doi: 10.48095/cccg202544

Prenatal diagnosis of parasitic conjoined twins

Prenatálna diagnostika parazitických zrastených dvojčiat

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Summary: Heteropagus or parasitic conjoined twins represent an extremely rare anomaly, occurring in approximately 1 in 1 million cases. This condition is characterized by the presence of a parasitic twin with significant congenital abnormalities attached to an otherwise typically healthy fetus. The well-developed twin is known as the "autosite" or "host," while the severely affected fetus is termed the "parasite." Survival of the defective twin depends on the cardiovascular system of the second, relatively normal fetus. We present the case of a 27-year-old primigravida in her 14th week of pregnancy with ultrasound findings indicating parasitic conjoined twins, specifically omphalopagus.

Key words: conjoined twins – parasitic twins – heteropagus – prenatal diagnosis

Súhrn: Heteropágy alebo parazitické zrastené dvojčatá predstavujú extrémne zriedkavú anomáliu, ktorá sa vyskytuje približne v 1 z 1 milióna prípadov. Sú charakterizované prítomnosťou parazitického dvojčaťa s významnou vrodenou abnormalitou, pripojeného k zvyčajne inak zdravému plodu. Dobre vyvinuté dvojča je známe ako „autosite" alebo „hostiteľ", zatiaľ čo ťažko postihnutý plod sa nazýva „parazit". Prežitie defektného dvojčaťa závisí od kardiovaskulárneho systému druhého, relatívne normálneho plodu. Predstavujeme prípad 27-ročnej primigravidy v 14. týždni gravidity s ultrazvukovým nálezom poukazujúcim na parazitické zrastené dvojčatá, omfalopágy.

Kľúčové slová: zrastené dvojčatá – parazitické dvojčatá – heteropágy – prenatálna diagnostika

Introduction

Heteropagus or parasitic conjoined twins are a highly uncommon anomaly with an estimated incidence of less than 0.1 in 100,000 births. Among all instances of conjoined twins, the incidence can range from 4.5% to 15% [1]. The earliest reliable report of this phenomenon likely dates back to the 16th century by the French surgeon Ambroise Paré, who described an acephalous body attached to the abdomen in one of his patients [2].

This anomaly is characterized by a parasitic twin with significant congenital abnormalities joined to an otherwise relatively normal fetus. Heteropagus twins are connected at one of the shared points, similar to intact conjoined twins. They often have externally attached supernumerary limbs, sometimes with some viscera, and rarely a functional brain or heart. The well-developed twin is referred to as the "autosite" or "host," and the critically defective fetus is referred to as the "parasite." Tissues of the severely defective twin depend on the cardiovascular system of the second, relatively normal fetus for survival [2–4].

Parasitic conjoined twins arise when monozygotic twins do not separate completely, resulting in the dominance of one embryo's development over the other [5]. The asymmetry between the autosite and parasite likely occurs due to vascular compromise, causing the parasite's tissue to rely on collaterals from the autosite. This leads to selective ischemic atrophy in the deprived portion of the parasite's body [6]. Sonographic examination plays a crucial role in prenatal diagnosis [7].

Case report

A 27-year-old primigravida was referred to our department for an ultrasound examination during her 14th week of pregnancy. Her family and personal medical history were unremarkable, and the pregnancy occurred after spontaneous

