and the risk for neural tube defects in offspring: systematic review and meta-analysis. *Epidemiology*. 2005 ;16(2): 216-219
- AlShail E, De Vol E, Yassen A, Elgamal EA. Epidemiology of neural tube defects in Saudi Arabia. *Saudi Medical Journal*. 2014;35(Suppl 1):S68.
- Jose M, Sreelatha HV, James MV, Arumughan S, Thomas SV. Teratogenic effects of carbamazepine in mice. *Annals of Indian Academy of Neurology*. 2017;20(2):132.
- Wegner C, Nau H. Alteration of embryonic folate metabolism by valproic acid during organogenesis: implications for mechanism of teratogenesis. *Neurology*. 1992;42(4 Suppl 5):17-24.
- Agopian AJ, Tinker SC, Lupo PJ, Canfield MA, Mitchell LE, National Birth Defects Prevention Study. Proportion of neural tube defects attributable to known risk factors. *Birth Defects Research Part A: Clinical and Molecular Teratology*. 2013 Jan;97(1):42-6.
- Kondo A, Matsuo T, Morota N, Kondo AS, Okai I, Fukuda H. Neural tube defects: Risk factors and preventive measures. *Congenital anomalies*. 2017 Sep;57(5):150-6.
- Greene NDE, Stanier P and Copp AJ. Genetics of human neural tube defects. *Human molecular genetics*. 2009; 18(R2): R113-R129.
- Harris MJ and Juriloff DM. An update to the list of mouse mutants with neural tube closure defects and advances toward a complete genetic perspective of neural tube closure. *Birth Defects Research Part A: Clinical and Molecular Teratology*. 2010; 88(8): 653-69.
- Estevez-Ordóñez D, Davis MC, Hopson B, Arynchyna A, Rocque BG, Fieggen G, Rosseau G, Oakley G, Blount JP. Reducing inequities in preventable neural tube defects: the critical and underutilized role of neurosurgical advocacy for folate fortification. *Neurosurgical focus*. 2018 Oct 1;45(4):E20.
- Ba G, Wu QJ, Chen YL, Huang YH, Gong TT. Prevalence and time trends of spina bifida in fourteen cities located in the Liaoning province of northeast China, 2006-2015. *Oncotarget*. 2017 Mar 21;8(12):18943.
- Zamłyński J, Horzelska E, Zamłyński M, Olszak-Wąsik K, Nowak L, Bodzek P, Horzelski T, Bablok R, Olejek A. Current views on fetal surgical treatment of myelomeningocele—the Management of Myelomeningocele Study (MOMS) trial and Polish clinical experience. *Ginekologia polska*. 2017;88(1):31-5.
- Behrooz A. Prevalence of neural tube defect and its relative factors in south-west of Iran. *Pakistan Journal Of Medical Sciences*. 2007;23(4):654.
- Rai SK, Singh R, Pandey S, Singh K, Shinde N, Rai S, Prasad R, Shama SN. High incidence of neural tube defects in Northern part of India. *Asian journal of neurosurgery*. 2016 Oct;11(4):352.
- Khan MY, Khan K, Ahmed M. Short term outcome of surgical management of patients with neural tube defect (spina bifida). *Journal of Postgraduate Medical Institute (Peshawar-Pakistan)*. 2011;20(3).
- Alabi NB, Thibadeau J, Wiener JS, Conklin MJ, Dias MS, Sawin KJ, Valdez R. Surgeries and health outcomes among patients with spina bifida. *Pediatrics*. 2018 Sep 1;142(3):e20173730.
- Chance A SD. Hydrocephalus in patients with closed neural tube defects. *Child's Nervous System*. 2015 1;31((2)): 329-32.
- Vinck A MB, Mullaart R, & Rotteveel J. . Arnold-Chiari-II malformation and cognitive functioning in spina bifida. *Journal of Neurology, Neurosurgery & Psychiatry*. 2006; 77 ((9)):1083-6.
- Müslüman AM, Karşıdağ S, Sucu DÖ, Akçal A, Yılmaz A, Şirinoğlu D, et al. Clinical outcomes of myelomeningocele defect closure over 10 years. *Journal of Clinical Neuroscience*. 2012;19(7):984-90.
- Wilson RD, Johnson MP, Bebbington M, Flake AW, Hedrick HL, Sutton LN, et al. Does a myelomeningocele sac compared to no sac result in decreased postnatal leg function following maternal fetal surgery for spina bifida aperta? Fetal diagnosis and therapy. 2007;22(5):348-51.