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Epidemiology of Spina Bifida Cystica in Sohag: A Hospital-Based Study

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Abstract

Background Data: Neural tube defects (NTDs) include a wide variety of pathologies due to the intrauterine failure of neural tube closure. It includes anencephaly which is incompatible with life, encephalocele and myelomeningocele and meningocele. Myelomeningocele (MMC) the commonest and worst form in which the spinal cord and the meninges protrude from a defect in the spine. Meningocele is a less severe form in which only the meninges protrude into a sac. MMC represents a major health problem with a higher incidence in developing countries. It is one of the death associated diseases in infants and early childhood. It compromises the patient's life quality and causes lifelong disabilities. Other congenital anomalies might be associated with the SB like congenital hydrocephalus, congenital talipus, and congenital heart diseases.

Purpose: To study patients' epidemiological data and possible risk factors in our locality.

Study Design: A cohort descriptive retrospective clinical case study.

Patients and Method: We reported 122 babies with spina bifida cystica came to neurosurgery clinic in Sohag university hospital between January 2009 and January 2015. We collect their epidemiological data and ask their parents about the possible factors.

Results: In 122 patients with spina bifida cystica, the age range varies from 1 day to 8 months (the mean age was 2 months). The study involved 66 males (54%) and 56 females (46%). 71 cases had associated hydrocephalus (58%), 33 had congenital talipus deformity (27%) and 26 cases had associated cardiovascular disease (21%). Seventy three percent of cases (N=89) came from rural area.

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Conclusion: spina bifida cystica is a common disease in Sohag government. Both sexes affected equally. Incidence was higher in rural areas, mothers who took medication in the first trimester, those who were not on regular folate intake and in families who had a previous baby with spina bifida cystica. (2015ESJ107)

Keywords: Spina bifida, meningomyelocele, congenital anomalies, neural tube defects.

Introduction

There is evidence that spina bifida existed in ancient Arabian and Greek civilizations.[18] Peter Van Forest first recorded a child with spina bifida in 1587 and in 1610 he performed the first reported surgical resection of the myelomeningocele sac. The term spina bifida was first used by Professor Nicolai Tulp of Amsterdam in 1652. He draws the first anatomic illustration in 1641. In 1761, Morgagni was the first to describe associated diseases as congenital hydrocephalus observed in patients with the myelomeningocele.[7]

Neural tube defects (NTDs) include a wide variety of pathologies due to intrauterine failure of neural tube closure.[5] It includes anencephaly which is incompatible with life, encephalocele and myelomeningocele and meningocele.[12] Myelomeningocele (MMC) is the commonest and worst form in which the spinal cord and the meninges protrude from a defect in the spine.[25] Meningocele is a less severe form in which only the meninges protrude into a sac.[25] MMC represents a major health problem with a higher incidence in developing countries.[16] It is one of the death associated diseases in infants and early childhood. It compromises the patient's life quality and causes lifelong disabilities.[28]

Spina bifida occurs in early gestational age (within 6 weeks of gestation) resulting in neurological dysfunction as paraplegia or paraparesis, sensory loss and urinary and fecal incontinence. SB has many hazardous complications as repeated urinary tract infection which is the major cause of death in these cases.[17] Other congenital anomalies might be associated with SB like congenital hydrocephalus, congenital talipus, and congenital heart diseases.[19]

From an embryological point of view, there are two well-established theories; the old theory (watery tumors of the vertebrae) assumed by Morgagni which postulated that the continuous pressure of the CSF in hydrocephalic babies leads to rupture of the neural tube and bone. The second recent theory assumed that there is failure of the embryonic neural plate to close in its caudal part with the failure of the vertebral arches to fuse.[8]

There are many associated factors that might contribute to the occurrence of MMC, like drug intake in the first trimester especially antiepileptic and chemotherapeutic agents, diabetic mothers and maternal infection during pregnancy (TORCH) infections.[26] Genetic risk factor plays a fundamental role; Loss of the Sox2 gene was observed in most cases of NTDs.[2] Positive family history of NTDs was in about 10%, and they increase markedly if there were two previously affected family members.[2]

The prevalence of NTDs at birth varied markedly in each country and race. It ranges from as high as 1% in China, to approximately 1 case in 5000 or less in Norway and Sweden.[6] Its incidence varies from 3% from total live births to 0.003 % in the Unites States (US),[27] while no accurate data recorded its incidence in Egypt. The common sites for MMC are lumbosacral and cervical regions (87%) as they are the last areas of closure of neural tube, very rare in the dorsal region. More than half of MMC cases are associated with hydrocephalus (Chiari malformation II).[6]

Due to the great importance and the health and socioeconomic burden of this health problem we conducted this study to highlight

the risk factors associated with the diseases and epidemiology in our locality.

