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Case Report

Anencephaly: A Case Report in Aného Hospital (Togo)

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Abstract

Anencephaly is a severe anomaly of the brain that results from the failure of the cephalic part of the neural tube to close during the fourth week of pregnancy. In anencephaly, the mortality rate is 100% during intrauterine life or within hours or days after birth. Its prevention is based on supplementation with folic acid during pregnancy. We report a case of complicated anencephaly with death of the fetus in the womb of a young woman who didn't take folic acid during her pregnancy.

Keywords: Anencephaly, neural tube, pregnancy, folic acid.

INTRODUCTION

Anencephaly is a severe malformation of the central nervous system, being the most common type of neural tube defect. The brain stem, cerebellum and diencephalon are usually present (Santana MVMC *et al.*, 2016). It results from the failure of the rostral neuropore to close during the fourth week of development (Moore KL *et al*., 2008). Anencephaly is still a common lethal anomaly. It occurs in 1.0–4.7 per 1000 births and is more common in females (female: male ratio of 4:1) than in males (Santana MVMC *et al.*, 2016; Sadler TW, 2011). It is the major cause of fetal loss and disabilities in newborns and it is considered as a significant public health problem. It is associated with substantial mortality, morbidity, and psychological costs (Nasri K *et al*., 2014; Anyanwu LC *et al*., 2015). We aim to remember through a case report of anencephaly in Togo, the prevention of this anomaly by folic acid.

OBSERVATION

It concerned a female newborn from a presumed full-term pregnancy in a 31-year nulliparous primigravida with unfavourable socioeconomic conditions.

There is no notion of consanguinity or history of spontaneous abortion or miscarriage. During the pregnancy, the pregnant had no prenatal consultation and therefore had no prophylaxis, namely with folic acid. She was admitted to the obstetrics department in labour with excessive fundal height. The delivery was made vaginally and enabled the extraction of a fresh stillborn baby weighing 3150 g, whose examination enabled to notice:

- An absence of the calvaria, giving a glimpse of a necrotic-looking rudimentary ancephalic tissue, covered with multiple blood clots. This defect of the cranial vault does not reach the spine.
- Eye sockets pointing upwards, with prominent eyebrows arches and bulging, drooping and tragic eyes.
- The forehead was almost nonexistent, the low ears set with large auricles and badly hemmed. The neck was short.

All the malformations of the cephalic extremity gave a batrachian aspect to the head of the newborn and enabled us to retain the diagnosis of complicated anencephaly with death in utero (figure).The clinical examination did not notice any other malformations.

Figure: Anencephaly with absence of the cranial vault and the aspect of a batrachian head

COMMENTS

The process of neural induction is the first step of neurulation, while the last step will be the closure of the caudal neuropore, by the end of the 4th week (Sadler TW, 2011; Schoenwolf GC *et al*., 2008; Moore KL, 2006). Neurulation is the formation of the neural plate and the neural tube and begins in days 22–23, close to the 4th to 6th pairs of somites (Moore KL, 2006). The closure of the cranial neuropore takes place at the 18- to 20-somite stage (i.e., approximately the 25th day) and starts from the initial closure site in the cervical region, while the closure of the posterior neuropore takes place approximately two days later, at the 25- somite stage (i.e., 27th day). If the rostral neuropore of the neural tube fails to close during the 4th week, exencephaly will appear (Sadler TW, 2011; Moore KL, 2006). Anencephaly is always associated with a crania and, as the calvaria is absent, the brain will not be protected and it will degenerate, but the brain stem will remain unaffected (Sadler TW, 2011; Moore KL, 2006). Moreover, the destructive process also appears because of the abnormal brain structure and vascularisation, defined by formation of new blood vessels (Moore KL, 2006) [8]. On macroscopic evaluation, the remaining part of the brain has a spongy aspect, forming a vascular mass composed almost of hindbrain structures (Moore KL, 2006) [8]. Anencephaly has an incidence of 1 to 5 in every 1000 births, and the mortality rate is 100% during intrauterine life or within hours or days after birth (Santana MVMC et *al.*, 2016; Sadler TW, 2011) [1, 3]. In United States, about one in every 4600 babies is born with anencephaly (Mai CT *et al*., 2019). Geographic variation is a factor to be taken into consideration: countries such as Mexico, China, Turkey or British Isles have a high rate of prevalence (Ekmekci E et *al*., 2019). The pooled birth prevalence of anencephaly in Africa was 0.14% (95% CI: 0.12, 0.15%) (Oumer M *et al*., 2021). Higher burden of anencephaly was detected in Ethiopia (0.37%, CI: 0.15, 0.58%), Algeria (0.24%, CI: 0.24, 0.25%), and Eritrea (0.19%, CI: 0.19, 0.19%) (Oumer M *et al*., 2021). The higher pooled prevalence of anencephaly was observed in the studies that included both live births and stillbirths (0.16%) (Oumer M *et al*., 2021).

The mortality rate is 100% during intrauterine life or within hours or days after birth and the termination of pregnancy percent is greater than 83% (Johnson CY *et al*., 2012).The etiology of anencephaly remains unclear, but various maternal-related environmental and genetic risk factors have been reported, which include diabetes, obesity, exposure to different drugs or toxins, genetic polymorphisms and mutations, as well as positive family history for neural tube defects. One of the most important nutritional factors in the development of anencephaly is folate deficiency. Supplementation with folic acid represents a major public health issue, reducing the risk of neural tube defects (van Gool JD *et al*., 2018).

The diagnosis of anencephaly can be made in utero with a high degree of certainty according to following factors (Munteanu O *et al*., 2020): alpha-fetoprotein level is elevated in the maternal serum and amniotic fluid; absence of the upper portion of the cranial vault, there is either no tissue or an ill-defined mass of heterogeneous density above the level of the orbits, hydramnios may complicate these pregnancies. In postnatal period, the appearance of the infant with anencephaly is unique, and the diagnosis can be made certainly when all following criteria are met: a large portion of the skull is absent; the scalp, which extends to the margin of the bone, is absent over the skull defect; hemorrhagic, fibrotic tissue is exposed because of defects in the skull and scalp; recognisable cerebral hemispheres are absent (Munteanu O *et al*., 2020).

When newborns with anencephaly are alive during few hours or days, there is no curative treatment.

CONCLUSION

Anencephaly is the most lethal neural tube defect. Its prevention is therefore important and based on supplementation by folic acid during the first trimester of pregnancy. Otherwise diagnosis in utero with perfomant ultrasonography in hand of

experimented radiologist is encouraged to avoid psychological complication on the mother when pregnancy is not stopped.

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