Cephalothoracopagus Janiceps Conjoined Twins – A Rare Case

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Abstract

Conjoined twin is an unusual congenital anomaly having severe mortality and morbidity. We are reporting a rare case of conjoined cephalothoracopagus janiceps twins diagnosed by ultrasonography at 30 weeks when they presented to our department for the first time. Two live male conjoint babies were delivered by caesarean section. They died in half an hour after birth. Mother was discharged from hospital at 7th post natal day. This case is extremely rare because of following features: (a) presence of separate hearts (b) male sex (c) bony fusion at the level of the base of skull. Literature suggests that early diagnosis by a combination of ultrasound is essential for management. It gives an opportunity for viability and process of surgical separation. With this early counseling of parents and termination can be done if indicated.

Key words: Conjoined Twins, Cephalothoracopagus, Janiceps

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Introduction

Conjoined twins are a category of monozygotic twin, in which incomplete embryonic division occurs on the 13th-15th day following conception, resulting in different degrees of fusion defects between two fetuses[(1)].

Conjoined twins are about 1% of monochorionic twins, with an estimated incidence ranging from 1: 30,000 to 1: 200000 live birth[(2,3)] and one in 650-900 twin deliveries[(4)]. An increased incidence of 1: 14,000 to 1: 25,000 is described in various parts of Southeast Asia and Africa[(5)]. The exact incidence of this anomaly is unknown because most of them end in abortion or still births and hence leading to scarcity of diagnosis[(6)]. Some 40-60% of conjoined twins are reported to be still born, and approximately 35% of live births do not survive beyond the first 24 hours. Till date there are nearly 250 successfully separated cases in the history. There is 3:1 female predominance seen in these cases. Classification is made according to the site of fusion, the thorax (thoracopagus 30-40%) abdomen (omphalopagus; 25-30%) sacrum (pyopagus; 10-20%) pelvis (ischiopagus; 6- 20%) skull (craniopagus; 2-16%) face (cephalopagus) or back (rachipagus)[(3)]. Cephalothoracopagus janiceps is a very rare form of conjoined twins, which occurs in one of every million births and in one of every 58 sets of conjoined twins making it almost impossible for every obstetrician to see 1 case during her life time[(7)]. The term janiceps is derived from Janus, who is the two-faced Roman god[(8)].

Case Report

A 28 year old female with one live birth by caesarean section and one abortion of low socioeconomic status was admitted at 30+ week pregnancy with recently diagnosed conjoined twins at 28 weeks of pregnancy by USG. She was admitted for termination of pregnancy. She was not a regular ANC. Her first USG was done at 28th week of pregnancy which demonstrated cranio-thoraco-omphalopagus conjoined twin with variable presentation and polyhydrominos. Patient was then referred to our hospital and repeat scan was done which revealed conjoint twin of 32 weeks gestation with variable presentation with both fetal head thorax and abdomen joint together, fetal heads were at the same level in the same plane and amniotic fluid was increased. Her trimesteric history was uneventful. She had a previous caesarean 5 years ago in view of gestational hypertension and one abortion 6 years ago at 2months of gestational age which was spontaneous abortion. Her previous medical and surgical history was unremarkable. There was no family history of twining on both maternal and paternal side. On abdominal examination symphysiofundal height was 34 weeks size; fetal heart rates were 150 beats per min and 140 beats per min, uterus-relaxed.

During hospital stay elective caesarean section was done very next day and two alive male fetuses joined at the head, chest and abdomen with combined weight of 1.5 kg, were delivered. Fetus together had 2 eyes, one nose with single nostril, one mouth and one ear each. A single umbilical cord had two arteries and one vein with monochorionic monoamniotic placenta. The neonates died after few minutes of birth.