Patients and Method

This retrospective descriptive study was conducted on 122 patients in the neurosurgery department at Sohag university hospital between January 2009 and January 2015. Demographic data of the babies regarding baby's gender, type of spina bifid cystica, mother's age, presence or absence of consanguinity between parents, history of mother's drug intake, parity of the mother, gestational age, and mode of delivery were recorded (Table 1). The data were analyzed by SPSS, Version 10. Chi Square, Fisher exact and Student's t-test was applied whenever necessary. The P-value of P=0.05 or less was considered statistically significant.

Inclusion criteria were: Spina bifida cystica came to Neurosurgery clinic in the period between January 2009 and January 2015. Exclusion criteria were: Spina bifida occulta and Other NTDs as encephalocele or anencephaly

Results

Through the period of January 2009 and January 2015, we collected 122 cases of spina bifida cystica. There were 83 cases of myelomeningocele (68%) and 39 cases of meningocele (32%), males were 54% (N=66) and females were 46% (N=56), with male/female ratio 1.18 with no statistical difference. The mean age of mothers was 24±7 years. Moreover, 100% of mothers were not on regular intake of folic acid in 1st trimester. Consanguinity was reported only in 25.4%, while no consanguinity among parents in 74.6%. Spina bifida cystica is more predominant in illiterate and low education mothers (72%) while it was very low in highly educated mothers (P=0.008%), the level of education affect in awareness of the importance of folic acid intake and to avoid any drugs or radiation, especially in the first trimester. Spina bifida cystica were more common in rural areas (73%, N=89 cases) than in urban areas (27%, N=33 cases). No significant difference between normal vaginal delivery and caesarean section.

The majority of mothers took medication in their first trimester 92.5% (Antibiotics-valproate-methotrexate and others). Only in 8 babies, they had twins with the other twin neurologically free. More than one-third of cases (36%) born to primi-parous women, while multi-parous women were 64% of cases. No significant relation between the gestational age and occurrence of NTDs, full term babies with spina bifida cystica were 64% while pre-term babies were 36%. Incidental or occupational exposure to radiation increases the risk of NTDs. About 16% of mothers subjected to radiation during her first trimester, one of them were a nurse in the orthopedic department in our hospital subjected frequently to C-Arm radiation. Some drugs associated with a high risk of NTDs.

Positive family history with the previous baby with spina bifida cystica occurred in 9%. Positive neonatal history occurred in 61% of babies including: 66 babies with jaundice, 5 babies with convulsions and 4 babies with cyanosis. Half of the babies 51% (N=63) were breastfed while 19% were bottle only fed and 30% were mixed fed. House wives constitute 65% of mothers while 35% of women were employed. Complications during pregnancy were frequent (56.5%) including: 42 mothers diagnosed with anemia, 20 complained of hyperemesis gravidarum, 5 mothers with fits and 2 mothers had a history of fever. More than 80% of babies had associated hydrocephalus (N=102), 40 babies have associated orthopedic anomalies, 34 babies with cardiac anomalies, 21 babies with multiple congenital anomalies and 27 babies with no congenital anomalies. More than two-thirds of babies have good leg movement while one-third of babies were paraplegics.

Table 1. Summary of epidemiological data of reported cases in our study

Parameters		Meningocele	Meningomyelocele	Total
1) Gender	Male	23	43	66
	Female	16	40	56
2) Mother Age	15-ys	4	8	12
	20-ys	16	26	42
	25-ys	14	22	36
	30-ys	3	19	22
	35-ys	2	6	8
	≥40yrs	0	2	2
3) Parity	Primi-para	19	25	44
	multi-Para			
	p2	12	16	28
	p3	5	13	18
	p4	2	18	20
	p5	1	11	12
4) Gestational age	full term	26	52	78
	Pre-term	13	31	44
5) Complaint at pregnancy	hyper emesis gravidarum	6	14	20
	Fits	1	4	5
	fever	0	2	2
	anemia	16	26	42
	no comp.	16	37	53
6) Complications during delivery:	bleeding	0	8	8
	Obstructed Labor	2	4	6
	PROM	11	4	15
	Normal delivery	26	67	93
7) Twins:		2	6	8
8) Mode of delivery:	Normal vaginal	18	40	58
	CS	21	43	64
9) 1st trimester drugs intake	Valproate Antibiotics Methotrexate Other drugs	4 19 4 5	14 37 8 22	18 56 12 27
10) Folic acid intake during pregnancy	No	10	18	28
	irregular intake	12	22	34
	late pregnancy	17	43	60

