

Pattern of Presentation of Spinal Dysraphism

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ABSTRACT

Objective: To assess the pattern of presentation of spinal dysraphism and compare with already available data on the subject.

Design: Prospective study.

Material and Method: This prospective study was done in the department of Neurosurgery, King Edward Medical University (KEMU) Lahore, Pakistan from January 2008 to December 2009. Cases of spinal dysraphism admitted and managed in the department during this period were included in the study. Data was collected on a Performa for age, sex, socio-economic status, family history, location of spinal dysraphism, associated cranial, spinal and systemic congenital anomalies. Spina bifida aperta and occulta patients were included in the study. All patients of spina bifida occulta had MRI whole spine and of aperta CT or MRI brain as well.

Results: Total patients admitted with spinal dysraphism during this period were 56. Of them, spina bifida aperta were 42 (75%) and occulta were 14 (25%). Age range of patients of spinal dysraphism was from 1 day to 27 years. Male to female ratio was 1.5 : 1. All the patients of spina bifida aperta had myelomeningocele i.e. 42 (75%). Of them maximum patients had myelomeningocele at lumbosacral region i.e. 18 (42%) followed by lumbar area i.e. 11 (26%). No patients had cervical spina bifida aperta. Hydrocephalus was present in 32 (76%) patient. Of 56 cases of spinal dysraphism, 14 (25%) had spina bifida occulta. Among spina bifida occulta, lipomyelomeningocele and congenital dermal sinus were 5 (9%) each while 4 (7%) patients had meningocele. All patients had x-ray spine of the affected region. All patients of spina bifida occulta had MRI brain and whole spine. Patients of spina bifida aperta were advised CT or MRI brain.

Conclusion: All patients of spina bifida aperta presented with myelomeningocele, commonest at lumbosacral area. Age at presentation was relatively late. Of spina bifida occulta, Lipomyelomeningocele and congenital dermal sinus were unusually equal. Meningocele was relative less.

Key Words: Spinal dysraphism, spina bifida aperta, spina bifida occulta.

Abbreviations: CDS: Congenital Dermal Sinus, LMM: Lipomyelomeningocele, MC: Meningocele, NTD: Neural tube defect.

INTRODUCTION

Spinal dysraphism refers to group of congenital anomalies of spine in which midline structure fail to fuse.¹ It is of two types i.e. spina bifida aperta or open type and spina bifida occulta or closed type.¹

In spina bifida aperta, spinal canal is not covered by skin.¹ The examples of spina bifida aperta are myelomeningocele, myeloschiasis.¹ Spina bifida occulta is a type of spinal dysraphism in which underlying neural

structure is covered by skin.¹ The examples are Lipomyelomeningocele, meningocele, congenital dermal sinus, Spilt cord malformation etc.¹ Lipomyelomeningocele is commonest among spina bifida occulta.¹

Spinal dysraphism results from defective embryonic neurulation causing failure of fusion midline structures during 4th weeks of gestation.²

The exact cause is not known however, Genetic, environment and nutritional factors were found to play a role in development of this anomaly.³

MATERIAL AND METHOD

The prospective study was conducted Department of Neurosurgery, King Edward Medical University/Mayo Hospital Lahore, Pakistan. The study period was 2 years starting from 1st January 2008 to 30th December 2009. Total number of patients were 56.

Performa was used to collect data. The patients were admitted through neurosurgical outpatient clinic and referral from pediatric and obstetric department. Data was collected on a Performa for age, sex, weight, socioeconomic status, family history and location of spinal dysraphism and associated cranial, spinal and systemic congenital anomalies. Both spina bifida aperta and occulta were included in the study.

All patients of spina bifida occulta had MRI spine and CT or MRI brain.

RESULTS

Total patients admitted during this period were 56. Patients of spina bifida aperta were 42 (75%) and occulta were 14 (25%).

Age range of patients of spinal dysraphism was from 1 day to 27 years. Age range of patients of spina bifida aperta was from 1 day to 12 months and of occulta was from 1 month to 27 years.

Male were 34 (61%) and female 22 (39%). Male were more in both spina bifida aperta i.e. 28 (66%) and spina bifida occulta i.e. 9 (64%).



Fig. 1: Dorsolumbar Myelomeningocele.

Table 1: Age Distribution of Spina Bifida Aperta and Occulta.

Age	Spina Bifida Aperta	Spina Bifida Occulta		
		CDS	LMM	MC
< 48 Hours	6	0	0	0
2 Days – 1 Month	21	0	0	1
2 – 6 Month	12	0	1	3
6 Month -1 Year	3	1	2	0
1 – 5 Year	0	2	2	0
5 – 10 Year	0	1	0	0
10 – 20 Year	0	0	0	0
20 – 30 Year	0	1	0	0

CDS (Congenital Dermal Sinus)

LMM (Lipomyelomeningocele)

MC (Meningocele)



Fig. 2: Congenital Dermal Sinus.

All the patients of spina bifida aperta had myelomeningocele i.e. 42 (75%) (Fig. 1). Maximum patients

had myelomeningocele at lumbosacral region 18 (42%). It was followed by lumbar 11(26%) and dorso-lumbar 8 (19%) areas. Minimum patients presented with dorsal and sacral myelomeningocele i.e. 2 (5%) each. No patients had cervical spina bifida aperta.

