

Dicephalus dibrachius dipus: A case report

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Abstract

Dicephalus twinning is an uncommon condition. We report a dicephalus twin with a neural tube defect and her autopsy finding.

Key words: Dicephalus, Twinning, Neural tube defect

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Introduction

In human pregnancies, twins occur with a frequency of 1 in 90 births [1]. Conjoined twins occur when division of the embryo takes place at around 15 days of fertilization. Dicephalus is a rare form of congenital malformation resulting from incomplete twinning. Fetus with dicephalus monosomus has two heads attached to one body. Though there are few reports of dicephalus twins, a combination of dicephalus twins with neural tube defect is rarely reported. The present case report describes a still born dicephalus baby with neural tube defect.

Case report

This female baby was born to a 25year old third gravida mother who has a normal healthy one year old baby. Her first pregnancy got terminated with a spontaneous abortion at three months of gestation. Second pregnancy was normal except that she had a hydatidiform cyst. She was treated with albendazole for one year. She conceived the last pregnancy spontaneously. She was booked and immunized. She was not exposed to any teratogenic drugs or radiation during antenatal period. She was taking iron and folic acid tablets as prescribed by the doctor. She perceived fetal movements first at 16 weeks of gestation. Her first ultrasound imaging was done during 29 weeks of pregnancy. It showed polyhydramnios with amniotic fluid index of more than eight centimeters and fetus in breech presentation. Fetus was diagnosed to have gross hydrocephalus with thinned out brain parenchyma. Third and fourth ventricles were normal. There was a 7 X 5 cm cyst in left cerebral hemisphere producing mass effect. Spina bifida and a 2.3 cm X 1.7 cm sacral meningocele were detected. She was admitted in labor at 29 weeks of gestation. PGE2 was instilled before delivery. A stillborn baby weighing 1.39 kg was delivered by assisted breech delivery. External examination showed two heads fused with each other at the adjoining sides (dicephalus). Heads were attached to the body by a single neck. Baby had four eyes, two nose, two mouths and three ears. Baby had two upper limbs (dibrachius) and two lower limbs (dipus) which were morphologically normal (Fig 1). Examination of the back showed single vertebral column with a meningocele at sacral region of the spine (Fig 2). The esophageal, choanal and anal patencies were normal. There was no other obvious anomaly.

On autopsy of the two headed female baby, two heads fused in the midline continuous with a single neck and trunk were observed. There was no webbing of the neck. At the site of fusion there was a projection of fused auricles. There were two upper limbs and two lower limbs and normal female genitalia.

There was transposition of great vessels of heart. Arch of aorta was giving rise to right brachiocephalic trunk, left common carotid and left subclavian arteries and continued as descending thoracic aorta. Right common carotid was arising from the right brachiocephalic trunk and dividing into two branches for the head and the cranial cavity. Similar finding was observed for left common carotid artery supplying the second part of the head. The left subclavian artery arising from the arch of aorta was normal without any deviation. There was a communication between the pulmonary trunk and descending thoracic aorta distal to the origin of left subclavian artery through ductus arteriosus.

Two brains were enclosed in a single calvarium. They were separated by a fold of a common duramater (fig 4). However, a well defined falx cerebri was identified between the cerebral hemispheres of each brain. The anterior cranial fossae were well defined with distinct margins of lesser wing of sphenoid forming the posterior boundary. The middle cranial fossae near the fused site were shallow and of reduced surface area compared to the unfused lateral aspect of the cranium. The posterior cranial fossa showed marked changes in the form of a protuberance of the petrous temporal bone and a common fused site lodging a part of a common hind brain and medulla oblongata continuing further with a single spinal cord.



Fig. 1: Still born dicephalus baby



Fig. 2: Meningomyelocele (arrow) at sacral region showing two heads and single body



Fig. 3: Infantogram with the arrow showing two mandibles



Fig. 4: Autopsy showing single calvarium with two brains separated by common duramater (arrow)

There was moderate hepatomegaly, spleen was lobulated, was subhepatic in position and appendix was splenic type. Uterus, fallopian tubes and ovaries were normal.

Discussion

Conjoined twinning is a very rare form of twinning in which the twins share many of the vital organs. Conjoined twinning occur one in 50,000 to 100,000 of all births [2]. There is high incidence of multiple congenital anomalies in conjoined twins. Because of multiple anomalies and sharing of vital organs, their life as a single individual is compromised. There is high incidence of mortality in conjoined twins. In a study conducted by Tippi et al., 28% of conjoined twins died during intrauterine period, 54% during immediate postnatal period and 18% survived [3].

Embryology of conjoined twinning is studied extensively. There are two opposing theories, fission and fusion theories. According to fission theory, the inner cell mass divides around day 13 to 15 of fertilization resulting in conjoined twins. Dicephalus twins are as a result of fission at the cranial end alone. According to the second theory, fusion theory, two separate embryos fuse with each other at areas where surface ectoderm is absent. This happens early in the embryonic life within hours to days [4, 7].

Dicephalus twins are very rare form of conjoined twins. They have two separate heads or heads fused with each other with one body. They encompass around 11% of conjoined twins and there is a female preponderance [5]. They are subdivided into many groups depending upon the number of upper limbs, lower limbs and number of torso [5]. Accordingly they are dicephalus tetrabrachius dipus, dicephalus tribrachius dipus and dicephalus dibrachius dipus. Most of the dicephalic twins are either still-born or succumb to neonatal death. As in any other form of conjoined twins they share vital organs and mortality is very high. In a study of natural history of conjoined twins, two cases of dicephalic twins with multiple anomalies dying immediately after birth are reported [3]. A case of 11 day survival has been reported by Grone et al. [6]. Abigail and Brittany are dicephalic twins of modern world. They were born as dicephalus tribrachius dipus. They have two separate heads and necks attached to one body. They share many of the internal organs. They had three arms of which one in the middle was rudimentary and was amputated early in life. They are now seventeen year old. They are developmentally normal and able to walk, run and swim. Each twin controls limbs on their side. They have different characters and personalities [4].

Early and accurate diagnosis is important in conjoined twins so that parents can be counseled for options of termination of pregnancy. Ultrasound imaging and antenatal magnetic resonance imaging are useful diagnostic tools in early pregnancy. Two-dimensional ultrasound can demonstrate conjoined twins as early as 12 weeks of gestation. Three-dimensional scan is superior to two-dimensional scan as it detects anomalies more precisely, and as early as nine weeks of gestation [8].

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