Burden of Encephalocele in Tigray Region, Northern Ethiopia: Hospital-Based Study

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Abstract

Background: Encephalocele is a severe neural tube birth defect, which is associated with child mortality, morbidity, and lifelong disability. In Ethiopia, there is a need to understand the burden of encephalocele as surveillance data are lacking.

Objective: To determine the prevalence and clinical pattern of the encephalocele in the Tigray region, Northern Ethiopia.

Methods: Deliveries with encephalocele were identified from eight hospitals of Tigray region, Northern Ethiopia between October 2016 and June 2017. The clinical diagnosis of the infants affected with encephalocele was reported by senior obstetrics and gynecology specialist and the data were collected and recorded by trained midwives and mid-level emergency surgery and obstetrics professionals under the supervision of senior clinicians and biomedical researchers. Data were collected applying a standard data collection method across the participating hospitals, and analyzed using SPSS version 20. The prevalence rate of encephalocele was calculated per 10,000 live and stillbirths.

Result: A total of 10 infants were born with an encephalocele from a total of 14,903 live and stillbirths during the study period. The overall prevalence of infants with encephalocele was 6.7 per 10,000 live and stillbirths, and the majority (60%) of babies born with encephalocele were stillbirths. The highest rate (per 10,0000 live and stillbirths) of infants with encephalocele was observed in Kahsay Abera Hospital from Western Zone (31.2) and Adigrat Hospital from Eastern Zone of Tigray (28.9). The commonest type was found as occipital encephalocele (70%) followed by nasal frontal type (30%).

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Conclusions: This multi-center study covering eight hospitals in Tigray region of Northern Ethiopia provides important insights on the prevalence of encephalocele. We found that encephalocele cases are encountered in hospitals in Ethiopia, and resources are needed to address these cases and improve their outcomes. Studies of this nature can provide reasonable information on a relatively rare condition, especially when resources are lacking to conduct large population-based birth defects surveillance. Future studies can investigate the prevalence using population-based surveillance approaches.

Keywords: Encephalocele, Surveillance, Hospital-based, Occipital, Nasal, Frontal, Skull defects, neural tube defects.

Introduction

Encephalocele is one of central nervous system malformations characterized by herniation of the brain tissue or meninges through a skull (1,3). It is associated with intellectual impairment, child mortality, morbidity, and lifelong disability (1, 2). Encephalocele is clinically characterized by a fluctuant, round, balloon-like mass that protrudes from the skull. These masses of deformed neural tissue could be palpable and be covered by an erythematous, translucent, or opaque membrane, or by normal skin (3). Encephalocele may consist of only meningoceles, contain brain tissue (meningoencephalocele or encephalocele) and include a ventricle (hydroencephaloceleningocele) (4) and these types of skull defects are fatal, (5).

Encephalocele commonly occurs in occipital (75%), followed by frontoethmoidal (13% to 15%), parietal (10% to 12%) region of the skull respectively (6). The occipital region was found to be the most common site of encephalocele in the United States and Europe, while fronto-ethmoidal encephaloceles are prevalent in Russia and Southeast Asia (7-10). Scientifically, the etiology of encephalocele is not well established and the causes of this condition remain indescribable. (11, 12).

It is estimated that the prevalence encephalocele is approximately 0.8–5.6 per 10,000 live births (3, 13, 14). In the United States, it is estimated that approximately 0.82 per 10, 000 live births affected with encephalocele born each year (14). Report from Texas has shown that during 1999–2002, the prevalence of encephalocele was 0.86/10,000 live births, and female were predominantly affected

compared to males (15). There is a scarcity of knowledge on the prevalence of encephalocele in Ethiopia, where recent studies have found a very high prevalence of other neural tube defects (16, 17). This information is vital to develop evidence-based prevention strategies. The current study aimed to determine the prevalence and clinical pattern of encephalocele in the Tigray region, Northern Ethiopia using hospital-based surveillance.

Methods

The detailed description of the study methodology is available elsewhere (16). Briefly, all deliveries in our study were identified prospectively between October 2016 to June 2017 at eight hospitals of Tigray, Northern Ethiopia. Approximately 57% percent of births to Tigray residents occur in a health facility (18). The eight hospitals included in the study were Mekelle and Ayder from Mekelle Zone, Lemelem Karl from Southern Zone, St. Mary from Central Zone, Sihul from the Northwestern zone; Adigrat and Wukro from Eastern zone and Kahsay Abera from Western Zone of Tigray region. These hospitals are the biggest public hospitals in the Tigray region, serving populations with diverse socio-demographic characteristics as well as health-related behaviors. These hospitals have a general pediatric ward, pediatric critical care unit, obstetrics, and gynecology wards. The hospitals also provide 24-hour obstetrics and gynecology care.