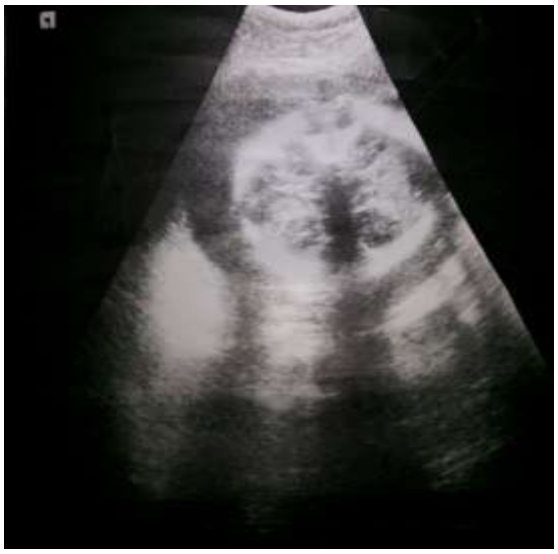

Fig. 1: USG showing fused cranium of twins

Fig. 2: Fused head face neck thorax and abdomen in janiceps twins

Fig. 3: Show single nostril and mouth

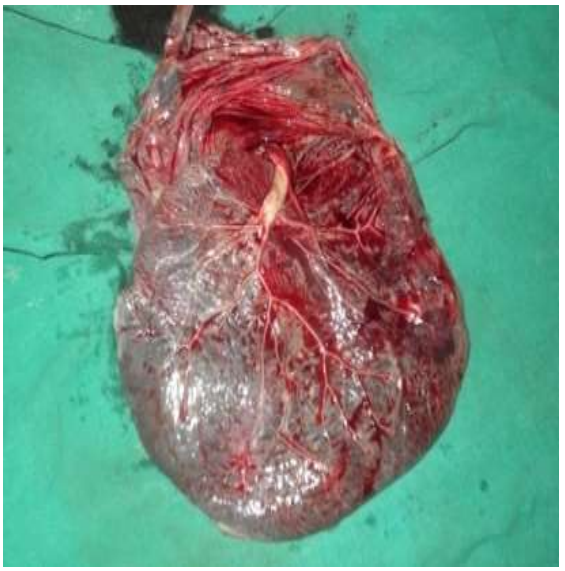

Fig. 4: Single placenta with single cord

Discussion

Conjoined twins are monochorionic monoamniotic twins where twinning is started after the embryonic disk formation and leads to its incomplete fusion[9]. Very few obstetricians get a chance to encounter janicep twins in their professional life[10].The diagnostic modalities for antenatal diagnosis of conjoined twins include X rays, ultrasound and magnetic resonance imaging. In cases of multiple pregnancies, an ultrasound should be performed at an early stage of gestation (10-14 weeks) to determine chorionicity and amniocity. This makes it possible to evaluate the gestational chorionicity precisely. Ultrasonologist should make sure that he Visualize two separate twins as conjoined twins are always monochorionic and monoamniotic.

Early diagnosis plays a vital role in the management of the pregnancy. With transvaginal

ultrasound, a conjoined twin can be diagnosed as early as 8 weeks gestational age[11]. Diagnostic criteria stated in literature for diagnosis of conjoined twins are: absent separating membrane, conjoined body parts, inseparable bodies or heads despite changes in fetal position, bifid appearance of the fetal pole in the first trimester, more than three vessels in the umbilical cord, complex structural anomalies, heads or bodies at the same level, hyper extended spine, unusual proximity of the extremities, persistence of the relative positions after movement or at the follow-up scan[12]. Polyhydramnios is commonly associated in 50–76% of cases. Recent reports have also shown the use of 3D ultrasonography in antenatal diagnosis of conjoined twins[13].

An early antenatal diagnosis of conjoined twins is practically seen only in very few cases. It is diagnosed mostly late in gestation or during parturition making the situation more worse[14]. If diagnosed antenatally it is mandatory to rule out the anatomical level of fusion between the two twins. A detailed survey of the vasculature of fused vital organs is very important in determining the prognosis of surgical separation and also owing to the high frequency of associated anomalies related to fusion, which include neural tube defect, orofacial cleft, imperforate anus and diaphragmatic hernia. In thoracophagus, twin fetuses usually have a common sternum, diaphragm, upper abdominal wall, liver, pericardium and gastrointestinal tract.

The prognosis for conjoined twins is mostly unfavorable, with approximately 40% of cases being stillborn[15]. The worst prognostic concern is seen with craniophagus twins and those having a single heart[16]. Structural anomalies are frequently found such as cardiac malformations, common omphaloceles[17], and neural tube defects[15].

The prognosis of treatment operation and its success mainly depend on the extent of cardiac fusion. Literature reports high success rates in cases with only pericardial fusion[18]. There are no reports of conjoined twins with ventricular conjunction having been successfully separated with both twins surviving. Magnetic resonance imaging and computed tomography both provide excellent anatomic and bone detail, demonstrating organ position, shared viscera, and limited vascular anatomy[19], but the best results was obtained by a combined evaluation of ultrasound and magnetic resonance imaging[20]. On diagnosis of nonviable conjoined twins, pregnancy needs to be terminated[15].

