Introduction

The older term ‘Acardiac Monster’, was used earlier but now the preferred term is ‘Twin Reversal Arterial Perfusion’ (TRAP), with an incidence of one in 35,000 deliveries. The development of this results from arterio-arterial and veno-venous anastomosis, leading to predominance of one of the twins and has no direct vascular communication with the placenta. The acardiac twin is grossly abnormal and outcome is invariably fatal. The normal twin provides circulation for itself as well as the acardiac and the perinatal mortality is about 50-70%, as a result of congestive heart failure. We present a rare case of acardiac amorphous twin with available review of literature.

Case Report

A female aged 28 years, G2P1L1, was admitted to VIMS, combined group of hospital, Bellary, with history of 9 months amenorrhea, in labour. It was an undiagnosed twin pregnancy. The first twin was female of 2.5 kilograms and second of the twin was fleshy mass with attached umbilical cord. III stage of labour was uneventful. Radiological findings like x-ray and CT scan of the mass revealed no cardia/brain, with rudimentary spine and pelvic bones.

Autopsy

Gross morphology: A formless blob covered with skin, measured 17x16x12 cms in size and weighed around 650 grams. At cephalic end, there was a tuft of hair. A small segment of umbilical cord (6cms) was seen attached to ventral surface (Fig.1). A rudimentary limb bud was noticed in lower end (Fig.2) and spine was palpable. No recognizable facial or other part differentiation noticed.

Section/autopsy: A longitudinal incision from the upper to lower part on the ventral surface was taken. On dissection, no recognizable organs were noticed, but a tuft of vessels was seen at the upper part. The rudimentary spine and pelvic bones were made out. At cephalic end, below the tuft of hair, a cavity filled with 250 ml of straw colored fluid was noticed. Bits were given from the cephalic end, umbilical cord and tuft of vessels.

Microscopy: Section studied from the cephalic end did not show any glial tissue. Section studied from the umbilical cord showed one artery and one vein. Section studied from the tuft of vessels showed feature of an artery.

Abstract

Acardiac twining also referred as the Twin Reversal Arterial Perfusion (TRAP) sequence, is a rare congenital malformation found in monozygotic twin pregnancy. This case report is based on physical examination and autopsy findings. Grossly, a formless blob covered with skin, where a small portion of cord was attached to ventral surface. The cephalic pole, had tuft of hair and at lower end a rudimentary limb bud was seen. No recognizable facial or other part differentiation noticed. On autopsy, organs were absent and tuft of vessels noticed in upper part. The umbilical cord histological showed one artery and one vein, without any glial tissue in cephalic end. The acardius syndrome is a rare complication of monozygotic twin pregnancy. The twin reversal arterial perfusion theory is most accepted etiology of this disorder. A vascular communication exists between the twins and usually normal twin may develop cardiac failure as a result of perfusion of abnormal twin. In this case there was absence of development of cephalic pole, it being most distal to retrograde perfusion.

Key words: Congenital, Twin pregnancy, Acardaic, Amorphous, TRAP, Autopsy.
Discussion

Acardiac amorphous is a rare abnormality of twins. The term Twin Reversal Arterial Perfusion is the preferred term for the older Acardiac Monster, so named as reversed deoxygenated arterial supply is associated with only rudimentary development of upper body structures such as heart, face and arms. The acardius syndrome is a rare complication of monozygotic twin pregnancies. Failure or disrupted growth of head is called Acardius Acephalus, a partially developed head with identifiable limb is known as Acardius Myelacephalus and failure of any recognizable structures to form is Acardius Amorphous. A vascular communication exists between the twins and usually normal twin or so called pump twin may develop cardiac failure, as a result of perfusion of the abnormal twin. The acardiac twin is generally abnormal with reduction anomalies affecting all tissues. Perinatal diagnosis of acardiac twin was reported by Pezzati. Nerlich reported an acardiac twin which was associated with a twin showing amniotic band syndrome. Mohanty et al., reported a case of acardiac anomaly spectrum, indicating the cephalic pole is the most severely affected, being most distal to the retrograde perfusion and hence partial or total absence of development of head, face and brain, along with upper extremities. Chromosomal abnormalities have also been reported in the donor fetus like Klienfelters syndrome. Hence careful search for chromosomal malformation in the normal twin is essentially advised. The prognosis was directly related to the respective weights of the acardiac fetus and pump twin, with a higher weight of the recipient twin resulting in a higher likelihood of development of plohydrannios, cardiac insufficiency in the donor twin and premature delivery.

Conclusion

Acardiac amorphous twin is a rare congenital malformation found in monozygotic twin pregnancy. The reversed circulation in the anomalous twin may alter the hemodynamic forces needed for normal cardiac morphogenesis and results in arcadia, thus TRAP theory is an accepted etiology of this disorder. The cephalic end is severely affected which is being most distal to the retrograde perfusion, thus in acardia there is partial or total absence of development of head, face, brain and upper extremities, as is evidenced in this case.
References