

Thoraco- Omphalo-Ischiopagus Tripus Conjoined Twins: Report of a Case

Naser Kamalian¹, Shapour Shirani^{2*} and Maryam Soleymanzadeh³

¹Associate professor of Pathology, Department of Pathology, Shariati Hospital, University of Medical Sciences, North Kargar Street, Shariati hospital, Tehran, Iran

²Assistant professor of Radiology, Department of Radiology, Tehran Heart Center, Tehran University of Medical Sciences, North Kargar Street, Tehran Heart Center, Tehran, Iran

³Research Assistant, Research Department, Tehran Heart Center, Tehran University of Medical Sciences, North Kargar Street, Tehran Heart Center, Tehran, Iran

Abstract

The frequency of thoraco- omphalo-ischiopagus tripus or conjoined twins (CT) is quite rare of mozygotic twinnings, and they are classified as unions of symmetrical CT. A case of conjoined twins with thoraco- omphalo-ischiopagus type is presented here as well as a review of differential diagnosis. The twins had two heads, two pairs of upper limbs and three lower limbs. They also shared thorax, abdomen and pelvis with a single beating heart.

Keywords: Conjoined twins; Monozygotic twins; Ambiguous genitalia; Surgical separation

Introduction

The earliest example of conjoined twins returns to a statue which belongs to sixth century, and now is being kept in a museum in Turkey [1]. The first successful attempt to separate conjoined twins occurred in 1689 by fatio [2]. The frequency of thoraco- omphalo-ischiopagus tripus or conjoined twins (CT) is quite rare entity with a estimation of 1 in 50,000 to 100,000 live births [3]. They develop from a single ovum and when the embryonic disc incompletely divides more than 13 days after fertilization [4], and have a strong relation with monoamniotic monochorionic type of placenta. Also there have been some reported attempts to induce conjoined twins experimentally following exposure to a variety of agents, but the ultimate reasons for this phenomenon are not clearly defined. Some studies suggest risk factors such as positive history of genetic talent for conjoined twins in each parent's family, history of drugs for treatment of infertility, or exposure to harmful radiation at the early time of pregnancy are also responsible for the matter.

Case Report

A 23-year-old woman, gravida 1, para 0 with 14 weeks of amenorrhea visited her radiologist for a routine checkup. The patient had a history of infertility and irregular menstrual periods and failed IUF before her natural pregnancy. She also did not have a history of drug abuse and exposure to environmental teratogen agents. Her husband had nothing positive in his history except a varicocelelectomy for correction of abnormal spermatogenesis process. No history of familial congenital abnormalities or twinning was found in the couple. The patient was examined by real-time ultrasonography by her obstetrician and the conjoined twin was already diagnosed for her. She was referred to our hospital and the first diagnosis was confirmed for her. The clinicians advised the patient about likelihood of future outcome, and she accepted termination of pregnancy.

After induction of delivery, the twins were examined. The placenta and its membranes were single. The attachment kind of was thoraco-omphalo-ischiopagus. The twins had two heads, two pairs of upper limb and three lower limbs (decephalus, tetrabrachius ,tripus)they also had common thorax, abdomen and pelvic. The external genitalia appeared ambiguous. Further evaluation by MRI demonstrated a single heart plus hydrocephalus in one head. Other internal organs were too small for autopsy and imaging evaluation (Figure1 and Figure 2).

Discussion

As mentioned earlier, the frequency of conjoined twins is estimated of 1 in 50,000 to 100,000 live births , and is believed to be related to a late cleavage of embryonic disk (day 13 and over) after fertilization. Most of conjoined twins report worldwide are females (up to 70-95%) [3]. Based on a large study in the United States, the incidence of conjoined twins was reported as below: the frequency was estimated 10.25 per million births. The most common types identified were as follows: thoraco omphalopagus (28.4%), thoracopagus (18.5%), omphalopagus (9.9%), parasitic twins (9.9%) and craniopagus (6.2%). The results of the study also suggested that only 60% of conjoined twins are born alive [3-4]. Furthermore, conjoined twins are often born with higher rates of abnormalities than other babies, thus survival likelihood is lower as the result [5]. Some environmental and genetic factors, such as history of twin and abnormality delivery, exposure to radiation, drugs for



Figure 1: Gross appearance.

***Corresponding author:** Shapour Shirani, Assistant professor of Radiology, Department of Radiology, Tehran Heart Center, Tehran University of Medical Sciences, North Kargar Street, Tehran Heart Center, Tehran, Iran, Fax: 0098-21 88029731; E-mail: sh_shirani@yahoo.com

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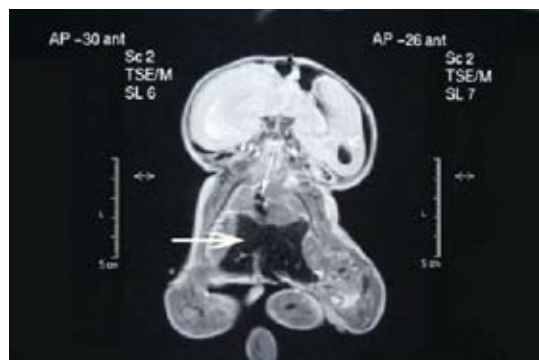


Figure 2: Sagittal T1 MRI of conjoined twins, short arrow dilated occipital horn of hydrocephalic fetus, thin long arrow common heart, thick long arrow common liver.

ovulation induction have been possible causes of conjoined twinning [6]. Prenatal diagnosis of conjoined twins is usually performed by ultrasonography, and is confirmed by a more definite imaging method such as MRI. Some important hints that are highly suggestive for conjoined twins are: lack of a separating membrane, detection of other anomalies, multiple (>3) vessels in umbilical cords and fetal bodies are never seen separately on regular sonography visits[7]. Ultrasonography can diagnose CT from the beginning of 12th week of pregnancy, and transvaginal sonography is also capable to detect it even in earlier times[8-9]. Utilization of MRI is also another unique imaging method for confirming ultrasonography findings. MRI technique is highly selective for evaluation the soft tissue defects, such as bladder extrophy, a common findings in CT [10-11]. Early diagnosis of conjoined twins, however is a great help in management of pregnancy and determining delivery technique. Even after live birth, imaging methods play an important role to detect fatal defects, especially in inner organs. Possible separation of conjoined twins should be beard in mind if there

are no life-threatening malformations [12]. Such operations usually have good satisfactory outcome unless for shared cardiac chamber or single extrahepatic billiary tree.

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