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Case Study

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OMPHALOPAGUS CONJOINED TWINS: ABOUT A CASE AND REVIEW OF THE LITERATURE

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ABSTRACT

Conjoined twins, popularly known as Siamese twins, named after the birthplace of the original Siamese twins born in 1811 in Siam, Thailand[7], refer to twins physically fused in utero and therefore at birth. This is a complicated phenomenon that requires an interprofessional team approach to manage effectively.

KEYWORDS: Siamese, embryology, antenatal ultrasound sign, prognosis.

INTRODUCTION

Conjoined twins, more commonly known as Siamese twins, are a rare and serious complication of monochorionic pregnancies in which the fetuses share not only a single amniotic cavity but also body segments[1], resulting from the incomplete splitting of one embryo into two separate twins or the early secondary fusion of two originally separate embryos. Currently, with the help of ultrasound, most of these pregnancies are detected early and therapeutic intervention can be offered. The prognosis depends on the location, nature and extension of the common organs as well as the association with other malformations.[2]

Clinical Case

This is a 34-year-old parturient, non-consanguineous marriage, with no family history of twin pregnancy, G3P2 (2 live children delivered vaginally), G3 : the current pregnancy was spontaneous, followed privately by a general practitioner, she had a T1 ultrasound which objectified a monoamniotic twin pregnancy, a morphological ultrasound at 25 SA was performed by a specialist with suspicion of conjoined twins; This led to the decision to perform a multibarette fetal MRI at 28 SA which was in favor of Siamese twins of the omphalopagus type (the 2 twins have a communication at the level of the umbilical region and share the diaphragm and the liver) (**Figure 1**). The subsequent course of gestation was uncomplicated. A cesarean section was performed at 38 SA due to rupture of membranes giving birth to conjoined twins (**Figure 2**) of female Apgar 10/10, who were transferred to the pediatric surgery department for management.