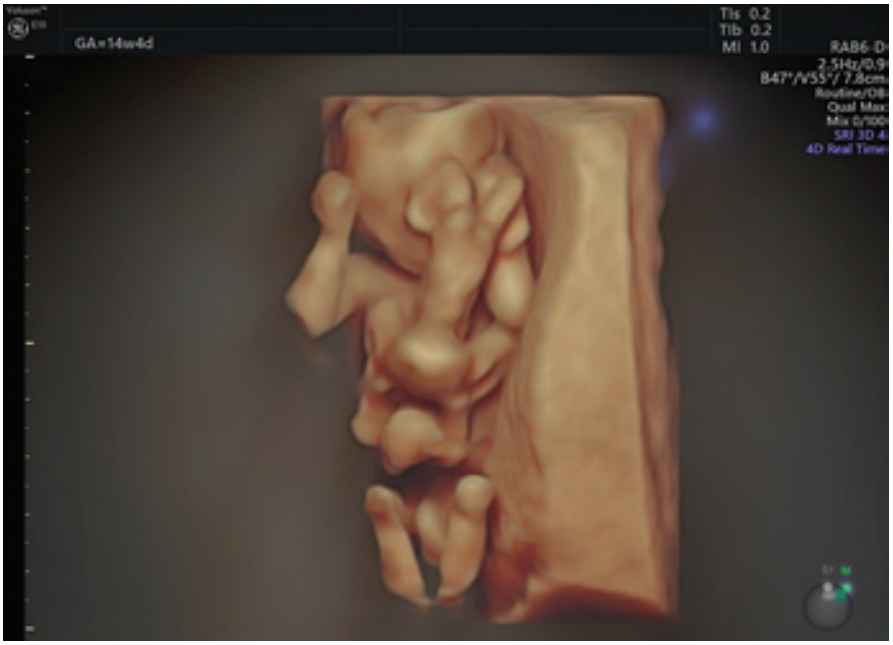

Fig. 1. High-definition three-dimensional ultrasound image at 14th weeks of gestation showing parasitic conjoined twins (omphalopagus), with the extrafetal body (parasite) attached above the umbilical cord insertion, displaying an omphalocele of the autosite.
Obr. 1. Trojrozmerný ultrazvukový obraz s vysokým rozlíšením v 14. týždni tehotenstva znázorňuje parazitické zrastené dvojčatá (omfalopágy) s extrafetálnou časťou (parazitom) pripojenou nad inzerciou pupočníka, zobrazuje omfalokélu autosite.

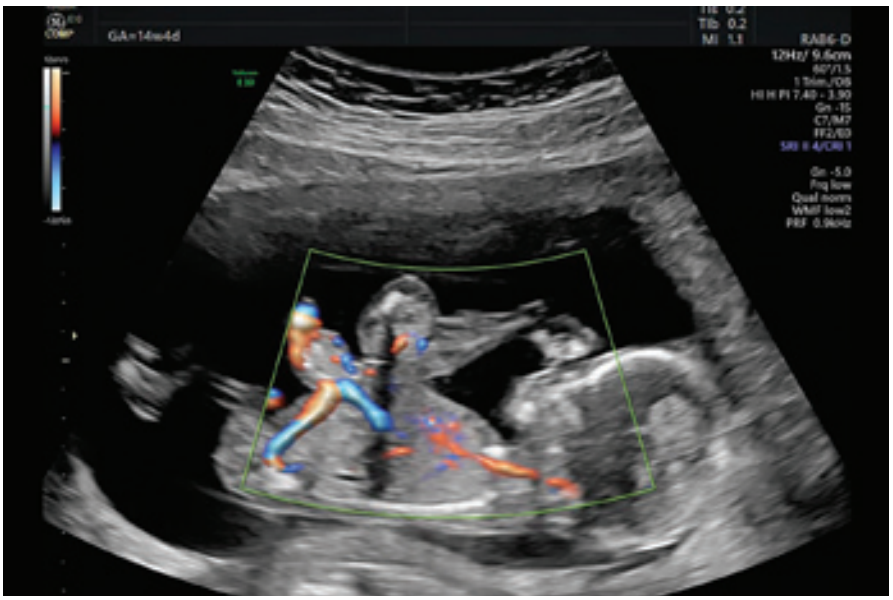

Fig. 2. Mode with color flow mapping ultrasound image at 14th weeks of gestation showing a parasitic fetus connected at the midline to the anterior abdominal wall of the autosite through soft tissue with weak vascularization. The soft tissue of the umbilical cord contained a total of five vessels – three in the umbilical cord of the autosite and two in the umbilical cord of the parasite.
Obr. 2. Ultrazvukový obraz s farebným mapovaním toku v 14. týždni tehotenstva zobrazuje parazitický plod spojený v stredovej línii s prednou brušnou stenou hostiteľa mäkkým tkanivom so slabou vaskularizáciou. Mäkké tkanivo pupočníka obsahovalo celkovo päť ciev – tri v pupočníku hostiteľa a dve v pupočníku parazita.

conception. She had not taken any medications or been exposed to teratogens, and all serological tests, including those for toxoplasmosis, rubella, cytomegalovirus, herpes simplex, HIV, hepatitis B surface antigen, and *Treponema pallidum hemagglutination*, had negative results.

During the ultrasound examination, conducted with a Voluson E10 system (GE Medical Systems Milwaukee, WI) using a convex 4–8 MHz abdominal transducer (RAB 4–8 L), bifetal gravidity was identified, complicated by parasitic conjoined twins. The crown-rump length of the first fetus was 86 mm, corresponding to 14 weeks and 4 days of pregnancy. A dominant omphalocele measuring 10 × 9 mm was present. The second twin, a parasitic fetus, was an acardiac and acranius conjoined above the navel in the epigastrium and distal part of the chest with the first fetus in the umbilical area, resulting in an omphalopagus conjoined twin (Fig. 1). This connection occurred through soft tissue with diminished vascular support. The fetuses had a total of five vessels in the soft umbilical cord tissue – three in the umbilical cord of the autosite and two in the parasite (Fig. 2).

