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Holoacardius Hemisomus Acephalus

— A case report —

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ABSTRACT

An autopsy case of holoacardius hemisomus acephalus is reported. She weighed 2,190 gm and the height was 38 cm. The head and upper extremities were absent, while the vertebrae and lower extremities were relatively well developed, but severely edematous. The heart, lungs, stomach, liver, spleen, and pancreas were missing, but the lower abdominal organs including kidneys, adrenal, urinary bladder, and genital organs were present. The intestine was blind-ended at jejunal level but opened into a normal anus. The umbilical cord had two arteries and one vein.

Key Words: Acardius, Heart, Twin, Malformation

INTRODUCTION

Acardius is one of the most severe and rare congenital malformations. The occurrence rate is estimated to be less than 1 in 34,600 deliveries. There is an acardiac in approximately one percent of monozygotic twin pregnancies and in 1 of 30 monozygous triplets[1]. Acardius is commonly associated with acephaly and failure of development of most of the thoracic organs and upper limbs. We experienced an autopsy case of holoacardius hemisomus acephalus and report it in view of it's rarity.

CASE REPORT

This fetus was born to a 32 years old woman as a twin. The mother had experienced two episodes of spontaneous abortion. During this gestation, ultrasonography showed that this was a twin pregnancy and only one head was present. Because of hydramnios and associated anomalies, termination of pregnancy was done at 33 weeks of gestation. An apparently normal baby was delivered first vaginally. It was a female baby weighing 1,600 gm. The second baby was an acardiac twin delivered via Cesarean section due to dystocia. The first baby was alive and unremarkable.

Pathological examination

This female fetus weighed 2,190 gm and top-heel length was 38 cm. Severe anasarca was present. On the left side of the midbody, poorly developed facial organs were present. The tongue with blind oral cavity was present. Upper extremities and head were absent, but vertebrae and lower extremities were relatively well developed (Fig. 1). An omphalocele was noted in the umbilical cord region. In the pubic area, labia and anus were present. On infantogram, there was a diffuse hazy density

