

A Rare Case Report- Syncephalus Conjoint Twins

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ABSTRACT

Conjoint twins or Siamese twins are occurrence of characteristics forms because of joining of twins which may start at either of pole. Conjoint twins are very rare with an incidence of 1 in 100000 to 200000 births. **Syncephalus:** One head with a single face but four ears and two bodies. We report a case of 21 year old primigravida, who came to the OPD for her antenatal check-up. Her past history was insignificant. Her menstrual cycle was regular with 28-30 day cycle. On examination general examination was normal, while per abdominal examination, her abdomen was disproportionately enlarged for her gestational age by LMP and found to be of 34 weeks size, where multiple fetal parts palpable and fetal heart rate heard. Her previous ultrasound report was unremarkable and showing single fetus of 25 weeks gestation. Because clinical findings were not corresponding, repeat ultrasound advised. After two weeks patient got admitted with complaint of draining per vaginum for two days, the recent USG report have conjoint twin with single head. A syncephalus stillborn baby delivered by caesarean section. Baby had single head, single face, four ears and two bodies. Internal anatomy could not be confirmed as couple declined autopsy for religious region. Conjoined twins especially cephalopagus and syncephalus are quite a rare occurrence. Diagnosis can be missed because of single head, so needs careful assessment clinically and sonologically.

KEYWORDS: Conjoint twins, Syncephalus

INTRODUCTION

Conjoint twins may result from aberration in the twinning process, traditionally ascribed to incomplete splitting of an embryo into two separate twins¹. Conjoined twins are commonly referred as Siamese twins after Chang and Eng Bunker of Siam, who were thoracopagus. Incidence of conjoint twins is 1 in 100000 to 200000 births and always monozygotic. Female to male ratio is (3:1). Approximately 50% are stillborn, and around 35% dies within 24 hrs. It is thought to result from incomplete, delayed division of the inner cell mass which occurs after the 14th day from fertilization. As differentiation of the chorion (placenta) and amnion have already occurred, they are also monoamniotic and monochorionic. Thus identification of a dividing membrane or two placentas excludes the diagnosis. Classification is based on symmetry and the size of the fused anatomic region. In symmetric forms, the name of the fused region is followed by the term pagus e.g. thoracopagus, omphalopagus, pyopagus, craniopagus, ischiopagus, etc. Among these Parapagus is most common. The embryonic disc may be divided incompletely at either of pole or at both pole, resulting in conjoint twins of different types. Cephalopagus: single conjoined head with two faces on opposite sides, upper part of the body is fused while the bottom part are separate. Syncephalus: One head with a single face but four ears, and two bodies with or without joined thorax.

CASE REPORT AND RESULT

Mrs. Xyz, a 21 years old primigravida, presented to our opd with 7 months amenorrhoea for antenatal checkup. LMP was 18.06.14. Past menstrual history was 3-4/30, regular. Her 1st and 2nd trimester was uneventful with one previous ANC at 6 months, 2 doses of tetanus immunization and irregular iron and calcium tablets intake. It was a spontaneous conception within 1 year of marriage and no significant positive history of any teratogen exposure. There was no family history of multiple pregnancy or congenital malformation.

On examination, general examinations and vitals were within normal limit except mild pallor. Systemic examination -NAD. On per abdominal examination uterine height was corresponding to 34 weeks size, disproportionately enlarged for gestational age, multiple fetal parts with two fetal pole being felt, fundal grip broad soft & irregular part s/o buttock, lateral grip- both sides knob like structure s/o limb, pelvic grip- smooth hard & globular part s/o head felt. Fetal thud palpable. FHS was 148, regular, midway between umbilicus and symphysis pubis on midline. She had a previous ultrasound (done 2 weeks back) report showing single live intrauterine fetus, cephalic, CGA- 25wks 3 days, EFW 936 grams, placenta posterior, AFI 14. As clinical findings were not corresponding with gestational age and previous USG report, so repeat USG advised.

