Asymmetric parasitic twins - Heteropagus

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.

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. 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. It is an extremely rare occurrence, with an incidence of one case in 1-2 million births.
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, monomniotic conjoined twin pregnancy was discovered (29 weeks of gestational age) in an 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 was 15.5 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).

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.

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 fetuses\(^3\).

Anencephaly is described more frequently in parasitic and diciphalus 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\(^3,33\).

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 heterópagos 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 monocoriónica, monoaomniótica 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 a 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.
REFERENCES

1. Kaufman MH. The embryology of conjoined twins. Childs Nerv Syst. 2004;20(8-9):508-25.
2. Spencer R. Parasitic conjoined twins: external, internal (fetuses in fetu and teratomas), and detached (acardiacs). Clin Anat. 2001;14(6):428-44.
3. Sharma G, Mobin SS, Lypka M, Urata M. Heteropagus (parasitic) twins: a review. J Pediatr Surg. 2010;45(12):2454-63.
4. Martínez-Frias ML, Bermejo E, Mendioroz J, Rodríguez-Pinilla E, Blanco M, Egués J, et al. Epidemiological and clinical analysis of a consecutive series of conjoined twins in Spain. J Pediatr Surg. 2009;44(4):811-20.
5. Calderoni DR, Mizukami A, Nunes PH, Kharmandayan P. Thoraco-om- phalopagus asymmetric conjoined twins: report of a case and complete review of the literature. Plast Reconstr Aesthet Surg. 2014;67(1):e18-21.