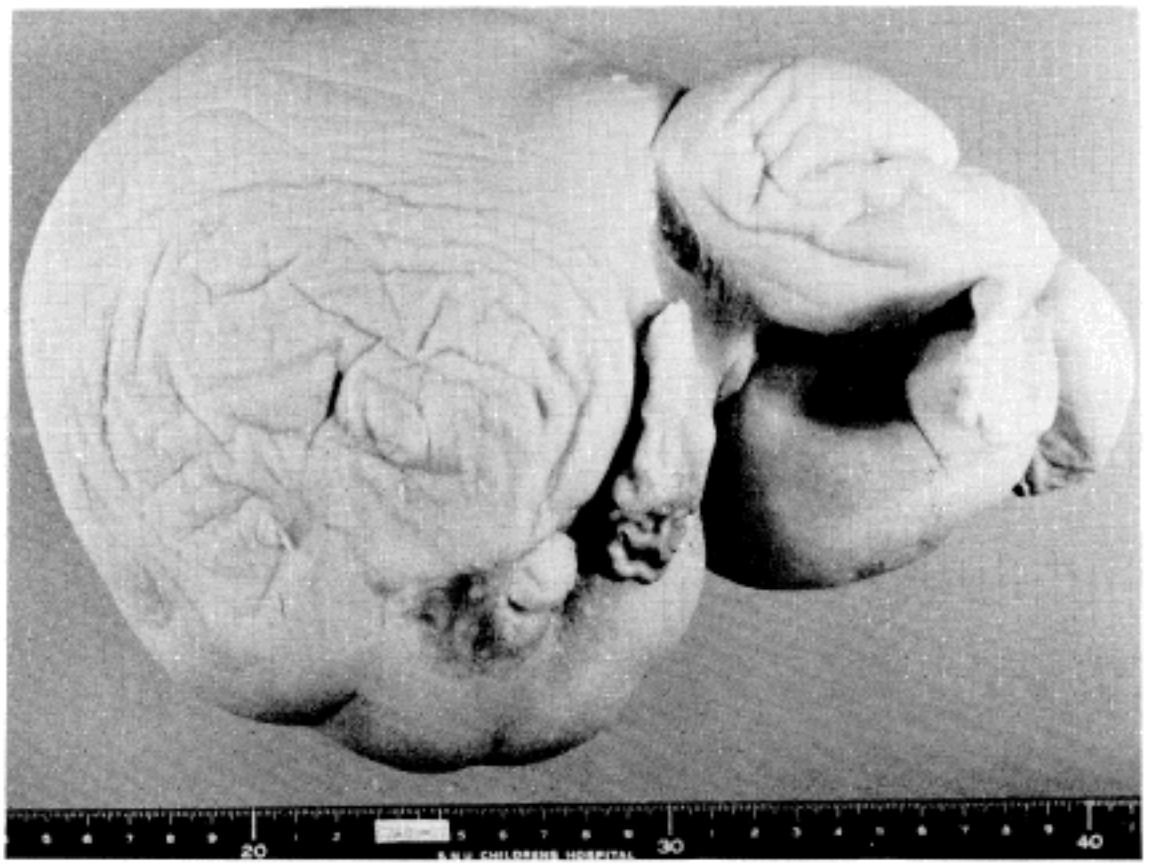

Fig. 1. Anterior View
It weighed 2,190 gm and top-heel length was 38 cm. Absence of head and upper extremities and fetal hydrops are seen.