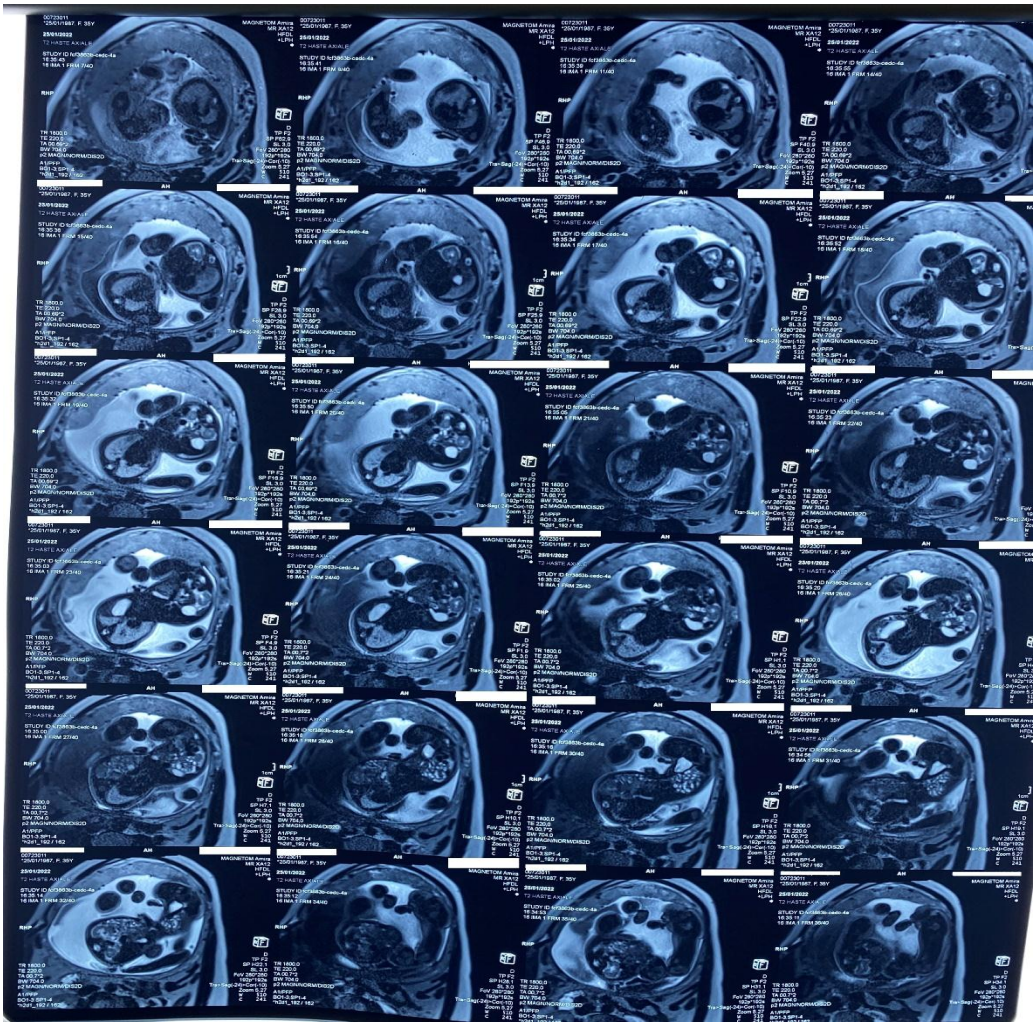

Figure 1: omphalopagus conjoined twins on MRI.

Figure 2: omphalopagus conjoined twins after postnatal extraction, overview.

DISCUSSION

This is a rare and specific complication of mono-amniotic monochorionic pregnancies. The incidence is estimated at 1/50,000 to 1/100,000 births with a female predominance (70%).[2,3] About 40-60% of cases are stillborn and 35% survive only the first 24 hours after birth.[4,2]

The pathogenesis of conjoined twins is unclear. The fission theory[5] and the fusion theory[6] are widely accepted. The fission theory suggests that the embryo undergoes incomplete division 13 to 15 days after fertilization, resulting in Siamese twins. The fusion theory assumes that two separate embryos undergo a second fusion 13 days after fertilization.

Several classifications have been described based on site of union, common organs and symmetry. Spencer's classification of Siamese is universally accepted at present. According to this classification, one finds dorsal junctions from the neural tube or ventral junctions from the anterior line. This results in 8 complete duplications distributed as follows.
- Dorsal unions occur in 87% of Siamese twins and are classified as: cephalopages, thoracopages, omphalopages, ischiopages and parapages.
- Dorsal unions occur in 13% of Siamese twins and are classified as: craniopages, rachipages and pygopages.[4,7,8]

The most frequent type seems to be the thoracopage type with a predominance of 75%.[9] Collins et al, reported that the vast majority of thoracopagus twins are complicated by major congenital heart disease (94.4%) mainly associated with monoventricular pathology.[10] The outcome of thoracopagus twins with conjoined hearts remains poor due to failure to separate conjoined and single ventricles.[11]

To this classification, we must add the incomplete duplications and the rare forms: diprosopes, dicephales, dipygus, parasitic twins and fetus in fetus.[12] There are no known cases of conjoined triplets or quadruplets.[13] However, many cases of conjoined twins in a triplet pregnancy have been described in the literature.[14] We have found cases of spontaneous triplet pregnancy or by monochorionic biamniotic assisted reproduction[14,15], spontaneous triplet, bichorionic and biamniotic pregnancies.[13]

The prenatal diagnosis of the conjoined twin is based on ultrasound. The characteristics of prenatal ultrasound diagnosis of Siamese twins include[16,7]: (1) single placenta without amniotic septum, (2) fetuses lying in the same constant position with head and body parts persistently at the same level, (3) inseparable body and skin contours, (4) fetuses facing each other with hyperflexion of cervical spines, shared organs, and a single umbilical cord with more than three vessels, (5) fewer limbs in some Siamese twins than in normal twins, and (6) abnormal flexion of the spine. MRI can be used to assist in the diagnosis if necessary.[17]

Although early diagnosis in the first trimester of Siamese twins is now a realistic option, there is a high rate of false positive diagnoses when the examination is performed before the 10th week of pregnancy[18], the diagnosis must be confirmed at ultrasound at 22 SA or the organs are more developed which allows a more precise localization of the area of attachment, analysis of duplicated structures and search for associated malformations, especially cardiac.[8]

