Case Report

Acephalus acardia

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Abstract

Acephalus acardia is a unique and uncommon congenital malformation seen in Monochorionic twin pregnancies.

Twin-twin transfusion syndrome (TTTS), is a complication associated with twin pregnancy where, as a result of sharing of single placenta, the blood supply of mono-chorionic twins get connected. The donor twin or the "pump twin" provides circulation for itself and to the recipient acardia twin. The acardiac twin which receives deoxygenated blood is usually grossly abnormal with severe reduction anomalies of the upper part of the body as seen in our case. It can cause diagnostic and treatment difficulties. Currently it is possible to stop the blood flow to the acardiac twin by destroying the blood vessels, which supply the acardiac twin. We report a case of Monochorionic twin pregnancy having acephalic acardia with autopsy findings. She was noted for complete lack of antenatal care, admitted as an obstetric emergency and underwent cesearean section in a peripheral hospital. Identification of this condition early in pregnancy can possibly save one or both babies.

Case report:

Acephalus acardia is an uncommon congenital malformation occurring approximately 1 in 35,000 deliveries.(1) We report a case with autopsy findings.

Pregnancy in a 28 year G2P1 Indian woman presented in labor, at 34 weeks (menstrual age), to a private hospital. She was married for 7 years and was a non-consanguineous marriage. She had a 6-year-old healthy girl, born from a caesarean section. She had no infection, or drug intake during early pregnancy. The patient reported no unusual events or exposures in the current pregnancy. She was noted for complete lack of antenatal care. Clinically there was over distention of uterus with severe Polyhydramnios.

In view of previous cesarean section, over distended uterus and suspicion of threatened uterine rupture, a decision was taken to do emergency cesarean section. An acephalic male fetus, weighing 1500 gm was delivered after which an apparently normal, live female fetus weighing 1750 gm was delivered.

The abnormal fetus had no head. The left upper extremity was absent and there was also a severe reduction of the right upper extremity The lower trunk was normal. Both feet of the abnormal fetus showed only three toes.. The external genitalia were that of a male.

Surgery was uneventful and the patient was discharged in good health. The acephalic fetus was sent for autopsy.

Autopsy revealed no recognizable head, but a mound above sloping shoulders. Heart, stomach, liver, spleen, pancreas were absent. With proper development of lower half, the upper half was made of a single mass with a cyclope.

There was normal development of GI tract from ileum downwards. Retroperitoneal structures were normal. The supra-diaphragmatic portion

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Conclusion:

The tissues of anomalous fetus are kept alive by their essentially parasitic relation to the co-twin, whose heart perfuses both the bodies. As TTTS cannot be prevented, early diagnosis of this disorder in an identical twin pregnancy can possibly save one or both babies. Currently it is possible to eliminate the anastamosis between the twins and stop the blood flow to the acardiac twin. This can be achieved either by high energy Radio frequency Ablation (RFA) or Fetoscopic placental laser surgery.

References

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