Original Article

Localization Of Cyst Of Myelomeningocele Among Pediatric Patients

Urooj Fatima, Hussain Mehdi, Farrukh Mustafa Memon, Rubina Ghani, Hasan Ali

ABSTRACT:

Objective: To determine the frequency distribution of myelomeningocele cyst location among pediatric patients at a tertiary care hospital of Karachi.

Study Design and Setting: This cross sectional study was conducted in outpatient department of neurosurgery ward of JPMC.

Methodology: All fifty male and female babies having myelomeningocele, newly born to more than 12 months of age, who presented in outpatient department of neurosurgery ward of JPMC during the period of six months were included in the study. Verbal informed consent of the parents was taken and babies were examined for the location, size and associated complains of myelomeningocele.

Results: The patients presented mainly in the age group of newly born to 3months. Majority of them were males. Atonic bladder and bowel along with paralysis of lower limbs were uniformly found associated features. Among male patients distribution of myelomeningocele was 3% each in cervical and thoracic while 96% in lumbar region whereas female patients had 6% cervical and 84% lumbar cysts with no cyst in the thoracic region. The average size of MMC cyst in lumbar region was 4.0×4.2 cm.

Conclusion: Cyst of myelomeningocele was found to be more in male children up to 3months of age with location in the lumbar region. The average size of cyst was 4.0×4.2 cm and accounted for atonic bladder and bowel along with paralysis of lower limbs.

Keywords: Atonic bladder, Cyst, Gender, limb paralysis, location, Myelomeningocele

INTRODUCTION:

The leading cause of infant mortality is due to congenital anomalies.[1] A group of birth defects characterized by failure of fusion of midline structure is referred to as Spinal Dysraphism.[2] These anomalies are usually referred to as Neural Tube Defects (NTDs). It is the most frequent abnormality and a vital issue of well being in children.[3] It occurs due to inappropriate neural tube closure. Neurulation is an embryonic process in which a notochord forms a broad, uniform neural plate. The neural plate in turns, wrap inside to form a neural tube. It is this neural tube which is the precedent to the central nervous system, later on forming brain and spinal cord.[4] The growth and closure of neural tube occurs after 28 days of fertilization. Neural tube defects (NTD) occur due to inappropriate closure of neural tube.[5] NTDs are usually referred to as "Spina Bifida" a defect in the vertebral arches, may be covered by skin, meninges or underlying neural tissue. Myelomeningocele (MMC) is the most commonly occurring form of spina bifida in which meninges and cerebrospinal fluid along with neural elements buldge out through the sac.[6] Many factors are involved in the etiology of disease, of them, folic acid deficiency has been found to be the common cause.[7] Various authors have published different frequency figures regarding abnormalities of central nervous system varying from 1.3% in Pakistan[8,9] to 8.8% in Tanzania.[10] Worldwide prevalence of NTDs varies from 0.5-10 per 1000 live births.[11] The prevalence of the disease has greatly been reduced worldwide by periconceptional use of folic acid.[12] The cyst of myelomeningocele could be present on any part of vertebral column.

Since much literature is not available on the current topic at local level therefore present study was designed to determine the frequency distribution of myelomeningocele cyst location among pediatric patients.

METHODOLOGY:

This cross-sectional study was a part of "Molecular variations of Myelomeningocele in relation to VANGL1 gene". After approval of IRB letter# *IRB-556/DUHS/Approval/2015/130* of Dow University of Health Sciences, male and female children with ages from newborn to more than 12 months having MMC were enrolled from Neurosurgery O.P.D. of Jinnah Postgraduate Medical Center (JPMC) Karachi. This

Urooj Fatima
Lecturer, Department of Anatomy,
Jinnah Sindh Medical University, Karachi
E-mail: urooj.fatima@jsmu.edu.pk

Hussain Mehdi,
Department of Surgery
Jinnah Medical and Dental College, Karachi

Farrukh Mustafa Memon,
Department of Anatomy,
Dow University of Health Sciences, Karachi

Rubina Ghani
Department of Biochemistry
Jinnah Medical and Dental College, Karachi

Hasan Ali
Professor and Head of Biochemistry Department
Bahria University Medical and Dental College

Received: 22-01-19
Accepted: 12-03-19

study was conducted for a period of six months. All fifty cases who presented with rare problem of myelomeningocele were included in the study after verbal informed consent of the parents during the specified period.

Children were subjected to physical (local) examination of the cyst regarding location, size and associated complains of myelomeningocele.

RESULTS:

The number of patients between ages 0-3, >3-6, >6-9, >9-12 and >12 months were 20, 15, 7, 5 and 3 respectively. The frequency of male patients was 31 and that of female patients was 19 (Table 1). The cyst of MMC was mostly located at lumbar region (92%) with mean size 4.0×4.2 cm followed by cervical cysts (6%) having mean size 2.6×3.6cm and thoracic region (2%) having mean size 4×4cm (Table 2). The uniformly associated features found in all patients with MMC were atonic bladder, bowel and lower limb paralysis. The age wise distribution of patients having myelomeningocele at lumbar region is, 20 patients newly born -3months, 14 patients >3-6 months, 7 patients >6-9 months, 6 patients > 9-12 months and 3 patients were >12months of age. The gender wise distribution of patients of myelomeningocele was 3% male patients each had cervical and thoracic cysts while 96% had lumbar myelomeningocele whereas regarding female patients 6% had cervical, 84% lumbar and no cyst in thoracic region (Table 3).

