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Brief Review

Medicolegal Aspects of a Case of Anencephaly

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ABSTRACT

The term 'anencephaly' is commonly used for the serious developmental defect of the central nervous system, in which the brain and skull are completely or partially absent. The cases of anencephaly are not uncommon in the Department of Gynecology and Pediatrics but it is rarely noticed in practice of forensic medicine. One such rare case of anencephaly was brought to us in the Department of Forensic Medicine at Government Medical College, Vadodara (Gujarat), for autopsy examination in which the death was alleged due to malnutrition. After autopsy examination, we concluded that it was a case of anencephaly in which the brain was almost completely absent in the thick and malformed skull. The case is presented here with the aim to discuss its different medicolegal aspects as it is rare of its kind in our field.

Keywords: Anencephaly, Neural tube defect and Autopsy

INTRODUCTION

Neural tube defects (NTDs) are the congenital disorders, which occurs due to defective development of the tissues, which grows into the brain and spinal cord. They are considered as a complex congenital disorder because they are caused by a combination of multiple genes and multiple environmental factors. The prevalence of NTDs varies in different geographic areas. According to Duke Center for Human Genetics, the chances of having NTDs are 1 in 1000 population in the United States of America, however, once a couple has one child with an NTD, their chance of having a second child with an NTD is increased to approximately 2–5%[1].

NTDs are one of the most common types of congenital malformations in India and anencephaly is one of the most severe type of it. Anencephaly means 'no brain' but this terminology is commonly used for the newborns who are not having forebrain, which includes the largest part of the brain consisting mainly cerebral hemispheres. It is basically due to an abnormal development of the brain, results from a NTD that occurs when the rostral (head) end of the neural tube fails to close with the lumber end resulting in the absence of a major portion of the brain, skull and scalp[2]. Anencephaly more commonly affects female fetus than males. Without a functioning cerebrum, anencephalic infants cannot gain consciousness so most anencephalic infants are either stillborn or die within a few hours or days after birth. Sometimes baby born blind, deaf, unconscious and insensitive to pain due to lack of brain and some may have a rudimentary brainstem that permits reflex actions such as breathing and response to sound or touch. In the present case, dead body of an unknown female newborn aged about one day was brought to us for autopsy examination with an alleged history of death due to malnutrition. During investigation it was found that the baby was recovered from an open ground in an alive state and referred to our centre for further treatment, where she was admitted to the pediatric Neotal Intensive Care Unit (NICU) for about 3 h duration before death.

CASE HISTORY AND PM EXAMINATION

An unidentified newborn female child was recovered by the police from an open ground of a rural area near Vadodara (Gujarat). The baby was immediately brought to the nearest hospital, where she was alive and after initial treatment referred to the S.S.G. Hospital, Vadodara, for further management. On arrival in the emergency department patient was alive and conscious with normal pulse and blood pressure. She was admitted to the pediatric NICU and survived about 3 h before the terminal event of death. As it was a medicolegal case, the body was transferred to the forensic medicine department for autopsy examination, which was conducted in next day morning. The police officer narrated the cause of death 'malnutrition' in inquest papers. During autopsy, we found that the dead body was an unidentified newborn female child with fully developed rigor mortis and reddish blue discolouration over face and lips. Vernix casiosa was absent over the body and patechial haemorrhages were present at places. Nails were projected beyond the finger tips in both hands. Length of the baby was 45 cm and weight was 2050 g. The stump of the umbilical cord was 3.5 cm in length with the clean cut and well-clamped end. She was having a malformed skull with almost absent forehead and slightly protruded eyeballs, due to which the frontal hair line came very close to the eyebrows, as shown in photograph-1. The scalp hairs were black and 3–4 cm long and protruding angiomatus tissues were present in mid parietal areas of the scalp. Fontanelles were absent and the skull vault was uniformly thick about 0.5 cm and calcified. Cranial cavity and brain tissues were almost completely absent below the thick cranial vault. Lungs were fully extended, spongy and crepitant and were positively responding to hydrostatic test. The stomach contained about 20 ml half curdled whitish milky material. All other viscera were appearing normal. Ossification centres for the lower end of femur, upper end of the tibia and cuboid were present. After autopsy, it was concluded as a case of anencephaly and death was attributed to cardio-respiratory failure resulted from anencephaly. Later the baby was identified as a newborn of a young female who delivered the child at home after a full-term normal delivery and due to her monster appearance she was thrown into an open ground from where it was recovered by the police.