Prospective registrations of all birth outcomes with any type of encephalocele presenting to the usual catchment area of the study hospitals within the study period were conducted. A case reporting format was used to collect data from newborns affected with an encephalocele. All live births and stillbirths were clinically evaluated for any externally visible congenital malformations, the anatomical location, typology, and their characteristics. For all patients admitted to the hospital, demographic, admission, and treatment information was collected. The clinical diagnoses were conducted by senior obstetricians and gynecologists. Data were collected by the trained midwives and mid-level emergency surgery and obstetrics professionals under the supervision of senior clinicians and biomedical researchers. For purposes of this study, newborns with a clinical diagnosis of encephalocele were eligible for inclusion. An encephalocele case was defined as any reported diagnosis of frontal, nasal, frontal-nasal, occipital encephalocele and encephalocele unspecified. The stillbirths included in this study were mainly from spontaneous

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abortion or miscarriages. Counting stillbirths were started from gestation age 20 weeks. Cases were included in the analysis if they met the case definition for an encephalocele, and were permanent residents of the catchment areas of the hospitals. Birth outcomes that are not stillbirths or live births meaning elective terminations and or early spontaneous abortions were excluded from the analyses. Fortunately, we did not find any cases of elective terminations or early spontaneous abortions.

Statistical data analysis

The data collected was coded, cleaned and analyzed using Statistical Packages for Social Sciences (SPSS version 20). Simple descriptive analyses (frequencies and percents) were conducted to characterize study subjects. The prevalence of encephalocele was calculated per 10,000 births based on birth outcomes (live births and stillbirths). Overall birth prevalence of encephalocele is defined as number of cases of livebirths or stillbirths with encephalocele after 20 weeks of gestation (numerator) among a total number of livebirths and stillbirths during the study period in the denominator. The Poisson distribution was used to calculate for prevalence estimates and 95% confidence intervals (CI).

Ethical consideration

Prior to the beginning of the data collection, appropriate ethical clearance was obtained from Mekelle University, College of Health Sciences, Health Research Ethics Review Committee (registration number: ERC 0837/2016). The study was conducted in accordance with the ethical principles of health research involving human subjects.

Results

During the study period, a total of 14,903 deliveries occurred in the eight hospitals. Of which, a total of 10 infants were recorded to have an encephalocele. The overall prevalence of encephalocele was 6.7 per 10,000 births (live birth and stillbirth) (95% CI, 3.2–12.3). When the encephalocele data were further disaggregated, 31.2, 28.9 and 7.4 per 10,000 births were observed in Kahsay Abera, Adigrat and Wukro hospitals, respectively (table 1). There were no cases in the other hospitals in the surveillance.

Table 1: Hospital-based prevalence of encephalocele in Tigray Region, Northern Ethiopia, 2017 (n = 14,903).

Administrative zone of Tigray region	Representative hospital	Total deliveries registered	Total Ence identified (n)	Ence per 10,000	Ence per 10,000 births		
					95% CI Poisson	LB	SB

				births	LL	UL	Ence	Ence
Mekelle	Ayder	2401	-	-				
	Mekelle	3529	-	-				
South	Lemelen Karl	889	-	-				
Western	Kahsay Abera	961	3	31.2	6.4	91.2	20.8	10.4
Central	St. Mary	1685	-	-	0.0	21.9	0.0	0.0
Eastern	Adigrat	2075	6	28.9	10.6	62.9	9.6	19.3
	Wukro	1358	1	7.4	0.2	41.0	0.0	7.4
Northwest	Sihul	2005	-	-				
Total		14903	10	6.7	3.2	12.3	2.7	4.0

Ence: Encephalocele; LL: Lower limit; UL: upper limit; LB: Livebirths; SB: Stillbirths

Anatomical location of encephalocele

The commonest types of encephalocele cases were occipital (70%) followed by nasal frontal encephalocele (30%). Moreover, omphalocele was found to be associated with encephalocele cases (table 2).

Typology of the encephalocele

Ruptured occipital (60%), followed by cystic nasal frontal (30%) and cystic occipital encephaloceles (10%) were identified in the study cases (table 2). Physical evaluation of the cystic nasal frontal encephalocele shows a bluish, soft, compressible mass, or intranasally with possible distortion of surrounding facial characteristics.

Characteristics of the encephalocele

The gestational age of the majority of the newborns affected with encephalocele was very preterm, followed by full term, moderate preterm, and early preterm. Males cases were predominated females. Majority of babies born with encephalocele were stillbirths. (table 2).

Table 2: Anatomical location, typology, and characteristics of encephalocele among patients from public hospitals in Tigray Region, Northern Ethiopia, 2017, (n=10).

Anomalies	Frequency	Percentage
Location		
Posterior (occipital) encephalocele	7	70
Nasal frontal encephalocele	3	30
Nasal	-	-
Parietal	-	-
Not specified	-	-
Salient characteristics of the observed encephalocele		
CysticNasal frontal encephalocele	3	30
Cystic occipital encephalocele	1	10
Ruptured occipital encephalocele	6	60
Associated anomalies		
Yes Omphalocele	2	20
No	8	80
Gestational age of the identified encephalocele cases		
Extremely preterm (<28 weeks)	-	-

Very preterm (28-31 week)	4	40
Moderate preterm (32-36 week)	2	20
Early term (37-38 week)	1	10
Full-term (39-40 week)	3	30
The sex of the observed infant affected with encephalocele		
Male	8	80
Female	2	20
Pregnancy outcome		
Live birth	4	40
Stillbirth	6	60

Discussion

This study has shown that the prevalence of encephalocele in the Tigray region of Northern Ethiopia was 6.7 per 10,000 live and stillbirths. Though this study is facility-based, studies of this type are important in resource poor-settings where often little or incomplete data are available, but where the public health burden is often the highest. Without this knowledge, governmental and non-governmental agencies cannot make data-informed public health policies or recommendations.