The patient was informed about the unfavorable prognosis and requested a termination of the pregnancy. Termination was performed through ultrasound-guided dilation and evacuation, using a combination of forceps and aspiration under general anesthesia. Aborted tissues were sent in for histological examination, confirming the presence of five vessels in the umbilical cord samples, with no other significant findings.

Discussion

Conjoined twins are characterized by the region of their conjoined bodies, resulting from incomplete monozygotic twinning. Their development reveals a duplication anomaly believed to stem from the conception of a single zygote, likely

arising from the splitting and incomplete separation of the inner cell mass within 3–15 days after fertilization. This anomaly might also arise from the fusion of two distinct embryonic discs [8].

Conjoined twins can be symmetrical (diplopagus) or asymmetrical. Symmetrical conjoined twins are approximately equal in size and joined symmetrically, while asymmetrical conjoined twins involve one nearly normal twin (autosite) and an incomplete twin (parasite), fully dependent on the autosite for growth [9]. Conjoined twins are classified based on the most prominent site of union, such as cephalopagus (head), thoracopagus (thorax), omphalopagus (abdomen), ischiopagus (pelvis), parapagus (side), craniopagus (cranium), pygopagus (sacrum), or rachipagus (spine, back) [10].

Death of the parasitic twin during embryonic development results in the autosite retaining vascularized body parts in the heteropagus twin. The exact cause of embryonic death is unknown, but may involve ischemic atrophy, insufficient cardiac function of the parasite, or vascular steal from the autosite. The parasitic twin typically lacks a heart, receiving blood supply from vessels originating in various regions of the autosite, such as the liver, epigastric artery, umbilical vessels, or falciform ligament [11].

Fetiform masses may also be located within the body of the autosite or protrude from the mouth, known as fetus in fetu (endoparasitic twin). The heart and brain are typically vestigial or entirely absent, similar to cases of external parasitic twins [4].

Autosite twins often have congenital cardiac malformations, potentially caused by hemodynamic changes associated with supporting the parasitic twin [11]. Omphalocele is also common in the autosite [12]. Prenatal ultrasound is crucial for diagnosing conjoined twins, with possible confirmation as early as the 12th week. By the 20th week, prenatal ultrasonography can provide detailed anatomical information [13].

Postnatal diagnosis is complemented by ultrasound, computed tomography, and magnetic resonance imaging to offer a comprehensive view of blood circulation and type of connection, contributing to enhanced surgical success rates and reduced postoperative complications. Cesarean delivery is recommended to prevent complications during childbirth and reduce the risk of birth injury due to frequent occurrence of a large parasite with an anomalous shape [14].

Surgical separation poses significant challenges and requires a multidisciplinary team. Optimal timing for surgical separation varies, with elective procedures recommended for infants between 9 and 12 months old. Early separation might be required in emergent conditions such as congestive cardiac failure, respiratory distress, intestinal obstruction, gastroschisis, and obstructive uropathy [13]. Preferred treatment is complete excision, although surgical challenges depend on the location [7]. Survival rates are generally favorable in heteropagus twins due to less extensive vascular and visceral connections between the autosite and parasite compared to symmetric conjoined twins [2].

Conclusion

Parasitic conjoined twins represent an uncommon anomaly of monochorionic, monoamniotic twins. Prenatal ultrasound diagnosis is crucial for identifying this anomaly, which can be established as early as the 1st trimester. Three-dimensional ultrasound provides deeper insights into parasitic twins, offering a clearer view and improved understanding of how their fetal parts are arranged spatially.

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Publication ethics: The Editorial Board declares that the manuscript met the ICMJE uniform requirements for biomedical papers.

Publikačné etika: Redakčná rada potvrdzuje, že rukopis práce splnil ICMJE kritériá pre publikácie zasielané do biomedicínskych časopisov.

Conflict of interests: The authors declare they have no potential conflicts of interest concerning the drugs, products or services used in the study.

Konflikt záujmov: Autori deklarujú, že v súvislosti s predmetom štúdie/práce nemajú žiadny konflikt záujmov.

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Submitted/Doručené: 10. 9. 2024
Accepted/Prijaté: 10. 10. 2024

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