Table 1. Summary of epidemiological data of reported cases in our study (continued)

Parameters		Meningocele	Meningomyelocele	Total
11) Other Risk factors during pregnancy:	radiation	8	12	20
	alcohol	0	0	0
	fever	12	31	43
	trauma	15	34	49
	smoking	4	6	10
12) family history	positive	4	7	11
	negative	35	76	111
13) Consanguinity	positive	13	18	31
	negative	26	65	91
	jaundice	12	54	66
	cyanosis	0	4	4
	convulsions	0	5	5
15) Mode of Nutrition of the baby	breast only	15	48	63
	bottle only	10	13	23
	mixed	14	22	36
16) Social state	rural	22	67	89
	urban	17	16	33
17) Education	illiterate	21	26	47
	Low	12	29	41
	moderate	5	28	33
	High	1	0	1
18) Occupation	housewife	29	50	79
	employee	10	33	43
19) Neurological examination of the fetus after birth:				
A) Leg move	Good leg mov.	39	42	81
	Paraplegic	0	31	31
B) Bladder	continent	39	33	72
	incontinent	0	49	49
C) Ant. Fontanel	Bulge	33	33	66
	Not	6	50	56
D) Associated Developmental anomalies	Hydrocephalus	44	58	102
	cardiac	13	21	34
	orthopedics	5	35	40
	multiple	0	21	21
	no anomalies	21	6	27

Discussion

This retrospective descriptive study was conducted on 122 patients in the neurosurgery department at Sohag university hospital between January 2009 and January 2015. Demographic data of the babies regarding baby's gender, type of spina bifid cystica, mother's age, presence or absence of consanguinity between parents, history of mother's drug intake, parity of the mother, gestational age, and mode of delivery were recorded.

In our study MMC cases were 68% and meningocele cases were 32%. Rankin collected 984 cases of NTDs including 50.2% spina bifida, nearly half of them were MMC cases (56%) and 44% cases were meningocele.[22]

There is a slight male predominance in our results where female represent 46 %. This matches with other studies including Houcher[15], David et al,[10] and Richard and Joseph,[23] who reported a figure of 52.4%, 57.7% and 52.4%respectively.

In our study the peak of maternal age is from 20-24 yrs (35 %) and 25-29 years (30 %) that match with other studies as Houcher et al,[15] and also David et al,[10] who reported figures of 20-24 years (26 %), 25-29 years (25 %) and 20-24 years (50.7 %), 25-29 years (27.2 %) respectively.

In our study the reported peak of maternal age was 35% in the age 20-24 yrs and 30% in the age 25-29 years. Similar results ws reporetd from other series including Houcher et al,[15] who reported 26% and 25%, and David et al,[10] who reported 50.7 % and 27.2 % for corresponding age groups. While we reported 10% of our mothers in the age group of 15-19 years, Houcher et al,[15] reported 4.3 % and David et al,[10] reported 16.2 %. Finally we reported 18% and 6% in the age group of 30-34 yrs and above 35 years respectively, Houcher et al,[15] reported 21.7 %, and 6%, and David et al,[10] 4.8%and 1 % for the same age groups.

In our study, there was an increased risk in primi-parous mothers as 36% of our cases belonged to primi-parous mothers. Similar results were also reported by others including David et al,[10], Robert J. Berry et al,[4] Northern China Region, and Moore et al,[20] Southern Region China who reported 57 %, 36.8%, and 42% respectively.

In this study, only 25.4% of spina bifida cystica cases happened to the babies of parents with consanguinity, On the other hand, other studies such as Afshar et al,[1] showed 50 % consanguinity and Houcher et al,[15] showed 57.1% consanguinity. Socioeconomic status including the education level of the mother and place of residence associated with a higher number of spina bifida cases, in this study, 38.5% of cases born to illiterate mothers, Robert J Berry et al,[4] Northern China Region reported 18 %, while, Moore et al,[20] reported 37 %, and one case only born to a highly educated mother. Rural residency has a higher number of cases (89 cases, 73%). Housewives were more than working mothers with a figure of 79 (64.7%) and 43 (35.3%) cases respectively. that match with some other studies where mothers are exposed to heavy manual work including, Robert J Berry et al,[4] Northern China Region who reported 87 %, and Moore et al,[20] Southern Region China who reported 64 %. In our case series, nearly whole mothers were not on regular folic acid intake in early pregnancy due to lack of a concept of giving folic acid in early pregnancy while in Salih et al.,[21] study showed that folic acid fortification leads to decrease in NTDs by 79%. In the United States (US), food fortification with folic acid leads to the marked decline of NTDs cases by 26%.[3] The Food and Drug Administration (FDA) recommends folic acid fortification to cereals, flour, and rice.[24] In Canada at which folic acid fortification is obligatory since 1998, a significant drop achieved in the number of NTDs cases.[11] On the other hand, in Chile after application of food fortified with folic acid, marked decrease by 31% occurred.[13] Since 1998, 50 countries started folic acid fortification programs.[9,14]