S.No.	Parameter	Frequency	%
1	**Age in months**		
	Newly born-3	20	40
	>3-6	15	28
	>6-9	7	14
	>9-12	5	12
	>12	3	6
2	**Gender**		
	Male	31	62
	Female	19	38

Table 1: Age and Gender wise Distribution of Study Patients N=50

Location	Frequency	%	Mean Size(cm)	Associated features
Cervical	3	6	2.6×3.6	Atonic bladder, bowel & lower limb paralysis
Thoracic	1	2	4×4	Atonic bladder, bowel & lower limb paralysis
Lumbar	46	92	4.0×4.2	Atonic bladder, bowel & lower limb paralysis

Table 2: Location, Size & Associated features of Myelomeningocele N=50

Gender	Cervical	Thoracic	Lumbar
Male	1(3.2%)	1(3.2%)	30(96%)
Female	2(6.4%)	0	16(84%)

Table 3: Gender wise distribution of myelomeningocele

DISCUSSION:

Neural tube defects (NTDs) comprises of fatal anomalies leading to lifelong disabilities and death of babies. Myelomeningocele has been reported as the most diagnosed cases and is related with the greatest degree of impairment among NTDs.[13] Myelomeningocele (MMC) is an inborn error of central nervous system (CNS) which occurs due to inappropriate closure of spinal column and the neural elements buldge out in the form of pouch through the bone and skin. Worldwide prevalence of neural tube defects is 1-10 per 1000 live births.[14]

The most common age group at which the babies presented in our study was newly born to 3months. Majority (46) of the patients had cyst on the lumbar region followed by cervical and thoracic regions. Results are in consistent with study conducted in 2016, *Ullah W* suggested lumbar region as the most common site followed by sacral, thoracic and cervical regions.[15] Other authors have also documented the same.[16-19] In contrary to this, *Asindi* found thoraco lumbar being the commonest site.[20]

Patients suffering from MMC usually have sensory and motor neurological defects underneath the lesion. It may lead to weakness of lower limbs or paralysis that hinders or restrain from walking and the chances of pressure sores increases due to lack of sensation.[21] The issues of bowel and bladder incontinence are frequent because the desire for defecation is although vanished but the recto anal inhibitory reflex is sustained.[22] Due to paralysis of external anal sphincter, fecal soiling is inescapable when internal anal sphincter relaxes.[23] This is coinciding with our study results as all fifty patients had complains of atonic bladder and bowel along with paralysis of lower limbs.

The associated features were atonic bladder, bowel and lower limb paralysis present in all patients irrespective of cyst at different sites in our study. Schletker has supported the view of such patients suffering from neurogenic bladder and bowel.[24] This dysfunction occurs virtually in all children having MMC irrespective of the site of location of the cyst. In case of bladder dysfunction there is failure of urine storage or failure to empty the urine. This in turn may be related to bladder itself or with the external sphincter of bladder or with both. Consequently there is increase risk of urinary tract infection following failure to empty the bladder properly and adequately. This sequelae culminates overtime into urinary reflux, hydronephrosis , renal damage and ultimately renal failure. Similar findings are reported by other researchers.[25,26] The findings of neurogenic bladder and bowel are consistent with our study results. We have found that the most common age period of patient presentation in the tertiary care hospital with lumbar myelomeningocele was newly born to 3 months. The same is the finding of *Chand MB*.[16] This could be justified by the statement that MMC is a congenital anomaly so patients usually present in the hospital settings with such little age.

Majority were males (31%) in this study. Hidrosefalis[27] and Ghani[28] supported the view of males being more commonly affected by myelomeningocele than females. Whereas Nnadi DC[29] and Sachdeva S[30] have documented that females were more commonly affected than males with neural tube defects in their studies. Pre-pregnancy counseling and administration of folic acid throughout pregnancy, awareness regarding neural tube defects and myelomeningocele in the community through print and electronic media are small steps that can play a major role in combating this problem. Early and timely referrals of such babies to specialized units can also improve the quality of life of these patients. Large multicenteric studies on this subject are open avenues for future research in our country.

CONCLUSION:

Frequency distribution of cyst of myelomeningocele is found to be more in male children in the age group of newly born to 3months with location in the lumbar region. The average size of cyst was 4.0×4.2 cm and accounted for atonic bladder and bowel along with lower limb paralysis.

