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Demographic and Clinical Characteristics of 63 Children with Myelomeningoceles

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Abstract

Background: Myelomeningocele (MMC) is the most common neural tube defect (NTD) characterized by the extrusion of the spinal cord into a sac filled with cerebrospinal fluid, resulting in lifelong disability. In the general population, the incidence of MMC ranges from 0.3 to 4.5/10,000 births. Live born infants with myelomeningocele have a death rate of approximately 10%. Many factors may play a role in the development of MMC such as environmental and genetic factors. In this study, we present our experience with a group of 63 children afflicted with MMC. **Methods:** This study was a retrospective analysis of 63 patients with MMC admitted to the neurosurgical department of Gabriel Touré Hospital from September 2017 to August 2018. A detailed history was obtained from the family at presentation. The family history and medical information before and during the pregnancy were compiled. Patients underwent complete physical and neurological examinations. Forty-seven (74.60%) patients underwent repair of the MMC and a ventriculoperitoneal shunt was placed in 12 (19%) patients with accompanying hydrocephalus. The risk factors, neurological status, and surgical results have been analyzed. **Results:** Of 63 children with MMC admitted to our neurosurgical department, 34 (54%) were boys and 29 (46%) were girls. Forty (63.49%) patients were the children of marriages of second cousins or closer. The mean age of the fathers was 34 years (16 - 65), while that of mothers was 26 years (16 - 38). The pregnancy was unplanned in all cases. Fourteen (22.22%) mothers had genitourinary infections, 9 (14.3%) had malaria and 57 (90.47%)

mothers used analgesics and antibiotics during the pregnancy. Fifty-nine (93.65%) children were born at term, 58 (92%) were delivered via normal spontaneous vaginal delivery, and 5 (8%) via cesarean section. Lumbosacral lesions were the most frequent in 27 (42.86%) patients. Forty-seven (74.60%) patients underwent repair of the MMC and a ventriculoperitoneal shunt was placed in 12 (19%) patients with accompanying hydrocephalus. Wound infection developed in 2 cases in the postoperative period. The mortality rate was 4.3%. **Conclusion:** Myelomeningocele is a congenital anomaly for which several risk factors are known as well as environmental and genetic factors. This emphasizes the importance of prevention with folic acid supplementation and genetic advice.

Keywords

Myelomeningocele, Neural Tube Defects, Hydrocephalus

1. Introduction

Myelomeningocele (MMC) is the most common neural tube defect (NTD) [1]. It is characterized by the extrusion of the spinal cord into a sac filled with cerebrospinal fluid, resulting in lifelong disability. In the general population, the incidence of MMC ranges from 0.3 to 4.5/10,000 births [2]. Live born infants with myelomeningocele have a death rate of approximately 10% [3] [4]. Many factors may play a role in the development of MMC such as environmental and genetic factors [4]. In this study, we present our experience with a group of 63 children afflicted with MMC. The objective was to evaluate the demographic and clinical characteristics for the first time in our department and to compare with data from the literature.

2. Methods

This study was a retrospective analysis of 63 patients with MMC admitted to the neurosurgical department of Gabriel Touré Hospital from September 2017 to August 2018. The selection criteria were all the children hospitalized in Gabriel Touré Hospital with myelomeningocele during the study period. A detailed history was obtained from the family at presentation. The family history and medical information before and during the pregnancy was compiled. Patients underwent complete physical and neurological examinations. Ultrasonography and CT scan were the preoperative imaging study in patients with hydrocephaly. Forty-seven (74.60%) patients underwent repair of the MMC and a ventriculoperitoneal shunt was placed in 12 (19%) patients with accompanying hydrocephalus. The risk factors, neurological status, and surgical results have been analyzed.

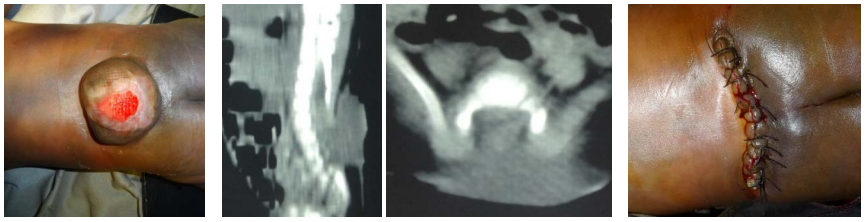
3. Results

We analyzed 63 children with MMC admitted to our neurosurgical department.