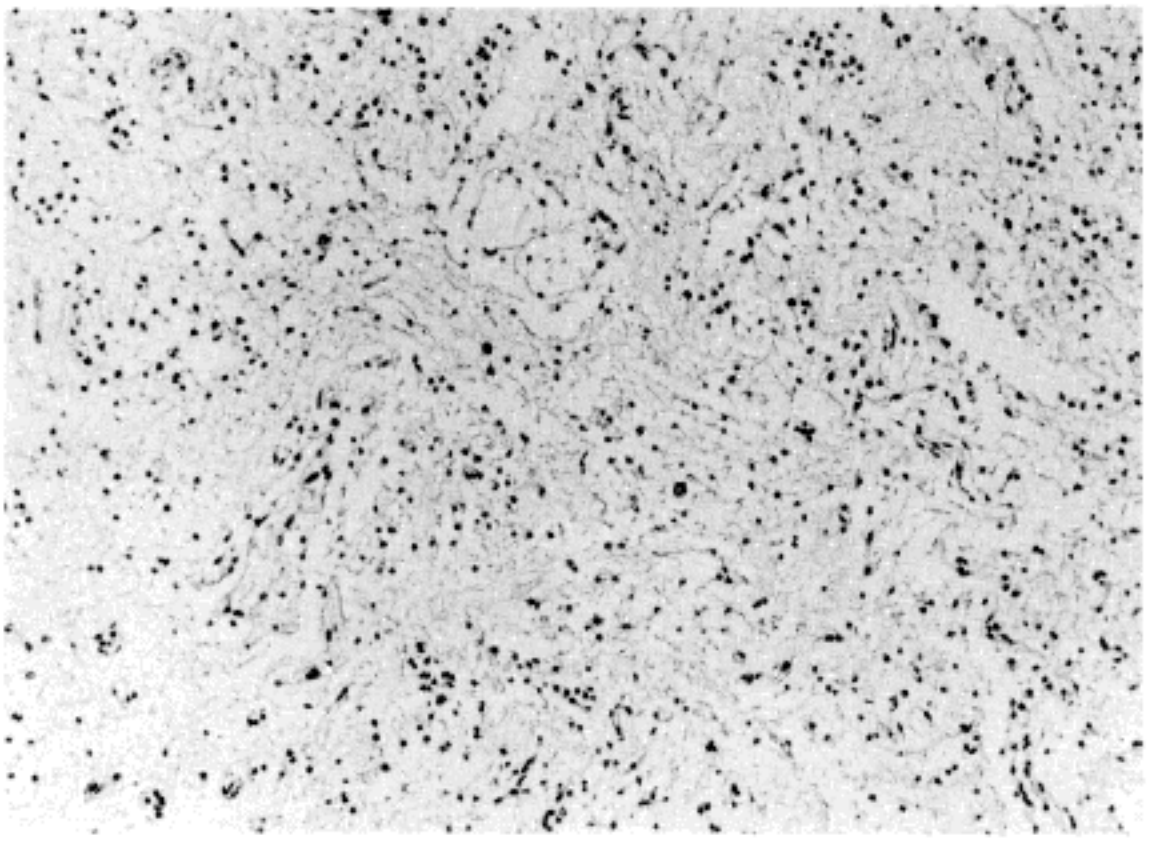

Fig. 2. Microscopic view of glial nodule (×100).
A round mass, measuring 0.5×0.3×0.3 cm, beneath the skull base consisted of glial cells with scattered calcifications.

through the whole body. The vertebral column was severely bent, convex to the left side, and pairs of ribs were also found. Rudimentary skull base was present. On section, a large amount of orange yellow clear fluid escaped from many loculated spaces in subcutis. On opening the abdominal cavity, the liver was absent and a large right kidney and an adrenal were seen. The left kidney was absent. Two ureters were attached to the right kidney. The pancreas and spleen were also absent. The intestine was 40 cm in length and blind-ended proximally. In the thoracic cage, there were no lungs, heart, and esophagus. On the top of the spine, soft brain-like tissue was present amounted about 3 cc. A round mass, measuring 0.5×0.3×0.3 cm, was recognized beneath the rudimentary skull. Microscopically it consisted of glial cells with

scattered calcifications(Fig. 2). On the cervical spinal cord, translucent fluid-filled arachnoid cyst, measuring 0.6×0.2×0.2 cm, was recognized. By specimen radiograph, skull and facial bones were not formed. There were two teeth in superior and inferior portions of the mouth, respectively. On the maxillary side, a cleft-like structure reminiscent of Rathke's pouch was recognized, measuring 0.4 cm in length. The placenta was not submitted. There were two umbilical arteries and one umbilical vein. Systemic vessels could not be searched due to severe anasarca and vascular collapse.

DISCUSSION

The most popular classification of acardius was developed by Das[2]. This case belongs to holoacardius hemisomus acephalus, the most common type. Hemisomus means partial development of somites. Holoacardius is used to designate the absence of the heart and to distinguish this condition from hemiacardius, in which there is a remnant of nonfunctioning heart muscle. The prerequisites for the formation of an acardiac fetus are a functioning cardiovascular system of the second normal fetus and a good vascular communication between the twins. For these reasons, acardius is only seen in monochorionic twin gestations, apart from a few case reports in diamniotic-dichorionic twins[3]. The reduced member depends for its circulation wholly or largely on the heart of the normal twin through the abnormal anastomoses of their umbilical vessels in or close to the common placenta. The blood flow in the umbilical and other main vessels of the malformed twin is in a reverse direction, deoxygenated blood from an umbilical artery of the normal twin entering the acardiac one by its umbilical artery and leaving it through its umbilical vein[4]. In over 50% of cases, the umbilical cord of the acardiac twin contains only a single umbilical artery accompanying the vein instead of the normal arrangement of two arteries with the single vein[5].

In the case presented there was dystocia of the acardiac twin necessitating Cesarian section. We have no information on the cardiomegaly of the other twin until the time of termination at 33 weeks. A small nodule of glial tissue in the top region was of interest particularly in such a case who had no head part at all. This really indicates that there was some resorptive mechanism involved in the pathogenesis of headless monster like this case.

참 고 문 헌

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＝국문초록＝

무심태아

— 1증례보고 —

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무심태아 1례를 부검하여 보고하였다. 환아는 여아였고 무심태아중 가장 흔한 형태인 *holoacardius hemisomus acardius*에 속하였다. 전신에 심한 부종이 있었으며 양상지가 없었으나 척추와 양하지는 비교적 잘 발달되어 있었다. 내부장기를 살펴보면 심, 폐, 위, 간, 비장, 췌장이 없었고 소장은 근위부에서 막혀있었고 원위부로는 정상이었다. 오른쪽 신장과 하나의 부신이 있었다.