Myelomeningocele about 100 Cases and Review of Literature

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ABSTRACT

Spina bifida is a group of vertebro-medullary malformations with a common defect in neural tube closure during embryonic life. Myelomeningocele (MMG) is the most frequent anatomical subtype. The aim of this study is to investigate the epidemiology, diagnosis and treatment of MMG. It is a retrospective study based on a series of 100 cases of MMG recorded over 10 years in the neurosurgery department of Chu Ibn Rochd, Casablanca, between 2009 and 2019. The frequency of occurrence is an average of 10 cases per year; consanguinity is found in 24% of cases, the most predominant location was lumbar (87%) and accompanied by a motor deficit in 61% of cases, cerebral CT is performed in 94% of cases. Hydrocephalus was present in 83% of patients, orthopedic disorders were found in 75% of cases, 90% of cases studied underwent surgery for the malformation and 83% benefited from ventriculoperitoneal shunting. Our results are similar to those reported in the literature, with a 100% rate of patients operated on in developed countries, and MMG frequently located in the lumbar region. The rate of hydrocephalus associated with MMG is 88%, the associated orthopedic malformations according to other studies vary from 46% to 93.9%, and the rate of patients benefiting from a DVP varies between 60% and 90%. Improving the long-term prognosis requires the collaboration of neurosurgeons, urologists, orthopedists, psychologists, physiotherapists and parents, which will facilitate the patient’s autonomy and social integration.

Keywords: Diagnosis, myelomeningocele (MMG), prognosis, treatment.

1. INTRODUCTION

Spina bifida is a group of vertebro-medullary malformations with a common neural tube closure defect occurring during embryonic life between 24 and 26 SA [1]. Several anatomic-clinical entities can be distinguished, ranging from spina bifida occulta, with a good prognosis, to spina bifida aperta, with a poorer prognosis [2]. Myelomeningocele, the subject of our study, represents the most frequent anatomical subtype of Spina bifida aperta (86%), but also the most severe form of the disease [3]. Its incidence varies from region to region and country to country, depending on ethnic and environmental conditions (in Algeria 2004–2006 at 43.4/10000 births, Congo 1993–2001 at 0.6/10000 births, Germany 6.43/10000 births, Morocco 2011–2016 at 3.8/10000 births) [4], however, it remains the most common viable central nervous system embryopathy [2].

Risk factors include the genetic component, but also environmental factors such as lack of folate supplementation, and iatrogenic intake during pregnancy [5]. Anatomically, it is characterized by herniation of nerve tissue through bone dehiscence, causing sensory-motor disorders that vary according to the level of injury [6]. Diagnosis can be made prenatally by obstetrical ultrasound and maternal alpha-fetoprotein measurement between 15- and 20–22-weeks’ gestation [6]. but in many cases, especially in our context, the diagnosis is made at the time of birth. Myelomeningocele remains a serious pathology with significant neurological, urological and orthopedic repercussions, requiring multidisciplinary management and long-term follow-up [3]. The aim of our work is to carry out a retrospective study of the epidemiology and management of this pathology in our department, in order to identify the shortcomings and deduce a better therapeutic strategy.
2. Method

This is a retrospective, descriptive, monocentric study reviewing 100 patients who were treated for myelomeningocele over 10 years, between September 2009 and March 2019, in the neurosurgery department of Chu Ibn Rochd in Casablanca.

2.1. Inclusion Criteria

All patients treated for myelomeningocele during the period 2009–2019, with a usable record or, failing that, a contact telephone number for questioning.

2.2. Exclusion Criteria

Patients with unusable records due to lack of data.

2.3. Ethical Considerations

Our study is based on the following ethical foundations:

- Authorization to access and collect information granted by the head of the neurosurgery department
- Undertaking not to use any data for this study