Fourteen patients had spina bifida occulta. Of them, lipo-myelomeningocele (LMM) and congenital dermal sinus (CDS) were 5(36%) each (Fig. 2) and 4 (29%) patients had meningocele (MC). The common location of Spina bifida occulta was lumbar region 9 (64%), while rest of the occulta were located at dorso-lumbar 2 (14%), lumbosacral 2 (14%), and dorsal region 1 (7%). No mother was taking folic acid before pregnancy.

The types of Neural tube defect (NTDs) reported in the study are shown below in the table 1, 2, and 3.

Table 2: Sex Distribution.

Sex	Patients	Percentage
Male	34	61
Female	22	39

Table 3: Location of Spina Bifida Aperta and Occulta.

Sites	Spina Bifida Aperta	Spina Bifida Occulta		
		CDS	LMM	MC
Cervical	0	0	0	0
Dorsal	2	1	0	0
Lumbar	11	4	3	2
Dorsolumbar	8	0	2	0
Lumbosacral	18	0	0	2
Sacral	3	0	0	0

CDS (Congenital Dermal Sinus)

LMM (Lipomyelomeningocele)

MC (Meningocele)

DISCUSSION

The incidence of neural tube defect (NTD) varies from country to country, geographic zone and among different racial and ethnic groups.⁴ Its incidence is high in Indian and eastern Mediterranean population. It is also

high in European white and Hispanic population compared to black population.⁴

Recently, highest incidence is observed in northern China i.e. 138.7 / 10,000 live birth.⁵

However, there are some publications from India and Pakistan which claims to have even higher incidence of disease i.e. 13.80 / 1000 live birth in Peshawar and on rise 5.7 / 1000 live birth in Pondecherry, India.^{6,7}

However, the incidence is decreasing in Canada and west.⁸ The incidence of disease is decreasing in the developed countries and it is mainly due to planned pregnancy with folic acid supplementation and early detection and termination of pregnancy.^{8,9}

The pattern of spinal dysraphism in the study is similar to that cited in the literature and many studies specially those, done in third world countries. However, some variations exist.

Patients admitted within 1 month of birth were 28 (50%) but only 6 (10.7%) of them were brought within 48 hours after birth. Patients presented between 2 to 6 months were 16 (28%). This shows a delay between time of delivery and visit to the hospital. This could be due to the distance that they had to travel to reach the tertiary care hospital as most of the patients were from rural areas and majority had delivery of baby at home. Besides this, poverty, illiteracy, insufficient counseling from health care professionals, parent’s own negligence and high patient load to the hospital with lack of beds for admission also plays a major role for their delayed treatment.

One patient of congenital dermal sinus presented at the age of 27 years. Spina bifida occulta like congenital dermal sinus can be easily missed when delivered at home.

The study shows a male predominance of 1.5 : 1 which is the same as reported by K. Raj and S. N. Singh.¹⁰

In the study, almost all the patients were from Punjab province of Pakistan so no racial and ethnic variation exist. However, consanguineous marriage plays role for the occurrence of disease¹¹ Pakistan is a country where consanguineous marriage is common but the trend is gradually decreasing due to difficulty in finding suitable match for the marriage. Six (10.7) patients had family history of consanguineous marriage.

In our series, Myelomeningocele (MMC) 42 (75%) is the commonest of all spinal dysraphism. Myelomeninngocele has been reported differently in literature ranging from 9.2% to 72%.¹¹

Our observation is similar to that made by Asindi Asindi and Amer AL–Shehri in Saudi Arabia (70%).¹² No other type of spina bifida aperta was recorded in our study.

Regarding the site of myelomeningocele, lumbosacral area (42%) was the commonest location. Many studies have reported lumbosacral area as the commonest site of myelomeningocele.^{10,12} However, contrary to our study, Asindi Asindi and Amer Al – Shehri, have reported dorso – lumbar region 44.4% as the commonest location for myelomeningocele.¹² we have reported 8 (19%) myelomeningocele cases in dorso-lumbar area. The other sites for myelomeningocele were lumbar 11 (28%), sacral 3 (7%) and thoracic 2 (4%). No cervical myelomeningocele was recorded in the study. Literature has mentioned its incidence less than 5%.¹³

The study identifies spina bifida occulta 14 (25%) less common as compared to spina bifida aperta (75%). Regarding spina bifida occulta, similar observations have been made by Raj Kumar and S. N. Singh (23%) and P. G. Saluja (15.7%).¹⁰

Among Spina bifida occulta LMM and CDS share equal percentage i.e. 9% each. However, Lipomyelomeningocele is commonest of all spina bifida occulta. Raj Kumar and S. N. Singh have observed LLM and CDS, 19% and 4.5% respectively.¹⁰

Meningocele was 4 (7%) which is similar to the study of Behrooz A from Iran 8.9% but less than Khat-tak ST et.al. 17.30% from Pakistan.^{6,14}

CONCLUSION

Myelomeningocele is commonest of all spinal dysraphism. Numerous such children are brought into this world everyday some terminated from the mother's womb and many might be dying at home due to meningitis. This is creating an adverse impact on the mother's health and the economy of the nation Therefore, the health sector should put effort from the beginning from prevention to rehabilitation. A proper mechanism should be implemented right from the fortification of food, supplementation of folic acid to occupational therapy. An extensive public awareness program about use of folic acid in prenatal and natal period.

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