ORIGINAL ARTICLE

Asymmetric parasitic twins - Heteropagus

Rita de Cássia S. Alves[1]
Adriana Ubirajara Silva Petry[1]
Josenel Maria Barcelos Marçal[1]
Bruno Hochhegger[1]
Andréa Souto Damin[2]
Adriana Vial Roehe[1]

1. Docente, Departamento de Métodos Diagnósticos, Universidade Federal de Ciências da Saúde de Porto Alegre (UFCSPA), Porto Alegre, RS, Brasil.
2. Docente, Departamento de Ginecologia e Obstetrícia, Universidade Federal do Rio Grande do Sul (UFRGS), Porto Alegre, RS, Brasil.

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SUMMARY

BACKGROUND: *Asymmetric or heteropagus conjoined twins is a rare occurrence, with an incidence of one case in 1-2 million. Conjoined twins are classified according to their symmetry, place of fusion, and grade of duplication.*

METHODS: *We report here an extremely rare presentation of parasitic twins not described before. We describe macro and micro anatomic alterations and discuss the aspects of this peculiar presentation and the importance of prenatal diagnosis.*

RESULTS: *The case of a twenty-three-year-old patient, with monochorionic, monoamniotic asymmetrically-conjoined twin pregnancy, discovered at 29 weeks of gestational age. We believe that this report calls attention to this presentation and the importance of prenatal care and management. The twins were delivered vaginally without life. The twins' combined weight was 1.300 gr. They were bonded in the left cervical region.*

CONCLUSION: *This report may help to find strategies for clinical decisions in future cases. Antepartum diagnosis is important to the management, preoperative planning, and outcomes. Prenatal imaging exams like echocardiography, CT, MRI, and ultrasonography are feasible and can provide relevant information about malformation severity and prognosis.*

KEYWORDS: *Twins, Conjoined. Congenital abnormalities. Twins, Monozygotic.*

INTRODUCTION

Conjoined twins are classified according to their symmetry, place of fusion, and grade of duplication, thoracopagus being the most frequent type (40%), followed by *thoracoomphalopagus* (28%). Asymmetrically conjoined twins are also known as parasitic twins or *heteropagus*, and usually, they do not fit well in any classification[1].

Asymmetric or *heteropagus* conjoined twins are defined by Spencer as one severely damaged twin attached to a relatively normal twin through an asymmetric anatomical location[2]. One twin of the pair (autosite) is mostly intact, even though some congenital abnormalities can be observed. Its counterpart, the one called the parasite is badly defective and depends on the cardiovascular system of the autosite for survival[3]. It is an extremely rare occurrence, with an incidence of one case in 1 - 2 million births[4,5].

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CORRESPONDING AUTHOR: Adriana Vial Roehe
Rua Sarmento Leite, 245, Porto Alegre, RS, Brasil – 90050-170
Tel: +55 51 3033-8723 / Fax: +55 51 3303-8810
E-mail: aroehe@gmail.com; adrianar@ufcspa.edu.br

METHODS

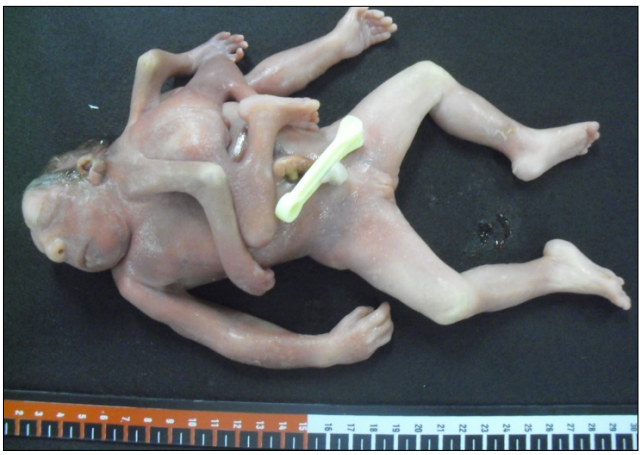
A twenty-three-year-old patient, P2L1, with no contributive medical history, went to the emergency service of the Femina Hospital, Porto Alegre, Brasil, with abdominal pain, in labor. The patient did not know about her pregnancy, but a monochorionic, monoamniotic conjoined twin pregnancy was discovered (29 weeks of gestational age) in a emergency ultrasonography examination. Intrauterine death was diagnosed and the fetuses were delivered vaginally without dystocia and induction; they proved to be conjoined twins with no cardiac activity.

