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Original Research Article

Clinical Profile of Spina Bifida Cystica: Six-Year Review from a Regional Centre, in Nigeria

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ABSTRACT

Background: Spina bifida cystica are the commonest congenital central nervous system anomalies compatible with life. Myelomeningocele can be associated with lifelong morbidity and mortality. Neural tube defects are abnormalities affecting neural tube fusion and occur between 18-21 days of gestation.
Genetic and environmental factors come together to cause neural tube defects. Preconception folic acid is paramount in preventing these disturbing anomalies.
Aims and objectives: The study aimed to describe the clinical pattern of cases of spina bifida in our centre and to determine the prevalence of hydrocephalus in patients with myelomeningocele.
Method: medical records of all patients with myelomeningocele and meningocele treated at department of Neurosurgery during the year 2011-2016 were reviewed. Biodemographics, clinical history and examination findings, associated anomalies were analysed.
Results: we had two hundred and forty-six patients over six-year period. Males were 56.5% and M: F ratio was 1.3:1. The mean age of presentation was two months. About sixty-five percent presented with ulcerated lesion, which were treated with antibiotics and dressing before excision and repair. There was history of maternal febrile illness in 83.7% of cases. Myelomeningocele was the most common lesion (84.6%), 15.4% had meningocele. About forty-eight percent had both hydrocephalus and myelomeningocele. All patients had excision, repair, and ventriculo-peritoneal shunt for associated hydrocephalus.
Conclusion: Health education and health insurance scheme is strongly advocated; as a tool in prevention and treatment of neural tube defects.

Key words: Spina bifida cystica, Myelomeningocele.

INTRODUCTION

Myelomeningocele is the most common congenital central nervous system malformation consistent with life. [1,2] It is described as protrusion of the spinal cord into a fluid filled sac as a result of failure of fusion of neural tube. [3-5] While meningocele is characterized by cystic dilation of meninges containing cerebrospinal fluid through a defect in the vertebral column with no neural tissue. [6-8]

Despite folic acid fortification of diet and health education, the incidence has been reported at 3.4 per 10,000 live births in USA. and prevalence of 2.2 per 1000 live birth in North-west Nigeria. [9] A number of genetic and environmental factors come to play to cause neural tube defects which occur within third to fourth week of intrauterine life. [10,11]

Numerous risk factors for development of neural tube defects have been identified, maternal febrile illness, maternal diabetes, use antiepileptic medications and family history of neural tube defects. [12,13]

Prenatal diagnosis allows early identification of spina bifida in the first trimester and gives room for termination of pregnancy where is legalized. Progress could be made in spina bifida through folic acid supplementation of diets but many women in our society are unaware of the existing relationship between folic acid and neural tube defects, do not attend regular antenatal care; in addition to the fact that spina bifida could occur before women are aware of their pregnancies.

Neurologic deficits in spina bifida results from failure of neural tube formation and spinal cord injury from prolonged exposure of neural elements to intrauterine environment. [1] Multidisciplinary approach is the key in the management of spina bifida.

The goal of post-natal repair is to cover the exposed neural tissues, prevents retrograde infection, prevents cord tethering, treat associated hydrocephalus and achieve good cosmesis. It is of note that damage to spinal cord and peripheral nerves is not reversible by post-natal repair.

Prenatal repair of myelomeningocele have been reported in developed nations with debatable benefits of improved neurological function, reduced hind brain herniation and better cosmetic results but the practice is still a mirage in our environment

METHODOLOGY

The medical records of all patients with meningocele and myelomeningocele treated at department of Neurosurgery of Usmanu Danfodio University Teaching Hospital Sokoto during the year 2011- 2016 were reviewed retrospectively. Biodemographic characteristics, clinical history, examination findings, date of first presentation and operation, associated anomalies, were noted and analysed.

RESULTS

We found two hundred and forty-six patients over the study period; males were 139 (56.5%). The male -to-female ratio is 1.3:1 in myelomeningocele alone and 1:1 in myelomeningocele associated with hydrocephalus. The mean age presentation was two months with range of one week to seven years. Majority (65.4%) of the patients presented with ulceration and infection, which were treated before surgery.

There was history of maternal febrile illness in 83.7% of cases. Paraplegia was observed in all patients with thoracic and lumbosacral myelomeningocele; while all patients with cervical lesions were neurologically intact.