3. Results

The geographical distribution of patients covered several regions, summarized in Table I. However, the majority of patients came from Casablanca. Patients ranged in age from 10 days to 2 years, with an average age of 4 months, and the results can be summarized as follows: 0–6 months: 84 cases (84%); 6–12 months: 10 cases (10%); 12–24 months: 6 cases (6%). There was a slight predominance of males, with a sex ratio of 1.2. Consanguineous couples accounted for 24% of the total number of cases. 74% of parturient had not taken folic acid supplementation. All cases were of 1st degree and 62% of pregnancies were monitored by an attending physician. 19% of the cases studied were discovered antenatally by a 3rd-trimester ultrasound scan showing a back mass or hydrocephalus, but 81% of the cases studied were discovered at birth. concerning the cranial perimeter (CP), we can subdivide our results into two main entities: patients with a normal cranial perimeter in 35% of cases, 53% of cases with macrocrania, and in 12% the CP was not specified. 61% of retained patients presented a lower limb deficit [49% with complete paraplegia and 12% with paraparesis of the MI. 83% of cases presented hydrocephalus on admission, while 17% of cases did not present ventricular system anomalies on CT. Orthopedic disorders were found in 75% of cases (Tables II, III; Fig. 1). 4%/90% of the cases studied underwent surgery after hospitalization (Figs. 2, 3). On the other hand, 10% of patients did not undergo surgery, due to: 7% lack of consent and refusal of surgery, and 3% early death from meningitis The immediate postoperative course was as follows: 21% of operated cases had complications (16 cases of postoperative meningitis, 2 cases of suture loosening (Fig. 4), 2 cases of CSF leakage, 1 case of ventricular dilatation), versus 79% of cases without complications. For the long-term evolution, Of the 61 patients with motor deficit, 40 remained stationary, 11 had moderate improvement and 10 were lost to follow-up.

4. Discussion

The prevalence of myelomeningocele has decreased in developed countries due to antenatal diagnosis and increasingly accessible folic supplementation. In the USA,
the prevalence of MMG was 1–2/1000 live births in 1980 and has recently fallen to 0.4/1000 live births [7]. Several studies have shown a variation in the incidence rate of myelomeningocele between 1–7/1000 live births according to geography, ethnicity, sex, socioeconomic conditions of the parents, maternal age, and parity, and although no reliable study has been able to link these different factors to the incidence of MMG [8]. We go from a frequency of 0.52%/ in France and 0.60% in Finland to a frequency of 5.5% in northern China [7]. In our study the average age was 4 months, however, R.H. Brau et al. [9] showed an average age of 42.2 days, W.C. Mezue et al. [10] the average age was 35.43 days, S. Alhindi et al. [11] showed a mean age of 44.1 days, and R. Kumar et al. [12] showed a higher average age of 3.1 years (Table IV). Y. Sogoba et al. [13] showed a male predominance of 54%, which is in line with our study, which showed a similar male predominance of 55%. Several studies have shown female predominance, such as that carried out by A.S.M. Hashim et al. [13] showed a rate of 50.7%, and that carried out by M.M. For consanguinity, first cousins present 2 times more neural tube defects than in the general population; whereas in France, the role of consanguinity is considered negligible [14]. According to Talbi et al., Morocco has one of the highest rates of consanguinity at 22.79% [15]. 5 cases of consanguineous marriage out of 17 cases of myelomeningocele were highlighted by Forci et al. [4] with a rate of 29.4% in Mali in a study by Sogoba [13] showed a rate of consanguineous couples of 63.5%, whereas our study showed a rate of 24%, which is in line with the rabat study, which showed a higher rate of consanguineous marriages, which is not the case in Mali, where customs still tolerate consanguineous marriages.

29% of the myelomeningocele cases considered in the study by Radouani [16] were from unattended pregnancies, in the study carried out in Iraq by Hassan [17] 40% of the cases studied arose from poorly monitored pregnancies, but our study showed that 28% of cases arose from poorly monitored or unmonitored pregnancies (Table V). Studies have shown that folic acid deficiency is considered by some authors to be the best-known risk factor and the easiest to prevent with simple measures [4]. In Ireland, according to a study by Mc Donnell RJ et al., the prevalence of MMG fell from 46.9/10,000 births to 11.6/10,000 between 1980 and 1994. These authors attribute these results to the use of folic acid in the periconceptional period [18].

![Fig. 2. The different aspects of MMG in our study.](image1)

![Fig. 3. Postoperative appearance after MMG cure.](image2)

![Fig. 4. Suture loosening.](image3)