RESULTS

The twins' combined weight was 1.300 gr. The autosite fetus crown-heel length was 27.5 cm crown-rump length was15.5 cm and head circumference was 15.6 cm. The gestational age was estimated at 29 weeks.

An ectoscopic examination demonstrated the parasite twin was bonded to the left cervical region of the autosite (Fig. 1).

FIGURE 1. MACROSCOPIC FRONTAL VIEW OF THE PARASITIC TWINS.



There were relatively well-formed upper and lower limbs. The parasite also had a rudimentary pelvis, with external female genitals and anus. CT scan features can be observed in Fig. 2 and 3.

Post mortem analysis of the parasite fetus showed pelvic organs inside the autosite fetus chest cavity. There was a rudimentary bladder and one kidney. These tissues were taken for histopathological examination. There was also a single intestinal loop from the parasite fetus fused with the intestinal loops of the autosite. No thoracic structures were observed in the parasite fetus.

FIGURE 2. (A) CORONAL CT SCAN AND (B) MINIMUM INTENSITY PROJECTION CORONAL CT SCAN OF THE PARASITIC FETUS REVEALS A HETEROGENEOUS MASS (ARROW) IN THE CERVICAL REGION WITH BONY COMPONENTS. THE BONY COMPONENTS (ARROW) RESEMBLE A VERTEBRAL BODY IN CROSS-SECTION. ALSO, CT DEMONSTRATED ANENCEPHALY.

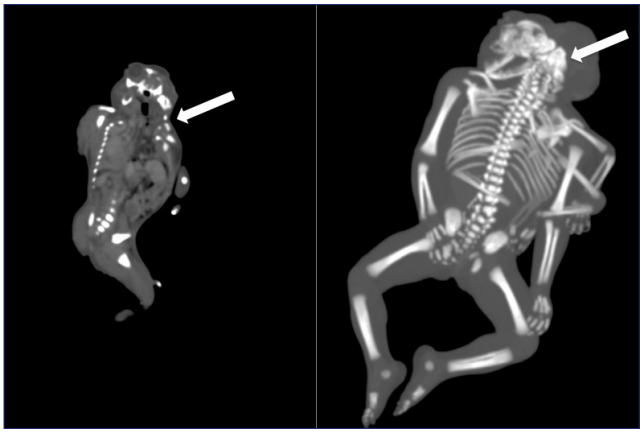
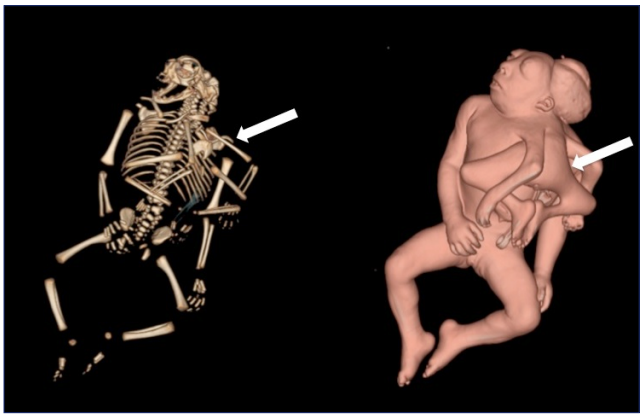


FIGURE 3. (A) VOLUME RENDERING BONE PROJECTION CT SCAN AND (B) VOLUME RENDERING SKIN PROJECTION CT SCAN OF THE PARASITIC FETUS DEMONSTRATED A HETEROGENEOUS MASS AT THE LEVEL OF THE AXILLA (ARROW) THAT CONSISTS OF AN OSSEOUS COMPONENT RESEMBLING THE PELVIS AND LUMBOSACRAL SPINE.



The autosite fetus was anencephalic. There were no facial or cervical malformations. The thoracic organs were topic. The lungs were hypoplastic and parts of the intestinal loops were located in the chest cavity (Fig. 6 of the Supplementary File). The other abdominal organs were topic and no abnormality was observed.