Table (statistical analysis)

Variable	Diagnosis Myelo.	Myelo+Hydro	Meningocele	P. value & χ^2
Age(months)				
1-5	91(58.0%)	43(27.4%)	23(14.6%)	P=0.990
6-12	35(54.7%)	18(28.1%)	11(17.2%)	χ^2=0.295
>12	14(56.0%)	7.0(28.0%)	4.0(16.0%)	
Sex				
Male	82(59.0%)	33(17.3%)	24(17.3%)	P=0.261
Female	58(54.2%)	35(32.7%)	14(13.1%)	χ^2=2.688
ANC				
Yes	85(57.4%)	46(31.1%)	17(11.5%)	P=0.068
No	55(56.1%)	22(22.4%)	21(21.4%)	χ^2=5.380
Maternal Febrile Illness.				
Yes	119(57.8%)	51(24.8%)	36(17.5%)	P=0.025
No	21(52.5%)	17(42.5%)	2.0(5.0%)	χ^2=7.353
Nature of the lesion.				
Ruptured	110(68.3%)	41(25.5%)	10(6.2%)	P=0.000
Intact	30(35.3%)	27(31.8%)	28(32.9%)	χ^2=37.1
Duration of Hospital Stay(wk)				
< 1	0.0(0.00%)	0.0(0.00%)	2.0(100%)	P=0.012
1-2	128(58.7%)	58(26.6%)	32(14.7%)	χ^2=12.87
>2	12(46.2%)	10(38.5%)	4(15.4%)	

Key Myelo=Myelomeningocele, Hydro=Hydrocephalus.

Thirty-eight patients (15.4%) had meningocele and 84.6% has myelomeningocele at presentation. About forty-eight percent (68) presented with myelomeningocele associated with hydrocephalus.

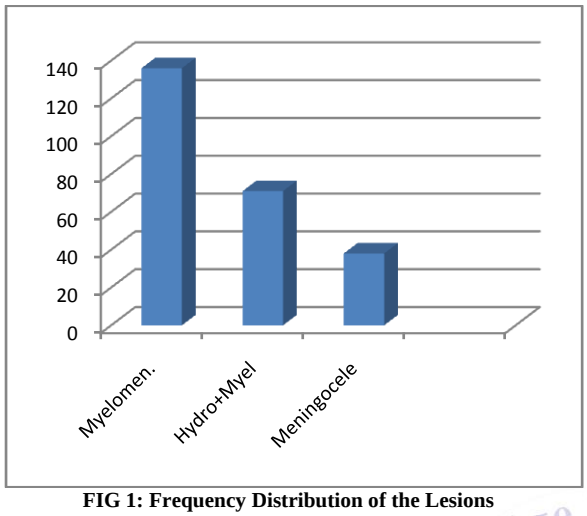
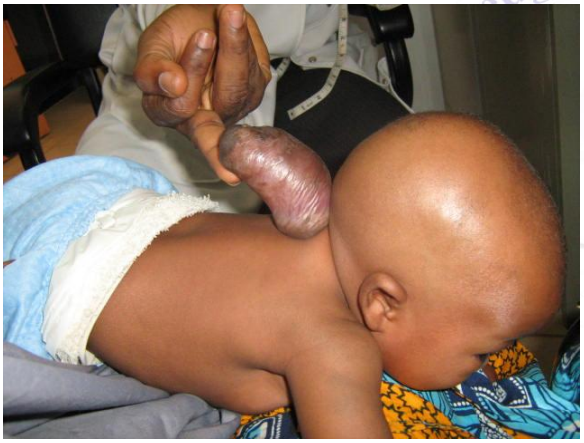
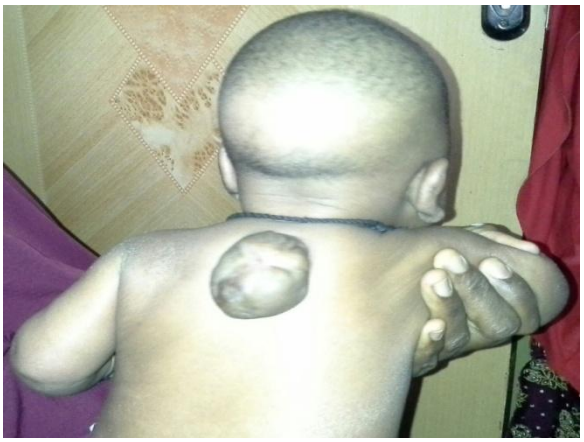