REFERENCES:

1. Sheridan E et al. Risk factors for congenital anomaly in a multiethnic birth cohort: an analysis of the Born in Bradford study. The Lancet. 2013; 382(9901): 1350-59.
2. McComb JG. Spinal and cranial neural tube defects. In Seminars in pediatric neurology 1997; 4(3):156-66. WB Saunders.
3. Mahapatra AK. Spinal dysraphism controversies: AIIMS experiences and contribution. Indian Journal of Neurosurgery. 2012;1(1):4-8.
4. Sadler TW. Langman's medical embryology. Lippincott Williams & Wilkins, Third to eight weeks: The embryonic period, 12th ed 2012: p.63.
5. Botto LD, Moore CA, Khoury MJ, Erickson JD. Neural-tube defects. New England Journal of Medicine. 1999; 341(20): 1509-19
6. Adzick NS, editor Fetal myelomeningocele: natural history, pathophysiology, and in-utero intervention. Seminars in Fetal and Neonatal Medicine; 2010: Elsevier
7. Gong R et al. Effects of folic acid supplementation during different pregnancy periods and relationship with the other primary prevention measures to neural tube defects. The Journal of Maternal-Fetal & Neonatal Medicine. 2016;29(23):3894-901
8. Toma BO et al. The prevalence and pattern of central nervous system anomalies in a neonatal unit in a tertiary hospital in Jos, north-central Nigeria. Journal of Medicine in the Tropics. 2018;20(1):63-67
9. Gillani S et al. Frequencies of congenital anomalies among newborns admitted in nursery of Ayub teaching hospital abbottabad, Pakistan. Journal of Ayub Medical College Abbottabad. 2011;23(1):117-21.
10. Mashuda F et al. Pattern and factors associated with congenital anomalies among young infants admitted at Bugando medical centre, Mwanza, Tanzania. BMC research notes. 2014 ;7(1):195
11. Alshalan AM, Hussain MA, El-Fetoh NM, Alenezi SZ, Alenazi AR, Alanazi MR et al. Spina bifida in Infants and Children in Arar, Northern Saudi Arabia. The Egyptian Journal of Hospital Medicine. 2018;72(1):3497-502.
12. 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):20-24
13. Leidinger A et al. Experience in the Early Surgical Management of Myelomeningocele in Zanzibar. World neurosurgery. 2019;121:e493-e499.
14. Maeda H et al. Myelomeningocele with Unilateral Right Renal Agenesis: A Case Report. AJP reports. 2018;8(1):e1-e3
15. Ullah W KM. Short Term Complications of Myelomeningocele Repair. An Experience in Neurosurgery Department Lady Reading Hospital Peshawar. Pakistan Journal Of Neurological Surgery. 2016;20(2):94-99
16. Chand MB, Bista P. Anaesthetic Challenges and Management of Myelomeningocele Repair. . Postgrad Med J NAMS. 2011;11 (1):41-46.
17. Tamburrini G, Frassanito P, Iakovaki K, Pignotti F, Rendeli C, Murolo D et al. Myelomeningocele: the management of the associated hydrocephalus. Child's Nervous System. 2013;29(9):1569-79
18. Haslam RHA. Congenital abnormalities of the central nervous system. In: Behnnan RC. Kliegman RM. Jenson ill (eds), Nelson Textbook of Pediatrics. 16th edn. WE Salmders Company, Philadelphia. 2000 : 1803-05
19. Eseoðlu M, Eroðlu 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):7-10.
20. Asindi A, Al-Shehri A. Neural tube defects in the Asir region of Saudi Arabia. Annals of Saudi medicine. 2001;21(1/2):26-29.
21. Copp AJ, Adzick NS, Chitty LS, Fletcher JM, Holmbeck GN, Shaw GM. Spina bifida. Nature reviews Disease primers. 2015;1:15007
22. Di Lorenzo C, Benninga MA. Pathophysiology of pediatric fecal incontinence. Gastroenterology. 2004 ;126:S33-40.
23. Di Lorenzo C. Chronic constipation and fecal incontinence in children with neurological and neuromuscular handicap. Journal of pediatric gastroenterology and nutrition.1997 ;25:37-39
24. Schletker J et al. Bowel management program in patients with spina bifida. Pediatric surgery international.2019;35(2):243-45.
25. Sawin KJ, Liu T, Ward E; NSBPR Coordinating Committee. The National Spina Bifida Patient Registry: profile of a large cohort of participants from the first 10 clinics. J Pediatr. 2015;166(2):444-450.e1
26. Alabi NB et al. Surgeries and health outcomes among patients with spina bifida. Pediatrics. 2018;142(3):e20173730
27. veHidrosefalisi M. When should ventriculoperitoneal shunt placement be performed in cases with myelomeningocele and hydrocephalus? Turkish neurosurgery. 2008;18(4):387-91
28. Ghani F, Ali M, Azam F, Ishaq M, Zaib J. Risks of surgery for myelomeningocele in children. Pakistan Journal of Neurological Surgery. 2016;20(1):53-57.
29. Nnadi DC, Singh S. The prevalence of neural tube defects in North-West Nigeria. prevalence. 2016;5(1):6-10.
30. Sachdeva S, Nanda S, Bhalla K, Sachdeva R. Gross congenital malformation at birth in a government hospital. Indian J Public Health 2014;58:54-56.