Photograph-1

DISCUSSION

Anencephaly is a congenital malformation of NTD, in which there is a complete or partial absence of scalp, calvarium and normal brain, which is replaced by an angiomatous mass[3], as that was present in this case also (as shown in Photograph-2). The eyes slightly bulged out because the frontal bones were absent and the orbits were shallow. Though such kind of cases are common in gynecology and pediatric departments but they are rarely noticed in practice of forensic medicine, probably because of two reasons: first, they can be easily diagnosed by ultrasonography and maternal alpha fetoprotein screening during pregnancy and an indication of medical termination of pregnancy and second due to their delivery as a stillbirth, which is a natural cause of death. In the present case, no routine antenatal sonography or screening was done to rule out the congenital defects and the pregnancy was carried out to full term, which ended into the normal vaginal delivery of an anencephalic child.

Most of the anencephalic infants are stillborn, but in the

Photograph-2

present case baby was alive after birth and referred to the higher centres for further treatment where she was admitted into the pediatric NICU for 3 h before death. The autopsy findings of respired lungs, positive hydrostatic test and half curdled milk in the stomach also corroborated the same that the child was not only born alive but also had lived for some times after birth. Absence of vernix caseosa over the body was also indicated that the baby was washed after delivery and suggesting that she survived for some time after birth. In this case, brain was almost completely absent in the rudimentary cranial cavity and the continuation of life after birth might be due to some functioning brain activity, which is also known as persistent vegetative state[4].

Another aspect of this case was that the 'Rule of Hasse' was not useful to us in determining the approximate age of this fetus, because of the presence of congenital malformation in the form of anencephaly, so the other parameters of age estimation like the presence of ossification centres for the lower end of femur, upper end of tibia and cuboid with nails projecting beyond the fingers of both hands were taken into consideration to conclude the approximate age, e.g., full term[5].

A baby born with anencephaly looks like monster and mostly born as still birth or may born blind, deaf, unconscious and insensitive to pain, which is a common reason of the unacceptance of such child after birth, which was there in this case and the parents left her in an open ground for the purpose of disposal. When such newborn infants, after it is born alive, are exposed in any place with the intention of abandoning it, and death does not supervene, the parent or person responsible for the care of such infant is guilty under section 317 of the Indian Penal Code 1860, and may be punished with imprisonment of either description for a term which may extend to 7 years, or with fine, or with both[6]. In the present case, mother also can be charged for an act of omission because she did not take any medical help during pregnancy and even after delivery did not try to protect her from the heat and cold and also did not supply her proper food and left the child at open ground with the intention of abandoning it.

Though anencephalic neonates usually born stillbirth but when they born as live they face certain and imminent death and such kind of cases of anencephaly can be a better source for the pediatric organ transplantation but there are legal and ethical issues according to the laws of the land. Some authors have proposed that the organs of anencephalic neonates can be used for organ transplantation after taking written informed consent from the parents and if there is compliance with the guidelines of the authority for organ transplantation in that particular area[7].

CONCLUSION

The cases of anencephaly are rare in practice of forensic medicine and that is why the information regarding its medicolegal aspects are also not discussed by most of the authors who have commented on it. The case was presented here with the aim to discuss the medicolegal aspects of the anencephaly and to highlight the need to review the criteria related with brain death so these anencephalic infant can be useful to meet the high demand of suitable donor for transplantation in infants and children.

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