In our series, in our study, 58% of mothers

have associated diseases with pregnancy that is comparable to other studies, Afshar et al,[1] reported 50 %. In our study, 47.5% has a normal vaginal delivery that in line with other studies. In our study twins represents 6% that match with other studies[10] reported 2.1 %. In our study 66.4%of cases had good leg movement, while other 33.6% were paraplegic, 59% were continent as regard urine and stool (72cases P=0.00001), while 37% were incontinent. In our study 77.8% has associated other congenital anomalies, while in David Whiteman et al,[10] study, 79.1% has associated other anomalies.

Conclusion

Spina bifida cystica is common in our locality, Sohag governorate; specially associated with previously affected babies, low educated mothers and rural areas. Both genetic and environmental factors are direct leading causes. Folic acid fortification should be implemented to reduce the incidence of NTDs. Providing folic acid tablets for both parents is mandatory when pregnancy planned and when there is no contraceptive method.

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الملخص العربي

معدل انتشار مرض القيلة المائية في محافظ سوهاج، دراسة في المستشفى الجامعي

البيانات الخلفية: أمراض فتق العمود الفقري متعددة وترجع لعد تكوين القناة العصبية بشكل سليم بعض هذه الأمراض ينتج عنها الوفاة بشكل مباشر وبعضها الأخر قد ينتج عنه شلل أو ضعف بالأطراف السفلية وفقد على الإحساس بالنصف السفلي من الجسم وعدم القدرة على التحكم بالبول والبراز. يعد مرض القيلة المائية أشهر نوع من أنواع فتق العمود الفقري ويكون هناك كيس من الأغشية السحائية والأعصاب البارزة عن طريقة فتحة بجلد الظهر. مرض القيلة المائية يمثل مشكلة صحية كبيرة وسبب من الأسباب الرئيسية للوفاة في الأطفال ويمثل حوالي ٠,٠٣ في الولايات المتحدة ولا توجد إحصائيات دقيقة لنسبة حدوثه في مصر. إن هذا المرض يوثر على حياة المريض كما أنه يؤثر بشكل مباشر وغير مباشر على عائلته والأقارب المحيطين به. هذا المرض ينتج عن حدوث خلل أثناء الأسابيع الأولى للحمل ويكون مصاحبا لبعض الأمراض الخلقية الأخرى كثقوب القلب واستسقاء المخ.

الغرض: تتبع الصفات الشخصية للمرضى ومحاولة معرفة الأسباب المؤدية والمصاحبة لهذا المرض في محافظة سوهاج

المرضي و الطرق: دراسة وصفية لمائة واثنان عشرون طفلا مصابون بمرض القيلة المائية في مستشفى سوهاج الجامعي في الفترة ما بين يناير ٢٠٠٩ ويناير ٢٠١٥. تم تسجيل كل البيانات والمعلومات الخاصة بالأطفال وسؤال الأبوين عن الأسباب المحتملة لحدوث هذا المرض.

النتائج: عمر المرضى كان يتراوح ما بين عمر يوم إلى عمر ٨ شهور (المتوسط العمري شهرين). شملت الدراسة ٦٦ ذكراً و٥٦ أنثى. واحد وسبعون طفلا كانوا مرضى باستسقاء المخ، ٢٦ كانوا يعانون من أمراض خلقية بالقلب. تسعة وثمانون مريضا كانوا من مناطق ريفية.

الاستنتاج: فتق العمود الفقري مرض شائع بمحافظة سوهاج. كلا الجنسين يصابوا بهذا المرض بصفة متساوية تقريبا. ينتشر هذا المرض بصفة أكبر في المناطق الريفية والمولودين لأمهات قد أخذن علاج دوائي في الثلاث شهور الأولى من الحمل وللأمهات المولود لهم أطفال بنفس المرض سابقا والأمهات الذين لم يأخذوا أقراص حمض الفوليك بشكل منتظم.