Thirty-four (54%) were boys and 29 (46%) were girls. Forty (63.49%) patients were the children of marriages of second cousins or closer. The mean age of the fathers was 34 years (16 - 65), while that of mothers was 26 years (16 - 38). Table 1 lists the demographic characteristics of MMC. The pregnancy was unplanned in all cases. Fourteen (22.22%) mothers had genitourinary infections, 9 (14.3%) had malaria and 57 (90.47%) mothers used analgesics and antibiotics during the pregnancy. Fifty-nine (93.65%) children were born at term, 58 (92%) were delivered via normal spontaneous vaginal delivery, and 5 (8%) via cesarean section. Lumbosacral lesions were the most frequent in 27 (42.86%) patients. The clinical characteristics are presented in Table 2. Forty-seven (74.60%) patients underwent repair of the MMC and a ventriculoperitoneal shunt was placed in 12 (19%) patients with accompanying hydrocephalus (Figure 1). Fourteen (22.22%)

Table 1. Demographic characteristics of myelomeningocele.

Demographic characteristics	n	%
Maternal age at delivery (years)		
16 - 25	29	46.03
26 - 35	30	47.60
>35	4	6.35
Paternal age (years)		
16 - 25	7	11.11
26 - 35	25	39.68
>35	31	49.20
Maternal education		
Yes	12	19.05
No	51	80.95
Paternal education		
Yes	20	31.75
No	43	68.25
Consanguineous marriages		
Yes	40	63.5
No	23	36.5
Pregnancy follow-up		
Regular	40	63.50
Irregular	19	30.20
none	4	6.30
Folic acid use		
Yes	59	93.65
No	4	6.35
Smoking		
Yes	0	0
No	63	100



(a) (b) (c)

Figure 1. (a) Clinical photograph of myelomeningocele; (b) CT scan showing the bone defect; (c) Clinical photograph after surgical repair.

Table 2. Clinical characteristics of patients with myelomeningocele.

Clinical characteristics	n	%
Site of myelomeningocele		
Thoracolumbar	5	7.94
Lumbar	17	26.98
Lumbosacral	27	42.86
Sacral	14	22.22
Neurological signs		
Paraplegia	41	65
Sphincter disturbances	35	55.6
Orthopedic deformity		
Yes	29	46
No	34	54
Hydrocephalus		
Yes	43	68.25
No	20	31.75
Congenital anomalies		
Yes	9	14.29
No	54	85.71

patients have been lost of view and 2 (3.2%) died before surgery. In the post-operative period, wound infection developed in 2 cases. In the short term follow-up, 8 (17%) children showed clinical improvement, 3 (6.38%) showed neurological deterioration, 15 (32%) remain unchanged and 21 (44.7%) had been lost of view. The mortality rate was 4.3%.

4. Discussion

MMC is commonly encountered congenital anomaly of the central nervous system (CNS). Most studies have found a female predominance [5] [6]. We found more afflicted boys in our study (54%). Our ratio is compatible with the survey by some authors [7] [8]. Its occurrence has been associated with a number of factors including extremes of maternal age. The mean age of mothers was 26

years in our study. Environmental factors may play a role in the higher risk of the pathologic embryo development. In our study, 57 (90.47%) mothers used analgesics and antibiotics during the pregnancy. The genetic contribution to malformation is described in the literature. Forty (63.5%) patients with MMC were the children of consanguineous marriages. This emphasizes the importance of prevention in families at higher risk to have a child affected by MMC, what can be achieved by the use of folic acid before and during pregnancy. Maternal folic acid intake is associated with reduced risk of spina bifida [9] [10] [11]. In the present study folic acid was used by mothers irregularly during pregnancy in most of the cases and none before pregnancy because the pregnancy was unplanned in all cases. Although some studies found an increased risk of MMC with smoking [12], no mother in our study was smoking. Low socioeconomic and educational status has been also observed as risk factors for MMC in epidemiologic studies. Fifty-one (80.98%) mothers and 43 (68.95%) fathers had no education in this study. The lumbosacral region was found to be most commonly involved in 27 (42.86%) children, as also reported in the literature. It is well recognized in the literature that there is a significant incidence of hydrocephalus associated with MMC [13]. We found a 68.25% occurrence of hydrocephalus in our series. Therefore, it is recommended that preoperative brain imaging is performed to assess the evidence of hydrocephalus in any patient who has spinal dysraphism. Orthopedic deformity, paraplegia and sphincter disturbances were preponderant in respectively 29 (46%), 41 (65%) and 35 (55, 6%) children, as also reported in the literature [14] [15].

5. Conclusion

Myelomeningocele is a congenital anomaly for which several risk factors are known as well as environmental and genetic factors. This emphasizes the importance of prevention with folic acid supplementation and genetic advice.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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