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She came after 2 weeks with c/o leaking pervaginum for last two days. On examination, there was mild pallor, no oedema, pulse 98/min, BP 116/80 mm of Hg, Systemic examination -NAD. & P/A finding was same as before with floating head, and fetal tachycardia- 180/min P/V examination-active draining present, OS – 1finger tight, membrane was absent, station was high up, pelvis average. A recent USG showed syncephalus monster twin with single head , two bodies, joined at thorax,

Investigation: Blood group :A+,Hb-10.2gm,TLC-11840, DLC-N80L17M2E1B0, FBS-80g/dl, Viral markers-negative.

USG- Monochorionic, monoamniotic, conjoint twins fused at head and thorax with common face, cephalic, fetus A- CGA - 31 wks, EFW-1.7 kg, fetus B- CGA- 28wks, EFW - 1.8 kg, Placenta common for both twin, posterior, grade 2, not low lying, AFI-10, As head was floating, not entering in pelvis on pushing, unfavourable cervix and prolonged rupture of membrane, decision of LSCS taken. Patient was taken for LSCS.

Baby was a syncephalus monster with 4 ears, 2 occiputs, and 1 face with 2 eyes, single nose, and single mouth. External genitalia was female, weight 3.4 kgs and it showed no signs of life. Lower parts of bodies were separate with a pair of lower limbs each; there were four arms and single umbilical cord with 5 vessels (Figure 1,2).



Figure 1,2: Picture of conjoint twins

DISCUSSION

Very few similar cases in literature were found. Gough² in 1934 described such a case with two complete lower bodies, one face, two occiputs and four ears (Syncephalus Thoracopagus, subgroup Disprospus Diophthalmus).

The diagnosis of conjoined twins can be made at mid trimester using sonography or 3-D USG or MRI. Counselling of parents to be done after a thorough examination of organs involved and survival chances of each of the twin baby.

The sonographic and radiologic findings have been described by Gray et al.³ and Koontz et al. and Gore et al. These include 1) lack of a separating membrane 2) non-separable skin contour with an inability to separate the fetal bodies 3) detection of other anomalies in a twin gestation 4) solitary umbilical cord with more than 3 vessels present 5) both fetal heads persistently at the same level 6) extension of the cervical spine (because most conjoined twins are fused ventrally) 7) bibechech or less commonly, bicephalic presentation and 8) constant relative fetal positions. It should be remembered, however, that discordant presentation does not exclude the diagnosis, particularly in omphalopagus twins. The fused area may be quite small allowing for rotation of one of the twins.

The prognosis for conjoined twins is quite poor. 75% of conjoined twins are stillborn or die within 24 hours. Of thoracopagus conjoined twins, 75% have extensively joined hearts that preclude successful separation. The unfortunate death of Laden and Laleh in an operation theatre in Singapore indicates the complications & risks of separating conjoined twins. There is a high incidence of congenital malformations like anencephaly in conjoined twins (10-20%) which are unrelated to the point of fusion. There is likewise a higher incidence of polyhydramnios (50-75%).

Management: The surgical separation is successful when organs essential for life are not shared. Consultation with the paediatric surgeon facilitates decision making. Twinning is a teratogenic event, so counselling for continuation or termination of pregnancy, is essential. To determine the feasibility of separation, one must carefully access how the twins share organ function. In the case of conjoint twins after 24 weeks gestational age, the choice between vaginal delivery or caesarean section should be made based on maternal safety and fetal condition. Vaginal delivery of conjoint twins is difficult, dystocia and traumatic injury to uterus and cervix is common. Caesarean is the method of choice for viable conjoint twins.³

COCLUSION

Early prenatal diagnosis and typing of conjoint twins allows better management of pregnancy including counselling of parents regarding continuation or termination of pregnancy, elective mode of delivery with

postnatal surgery. Conjoined twins especially cephalopagus and syncephalus are quite a rare occurrence. Diagnosis can be missed because of single head, so needs careful assessment clinically and sonologically. These twins generally cannot survive due to severe malformations of the brain. Fetal prognosis not good in these cases and also maternal risks increases.

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