The burden of encephalocele observed in our study is higher than other studies done elsewhere in world (3, 13, 14, 19). A very high burden of encephalocele was observed in Adigrat Hospital (28.9 per 10,000 births) and Kahsay Abera hospital (31.2 per 10,000 births). The prevalence noted in the current study is higher than the report from Southern Alberta (0.00–3.8 per 10,000 live births) (20), Latin America (1.5/10,000 live births) and Mexico (3.1/10,000 live births) (19) and less than the report from Southern India (8.5/10,000 live births) (21). Moreover, the prevalence of the current study is much higher that reported from Atlanta (1.4 per 10,000 live births) (22). Our findings may not be directly compared to other studies due to variation in study designs; however, the high prevalence of encephalocele noted in Tigray is not unexpected given high prevalence of nutritional deficiency and lack of preconceptional care that can impact the formation of nervous system in the current study population.

Posterior (occipital) and nasal frontal encephaloceles were the common types of encephalocele identified in this study. Similar findings were reported from North America and Western Europe, where the occipital encephaloceles accounting for 0.3 – 1 per 10,000 live births and they comprise 85% of all encephaloceles (23, 24, 25). Likewise, a study done in Kenya reported that the occipital encephaloceles

were the most common type, contributing to 53% of all encephalocele cases and the anterior or sincipital group contributed only 33% (26). A similar pattern has been reported in Nigerian studies with 69% of lesions in the occipital region (27, 28).

In contrast to our study, anterior encephaloceles were the common types of encephalocele in South-East Asia, Russia, and Central Africa – with a prevalence of 2 to 2.8 per 5,000 births (29). The reason given for the predominance of anterior or posterior encephaloceles is not well known. But this might link with different mechanisms that are involved in the genesis of encephalocele or brain development.

The present study, majority of the observed encephalocele cases were preterm infants. The increased rate of encephalocele among low birth-weight and preterm infants has been described in previous studies (30, 31).

Our finding has shown that majority of babies born with encephalocele were stillbirths. The risk of mortality for infant with encephalocele is the highest during the first day of life, continuing to occur through adolescence and is influenced by several clinical and demographic characteristics, including the site of the defect, the contents of the sac, lower birth weight, and other associated birth defects (1, 32).

The present study, showed that males were more susceptible to encephalocele compared to their female counterparts. This finding is in contrast with reports from North America, Western Europe, and Central Africa countries that showed the predominance of encephalocele in females (7-10, 17, 29). The exact reason for male preponderance is unknown, and there may be genetic factors involved, which require further investigation (11, 22, 32).

Strength of the study

One of the strengths of this study is that all deliveries were carefully assessed and data were collected at a postpartum or postabortal period and before the discharge of the women by trained midwives and mid-level emergency surgery and obstetrics professionals under the supervision of senior clinicians and biomedical researcher. This made it possible to capture all encephalocele, with high case specificity. among stillbirths or live births in the study hospitals. Another, strength of this study is the involvement of multiple public hospitals, representing the majority of deliveries to Tigray region, northern Ethiopia residents and covering people with a diverse health related behavior.

Limitation of the study

The study only included hospital-based deliveries. This may have either led to an underestimation or overestimation of birth prevalence of encephalocele. With about 60% of the births in the Tigray region occurring in health facilities (19), our findings should be interpreted with caution. Several important clinical patterns of the encephalocele were not identified in this study due to constrained prenatal diagnosis. Detailed demographic characteristics of the mothers, who have given birth to fetuses with encephalocele were not sought. Furthermore, this study may also suffer with selection problems as the study depends on facility-based deliveries.

Conclusion

Our study has shown a high burden of encephalocele in northern Ethiopian region of Tigray. The majority of the newborns with encephalocele were stillbirths suggesting that encephalocele is contributing to perinatal mortality. Though this study is facility-based, studies of this nature are very important, especially in in resource poor-settings where often little or incomplete data are available, but the public health burden is often believed to be high. Future studies should examine the prevalence using population-based surveillance approaches.

Declarations

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Availability of data and materials

All relevant data are within the paper.

Authors' contributions

BAB has designed the study and put a substantial contribution to carry out the collection of data, and the statistical analysis. HKM, ALW, YB, TM, SA, KG and AM also participate in study design and coordination and helped to draft the manuscript. All authors read and approved the final manuscript.

Competing interests

The authors declare that they have no competing interests. All authors of the manuscript have read and agreed to its contents.

Consent to publish

Not Applicable.

Ethics and consent to participate

Appropriate consent was obtained from Mekelle University College of Health Sciences; Health Research Ethics Review Committee (ERC 0837/2016). Consent was obtained from the participating mothers in written form.

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