ABSTRACT

Conjoined twins are rare variants of monozygotic twins, which result from an incomplete late division of the embryonic disc. Here, we report a rare case of conjoined twins of cephalothoracoomphalopagus variety where the line of fusion extended from the cranium up to the level of the umbilicus. The crown-rump length of each twin was 23 cm, which corresponded to the prenatal age of week 24 in a normal gravidity. The anatomical features of these conjoined twins were observed and external anomalies such as indefinite single nostril and male hypoplastic genitalia were described and documented. The old formalized specimen was studied to reveal anomalies by using current imaging techniques, thus keeping it intact for posterity. Computed tomography (CT) findings suggest cephalothoracopagus twinning, with the gestational age being 24 weeks (approximately). An attempt was made to correlate the findings on embryological basis.

Key words: Cephalothoracoomphalopagus, computed tomography (CT), conjoined twinning, cranium, magnetic resonance imaging (MRI), omphalo-cephalothoracopagus, umbilicus

INTRODUCTION

Conjoined twinning is one of the most fascinating embryological malformations seen in humans and has also been reported in other mammals, fishes, birds, reptiles, and amphibians. Treating conjoined twins can be a serious challenge for the operating surgeon. Furthermore, these cases often raise religious, moral, ethical, and legal issues. Approximately, 75% of conjoined twins are females and 70% of them are fused at the thorax (thoracopagus) or at the abdomen (omphalopagus). The union can be in the frontal, transverse, or sagittal plane. These can be classified into two main categories, namely, symmetrical or equal conjoined twins (two babies with an equal sharing of body parts) and asymmetrical or unequal conjoined twins (a small part of the body is duplicated or an incomplete twin is attached to a fully developed twin). In broad terms, conjoined twins may be regarded as a doubling anomaly. The later the incomplete embryologic separation occurs, the higher the likelihood of a complicated fusion. The incidence of conjoined twins is between 1 in 50,000 and 1 in 250,000.[1,2] Currently, conjoined twinning is commonly believed to be the result of partial fission of the primitive node and the primitive streak.[3] A hypothetical mechanism postulated by Spencer suggests that after separation, monovular twins fuse in one of the eight predictably homologous sites.[4] It was the intention of this report to investigate the internal anatomical structures of the
specimen without destroying it by physical dissection. By using noninvasive techniques such as magnetic resonance imaging (MRI) and computed tomography (CT), it is possible to reveal the internal structural anomalies of preserved conjoined twins without dissecting the specimen and to compare it with the previously reported cases. CT scan was done to look for bony abnormalities and MRI to comment on the internal structures. Though the bony fusions were clearly delineated, MRI was noncontributory; it may have been due to internal putrefaction of the organs as it was an old formalized specimen.

**CASE REPORT**

The specimen under study was procured from the Museum of Anatomy Department (Department of Anatomy, Government Medical College, Patiala, Punjab). This was preserved and stored in formaldehyde for more than three decades. A preliminary external examination of the specimen led to the conclusion that this was a case of symmetrical conjoined twinning of the cephalothoracoomphalopagus variety, having an unusually large head and thorax. Both fetuses were fused ventrally in a craniocaudal manner. The line of fusion started from the vertex up to the level of the umbilicus, along with fusion of the umbilical cords. The cut section of the fused umbilical cord showed five vascular openings. A completely formed face was seen on one side comprising one oral aperture, one nose with two nostrils, two orbits, and two external ears [Figure 1]. Patency of the oral aperture tested with an infant feeding tube was found to be positive, which suggested that the buccopharyngeal membrane was obliterated. A highly underdeveloped face was seen on the other side with two fused auricles at the level of the neck; an indefinite opening was seen that may have been of the nostrils leading to two lateral foramina that indicated oblique fissure [Figure 2]. Both the fetuses were males with hypoplastic genitalia.

Radiological examinations were most contributory to the specimen evaluation though MRI study was inconclusive due to putrefaction of the internal organs.

1. One completely formed facial skeleton was seen comprising two frontal bones, two orbits, nasal bones, maxilla, and the mandible [Figure 3]. Another incomplete facial skeleton was seen opposite the aforementioned face in the fused skulls, comprising an underdeveloped frontonasal process approaching the mandibular processes. But there was no demarcation between the maxillary and mandibular processes as they merged forming one process [Figure 4].