Early diagnosis in these cases can enable timed counseling of the family. It also makes the termination of pregnancy much easier. However, in our case due to lack of awareness and low resources, she was diagnosed late and outcome was bad.

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References

1. Sen C, Celik E, Vural A, Kepkep K. Antenatal diagnosis and prognosis of conjoined twins- a case report. J Perinat Med 2003;31:427-430.
2. Spitz L. Conjoined twins. Prenatal Diagnosis 2005;25:814-9.
3. Edomonds LD, Layde PM. Conjoined twins in the United States, 1970-1977. Teratology 1982;25:301-8.
4. Quiroz VH, Sepulveda WH, Mercado M, Bermudez R, Varela J. Prenatal ultrasound diagnosis of thoracophagus conjoined twins. J Perinatal Med 1989;17:297-303.
5. Diaz JH, Furman EB, Perioperative management of conjoined twins. Anesthesiology 1987;67:965-73.
6. Milham S: Symmetrical conjoined twins: An analysis of the birth recorded of twenty two sets. J Pediatric 1966;69-643.
7. Abossolo T, Dancoisne P, Tuaillon J, Orvain E, Sommer JC, Rivière JP. Early prenatal diagnosis of asymmetric cephalothoracopagus twins. J Gynecol Obstet Biol Reprod 1994;23:79–84
8. Chen CP, Shih JC, Shih SL, Huang JK, Huang JP, Lin YH, et al. Prenatal diagnosis of cephalothoracopagus janiceps disymmetros using three-dimensional power Doppler ultrasound and magnetic resonance imaging. Ultrasound Obstet Gynecol 2003;22:299–304.
9. George D, Athanasios P, Ioannis T, George M, Ioannis K, Aris A. First trimester diagnosis of dicephalus conjoined twins. Eur J Obstet Gynecol Reprod Biol 2004;112:110–3.
10. Grover V, Chawla R and Mishra SL: Management of conjoined twins Int J Gynecol Obstet 1990;31:67-69.
11. O'Neill JA, Holcomb GW III, Schnauffer L, Templeton JM Jr, Bishop HC, Ross AJ III, Duckett JW, Norwood WI, Ziegler MM, Koop CE. Surgical experience with thirteen conjoined twins. Ann Surg 1988;208:299-312.
12. Harper RG, Kenigsberg K, Sia CG, Horn D, Stern D, Bongiovi V. Xiphopagus conjoined twins: a 300-year review of the obstetric, morphopathologic, neonatal, and surgical parameters. Am J Obstet Gynecol 1980;137:617–29.
13. Kuroda K, Kamei Y, Kozuma S, Kikuchi A, Fujii T, Unno N, et al. Prenatal evaluation of cephalopagus conjoined twins by means of three-dimensional ultrasound at 13 weeks of pregnancy. Ultrasound Obstet Gynecol 2000;16:264–6.
14. Viljeon D and Beighton P. Obstetric implication of conjoined twining. J Obstet Gyenaecol 1984; 4: 233.
15. Puech F., Vaast P., Codaccioni X., Hubert D. - Grossesses gémellaires et multiples. Etude anatomo-clinique et prise en charge. Editions techniques- Encycl Med Chir. Obstetrique, 5-030-A-10,1993,16p.
16. Hill L.M. - The sonographic detection of early first-trimester conjoined twins. Prenat Diagn 1997;17:961-3.
17. Cazeneuve C., Nihoul-Fékéte C., Adafer M., Yassine B., Boury R. et al- Conjoined omphalophagous twins separated at fifteen days of age. Arch Pediatr 1995;2:452-5.
18. Sanders SP, Chin AJ, Parness IA, Benaceraff B, Greene MF, Epstein MF, Colan SD, Freigoletto FD. Prenatal diagnosis of congenital heart defects in thoracoabdominally conjoined twins. N Eng J Med 1985;313:374-80.

19. Spielmann AL, Freed KS, Spritzer CE. MRI of conjoined twins illustrating advances in fetal maging. J Comput Assistd Tomogr 2001;25:88–90.
20. Garel C, Brisse H, Sebag G, Elmaleh M, Oury JF, Hassan M. Magnetic resonance imaging of the fetus. Pediatr Radiol 1998;28:201–11.