DISCUSSION

Conjoined twins are classified according to their symmetry, place of fusion, and grade of duplication. Different from the usual conjoined twins that have the same sites fused, parasitic twins are a subset in which "asymmetric" joining occurs, having their

own variants and independent classification. In this case report, the parasite twin was attached to the left lateral thoracervical region of the autosite, in an extremely rare presentation, not classified and not described previously[3].

According to Sharma et al.[3], there can be a single heart, single liver, small bowel loops crisscrossed from one fetus to the other. Other authors describe some cases of urinary tract communication between the parasite and autosite fetus[3]. Some cases of urinary tract communication between the parasite and autosite fetus have also been described[3]. In our case, the parasite fetus has a rudimentary urinary system without communication with the autosite urinary system. The parasite fetus had a single intestinal loop connected to autosite intestinal loops. This feature is described in 28% of parasitic fetus[3].

Anencephaly is described more frequently in *parapagus* and *dicephalus* twins, and most *heteropagus* parasite twins are anencephalic[1,3]. The autosite fetus reported here was also anencephalic. There was an amorphous mass of fibroangiomatous tissue in the occipital and cervical region between the autosite and parasitic fetus, probably corresponding to an anencephaly of the parasitic fetus as well. According to the CT scan, there was cervical spinal duplication. Except for anencephaly, there was no other significant malformation in the autosite fetus. In many cases of *heteropagus* twins, the prognosis is determined by cardiac abnormalities. In this case, the autosite fetus did not have any cardiac malformation.

As in our case, none of the reviewed cases presented a history of consanguinity and most of them did not have significant pregnancy complications. The etiology of conjoined twins is still unknown and the pathogenesis is obscure. Some studies refer to a discreet prevalence of female twins, while gender differences are less pronounced in *heteropagus* twins[2,3].

In this case report, the mother did not know about her pregnancy and had not made any previous visits to a doctor. Frequently, the prenatal diagnosis of this condition is made between 9 and 28 weeks. Spontaneous abortion is observed in 12% of the cases of *heteropagus* twins. In our case, the abortion occurred lately[3].

Before the widespread use of ultrasound, most conjoined twins could not be identified intra-uterus. Imaging diagnosis advances have improved the approach to conjoined twins. Exact and detailed prenatal diagnosis of conjoined twins is possible and essential for optimal obstetric management and parental counseling.

CONCLUSION

In conclusion, as each set of conjoined twins is unique, the description of each new presentation is contributive to correlate and to guide imaging diagnosis. Imaging exams play an important role in proposing medical conduct, mostly because asymmetric *heteropagus* twins have less extensive vascular and visceral connections, determining a better prognosis to autosite fetus.

Author's Contribution

Rita de Cássia Sant'Anna Alves, Adriana Ubirajara Silva Petry, Andréa Pires Souto Damin – literature review and text formatting; Josenel Maria Barcelos Marçal, Adriana Vial Roehe – necropsy performance and text formatting; Bruno Hochhegger – image examinations.

RESUMO

BACKGROUND: *Gêmeos parasitas assimétricos ou heteropagos são uma ocorrência rara, com incidência de um caso em 1-2 milhões. Os gêmeos siameses são classificados de acordo com sua simetria, local de fusão e grau de duplicação.*

MÉTODOS: *Relatamos aqui uma apresentação extremamente rara de gêmeos parasitários não descritos anteriormente. São descritas alterações macro e microscópicas e discutidos aspectos relevantes dessa malformação e da importância do diagnóstico pré-natal.*

RESULTADOS: *Um caso de uma paciente de 23 anos de idade, com gestação monocoriônica, monoamniótica de gêmeos siameses assimétricos diagnosticada com 29 semanas de idade gestacional. Acreditamos que este relato chama a atenção para esta apresentação e para a importância do cuidado e manejo pré-natal. Os fetos nasceram de parto vaginal já sem vida, pesando em conjunto 1.300 gramas, e eram unidos pela região cervical esquerda.*

CONCLUSÃO: *Este relato pode ajudar a encontrar estratégias para a decisão clínica em casos futuros. O diagnóstico pré-natal é fundamental para o manejo e planejamento pré-operatório. Exames de imagem como ecocardiografia, tomografia computadorizada, ressonância magnética e ultrassonografia são factíveis e podem fornecer informações-chave sobre a gravidade e prognóstico da malformação.*

PALAVRAS-CHAVE: *Gêmeos unidos. Anormalidades congênitas. Gêmeos monozigóticos.*

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