FIG 1: Frequency Distribution of the Lesions

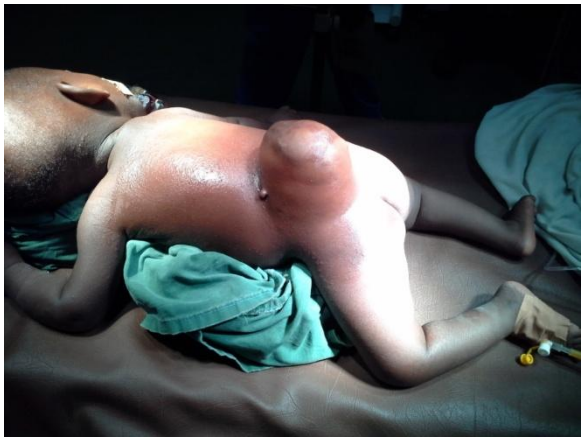


An infant with Cervical Myelomeningocele

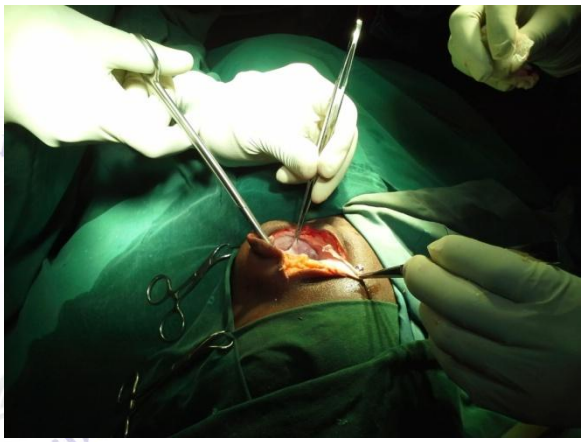


Thoracic Myelomeningocele

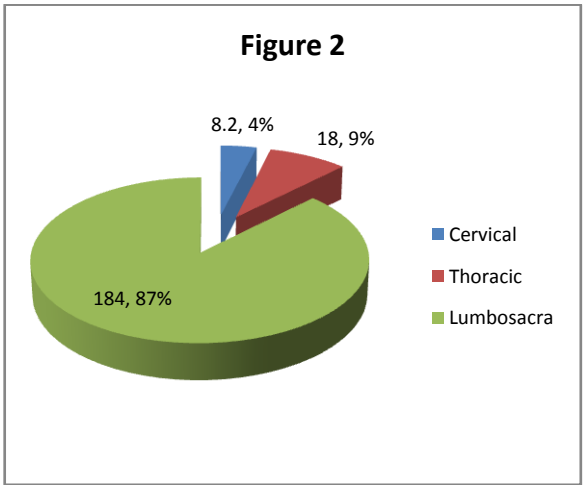
Eighty-eight percent (184) patients, presented with lumbo-sacral myelomeningocele, eighteen (8.5%) had thoracic and only six (2.8%) patients were diagnosed with cervical lesions.



Lumbosacral Myelomeningocele



Intra-operative picture showing another cyst after opening the dural sac



Anatomical classification of Myelomeningocele

All patients had excision and repair after adequate resuscitation and treatment of infection where present. Outcome was good in all the patients.

DISCUSSION

Myelomeningocele is the commonest neural tube defects in our environment. This is in tandem with, what is found in the literature. [1,2] Male children are commonly affected. Similar finding was reported in a study done in India. [14] However, other previous studies revealed female preponderance. [15-17]

In our series, we found lumbosacral myelomeningocele in eighty-eight percent of cases and cervical spine region was the least affected. This is comparable to the findings of A Sattar and colleagues in Karachi. [15]

Hydrocephalus was diagnosed in forty-eight percent of cases of myelomeningocele and all had ventriculo-peritoneal shunt insertion in addition to excision and repair. This is quite different from the report of many studies. [15,6] The variations could be attributed to differences in genetic and environmental factors across nations.

Majority of patients sought for care late, this could be due to lack of awareness and cost of treatment (about $ 300-400) per case, which is even higher if there is associated hydrocephalus.

We observed significant history of maternal febrile illness as possible risk factor. Significant number of our patients presented with ulcerated myelomeningocele that requires appropriate antibiotic therapy, regular dressing to achieve healing, before surgical interventions.

CONCLUSION

Myelomeningocele is still the most common neural tube defects. Prevalence of associated hydrocephalus is lower than what is obtainable in literature. Preoperative treatment of infection is paramount to prevent ascending infection. Health education and health insurance scheme will reduce the burden of neural tube defects and enhance early seeking for surgical care.

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