2. Fused skull vault comprised four parietal bones and two occipital bones.

3. Sternums of both fetuses were not visualized; however, the ribs (11 pairs), clavicle, and scapula were normally seen in each other.
4. Spine showed no evidence of dysraphism.
5. Pelvic bones were normal.
6. Upper and lower limb bones were normal.

**DISCUSSION**

Although fetal autopsy remains the gold standard for the confirmation of fetal pathology, the use of noninvasive MRI is frequently advocated when there is fear of disfigurement or for religious beliefs. Though many authors have reported different types of conjoined twins, there is a paucity of literature reporting any variety of this nature. Khanna et al. described a classical cephalothoracopagus janiceps disymmetros as described by Hovorakova et al. fetus with frontally fused cerebra, separate spines, identical faces, and four limbs each. Anastasakis et al. described a least common case of dibrachius tripus (three legged) parapagus (ventral orientation of lower limb) twinning. Salmaso et al. reported a rare case of gnatho-thoracopagus where the conjunction begins more caudally and the conjoined twins have almost fully separate heads. The classification scheme utilized in this study is from Spencer’s categorization of conjoined fetuses based upon the investigation of over 1,200 specimens. The specimen of the present study can be categorized as cephalothoracopagus (or cephalothoracoomphalopagus) in which the twins are united ventrally, sharing structures as distal as the abdomen. Typically, they share a face and the spines are ventrally oriented toward each other. It has been postulated that these conjoined twins result from the union of cardiac primordia, septa transversa, and oropharyngeal membranes. The precise etiology of conjoined twinning is unknown. The most common explanation is fission of a single zygote or alternatively, fusion of two dizygotic or monozygotic embryos in their very early embryonic development between 13 days and 15 days after conception. Because conjoined twins develop after differentiation of the chorion and the amnion, all conjoined twins are monochorionic-monoamniotic. Using a variety of radiological modalities, we demonstrated the possibility of studying very old damaged human material for documentation and scientific discussion without its destruction.

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**Conflicts of interest**
There are no conflicts of interest.

**REFERENCES**

1. Spitz L, Kiely EM. Experience in the management of conjoined twins. Br J Surg 2002;89:1188-92.
2. McHugh K, Kiely EM, Spitz L. Imaging of conjoined twins. Pediatr Radiol 2006;36:899-910; quiz 1002-3.
3. Sadler TW. Langman Medical Embryology. 9th ed. Baltimore: Lippincott Williams & Wilkins; 2004. p. 143.
4. Spencer R. Conjoined Twins: Developmental Malformations and Clinical Implications. Baltimore: Johns Hopkins University Press; 2003. p. 1-476.
5. Huisman TA, Wisser J, Stallmach T, Krestin GP, Huch R, Kubik-Huch RA. MR autopsy in fetuses. Fetal Diagn Ther 2002;17:58-64.
6. Khanna PC, Pungavkar SA, Patkar DP. Ultrafast resonance imaging of cephalothoracopagus janiceps disymmetros. J Postgrad Med 2005;51:228-9.
7. Hovorakova M, Peterkova R, Likovsky Z, Peterka M. A case of conjoined twin’s cephalothoracopagus janiceps disymmetros. Reprod Toxicol 2008;26:178-82.
8. Anastasakis E, Zhang EG, Bates AW, Abdel-Aal MA, Kadir RA. Parapagus diencephalus dibrachius tripus. An unusual case of conjoined twins. Prenat Diagn 2007;27:1165-6.
9. Salmaso R, Manara R, Fassan M, Severino MS, Visentin S, Macchi V, et al. Radiological- pathological comparison in a case of conjoined gnatho-thoracopagus twins. Fetal Diagn Ther 2009;26:223-6.
10. Lee AW, Farnquist B, Islam O, Mackenzie J, Taylor SA, Pang SC, et al. Noninvasive investigation of asymetrically conjoined twins with features of rachipagus, parapagus diencephalus and cephalopagus. Clin Anat 2012;25:1023-9.