**TABLE IV: MMG AVERAGE AGE BY STUDY**

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<tr>
<td>Average age of cases</td>
<td>42.2 J</td>
<td>35.43 J</td>
<td>44.1 J</td>
<td>3.1 J</td>
<td>4 months</td>
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The incidence of spina bifida reaching 65%–90% [3]. In our study, the incidence of hydrocephalus was 83% of cases, whereas 53% of our patients had macrocrania at birth, which is consistent with the series by Sayad [3] where macrocrania was seen in 60% of cases. Bashir [22] showed that 8 patients of the 60 total cases had IM paralysis in the lumbosacral form with a rate of 13.3% and 2 cases in the sacral form of myelomeningocele with a rate of 3.3%, whereas 7 cases had IM paresis in the lumbosacral form with a rate of 11.6% and 2 cases in the sacral form of myelomeningocele with a rate of 3.3%. Then Kumar [12] showed that 70 of the 102 cases studied had a motor deficit of the MI varied at a rate of 68.8% [49.1% had paraparesis of the MI and 19.5% had paresis of an MI] [12]. In contrast, our study showed that 61% of cases had an MI motor deficit subdivided into 49% with complete paraplegia and 12% with unrated MI paraparesis. In our study, 87% of cases were of lumbar MMG, 7% of dorsal MMG and 6% of sacral localization (Table VI). However, Ulsenheimer et al. showed that 10 cases of dorsolumbar localization (32%), 4 cases of lumbar localization (13%), 14 cases of lumbosacral localization (46%) and 1 case of sacral localization Y. Sogoba [13] showed different MMG locations [dorsolumbar 7.94%/lumbar 26.98%/lumbar 42.86%/sacral 22.22%]. Hashim [22] showed several MMG locations [cervical 5%/dorsal 7%/dorsolumbar 82%/lumbosacral 61%]. Hydrocephalus is the anomaly most frequently associated with MMG (Fig. 6), with the incidence of spina bifida reaching 65%–90% [3]. In our study, the incidence of hydrocephalus was 83% of cases, which is close to the findings of A.S.M. Hashim et al. du who showed a rate of 88% [22] which agrees with A.B.D. Rodrigues et al., where the rate was 83.5% but we found a lower rate than Beuriat et al., who showed a rate of 67%. The treatment of MMG malformation requires careful evaluation to select the surgical method that will minimize potential complications. The focus remains on the application of meticulous surgical techniques and reconstruction with appropriate closure of the different planes [3]. Physiotherapy plays an important role in improving the functional prognosis of MMG patients. Re-education should be adapted according to regular check-ups, and to the age and experience of the S.B. child, both on a motor and psychological level [3]. Post-operative complications include worsening motor deficits: it’s not uncommon to see some improvement after surgical treatment of the MMG placode, but it’s also common to see neurological deterioration post-op due to avoidable technical errors such as the use of povidone-iodine (Betadine), known to be toxic to nerve tissue during sterilization, and which should be used only for the surrounding skin. Excision of the

<table>
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<tr>
<th>Location</th>
<th>Percentage</th>
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<tr>
<td>Lumbar</td>
<td>87%</td>
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<tr>
<td>Back</td>
<td>9%</td>
</tr>
<tr>
<td>Sacred</td>
<td>4%</td>
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Fig. 6. EFT showing hydrocephalus in a newborn with MMG.

TABLE V: PREGNANCY RATES MONITORED BY STUDY

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<tr>
<td>Pregnancy follow-up rate</td>
<td>29%</td>
<td>40%</td>
<td>28%</td>
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TABLE VI: MMG LOCATION ACCORDING TO OUR SERIES
of large myelomeningocele cases are associated with con-

malnutrition due to hydrocephalus, postoperative ileus,

are diminished. This may sometimes be associated with

hormone, all anabolic processes, including wound healing,

to the increase in cortisol, ACTH, thyroxine and growth

casue by the hormonal alteration of operative stress due

dehiscence: as a result of the nonspecific catabolic response


arachnoid must be as delicate as possible to clearly dif-

Fig. 6. Major hydrocephalus.

dentiate the limits of the functional placode [2] wound
dehiscence: as a result of the nonspecific catabolic response

caused by the hormonal alteration of operative stress due

to the increase in cortisol, ACTH, thyroxine and growth

hormone, all anabolic processes, including wound healing,

are diminished. This may sometimes be associated with

malnutrition due to hydrocephalus, postoperative ileus,

Arnold Chiari malformation, etc., [3]. CSF leakage: 85% of

large myelomeningocele cases are associated with con-

genital hydrocephalus. This excess CSF pressure poses an

additional risk of CSF leakage [1].

5. Conclusion

Myelomeningocele is the most frequent form of Spina Bifida. Its etiology is multifactorial, but the functional consequences remain extremely serious. The development of ultrasound monitoring programs for pregnant women, folic acid and multivitamin supplementation and the increasing availability of genetic tests such as karyotyping have all helped to reduce the frequency of such pathologies. Early diagnosis by prenatal ultrasound and even fetal MRI is essential for such early surgery to avoid complications. Given the complexity of the associated malformations, multidisciplinary management is essential to improve our patients’ quality of life.

Author Contributions

All the authors contributed to the patient’s care. They also declare that they have read and approved the final version of this manuscript.

Conflict of Interest

Authors declare that they do not have any conflict of interest.

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