Other imaging techniques, such as magnetic resonance imaging and 3D imaging, are useful for diagnosis and can provide additional information for prognostic purposes.[7,8]

Delivery by vaginal route is possible before 24 days of age, after this term, delivery is by caesarean section in order to avoid possible maternal complications. It should be noted that termination of pregnancy may be recommended in the case of non-viable siamese babies, but this is still debatable in view of our ethical context.[19,20]

The prognosis of conjoined twins is generally poor, they have a low survival rate. 25% of live births live to the age of surgery.[21] Only 60% of surgically isolated cases survive.[22] The prognosis is mainly related to the location and degree of the conjoined parts and the presence of associated malformations. Because of the high incidence of complex cardiac malformations, the survival rate of thoracic surgery is the lowest.[23]

A careful anatomical and vascular mapping, to determine the extent of organ sharing and a paramount consideration for separation surgery; it is an expensive surgery, involving multidisciplinary teams.

CONCLUSION

Conjoined twins represent one of the rare congenital anomalies, and one of the greatest challenges of modern pediatric surgery to date. A temperamental diagnosis of Siamese twins pregnancy is crucial to choose the best therapeutic option in terms of pregnancy termination or continued gestation with surgical separation of the twins.

REFERENCES

1. Sebire NJ, Sepulveda W, Jeanty P, Nyberg DA, Nicolaides KH. Multiple gestations. In *Diagnostic Imaging of Fetal Anomalies*, DA Nyberg, JP McGahan, DH Pretorius, G Pilu (eds). Lippincott Williams & Wilkins: Philadelphia, PA, 2003; 777– 813.
2. Boujoual M, Madani H, Benhaddou H, Belahcen M. Jumeaux conjoints au niveau d´une omphalocèle commune avec extrophie cloacale et ambigüité sexuelle. Pan Afr Med J, 2014; 17: 243.

3. Nahar N, Khan N, Islam SM, Chakraborty RK, Rima SZ, et al. (2015) Conjoined twins with single heart and liver - a case report. Mymensingh Medical Journal MMJ, 24: 172-176.
4. Bousselamti Amal. [Thèse médecine] Rabbat: Université mohammed-V Rabat; 2016. Jumeaux siamois à propos de 8 cas avec revue de la littérature.
5. Kaufman MH. The embryology of conjoined twin. Childs Nerv Syst, 2004; 20(8–9): 508–25.
6. Boer LL, Schepens-Franke AN, Oostra RJ. Two is a crowd: two is a crowd: on the enigmatic etiopathogenesis of conjoined twinning. Clin Anat, 2019; 32(5): 722–41.
7. Mathew RP, Francis S, Basti RS, et al. Conjoined twins–role of imaging and recent advances. J Ultrason, 2017; 17(71): 259–66.
8. Benchiba I, Saoud MK, Mamouni N, Errarhay S, Bouchikhi C, Abdelaziz B. Diagnostic à terme de jumeaux conjoints. Pan Afr Med J, 2020 Jan 10; 35: 8.
9. Ozcan HC, Ugur MG, Mustafa A, et al. Jumeaux siamois dans une grossesse triplée. Un dilemme obstétrical rare. Saudi Med J, 2017; 38(3): 307–9.
10. Collins, R.T.; Weinberg, P.M.; Gruber, P.J.; Sutton, M.G.S.J. Conjoined Hearts in Thoracopagus Twins. *Pediatr. Cardiol*, **2011**; *33*: 252–257.
11. Tan, K.L.; Tock, E.P.C.; Dawood, M.Y.; Ratnam, S.S. Conoined Twins in a Triplet Pregnancy. *Am. J. Dis. Child*, **1971**; *122*: 455–458.
12. Guèye M, Kane Guèye SM, Ndiaye Guèye MD, Diouf AA, Mansour Niang M, Diallo M *et al*. Accouchement de jumeaux conjoints de découverte fortuite au cours du travail au CHU de Dakar. Pan Afr Med J, 2012; 12: 102. Epub 2012 Aug 13.
13. Ozcan HC, Ugur MG, Mustafa A, Kutlar I. Conjoined twins in a triplet pregnancy: a rare obstetrical dilemma. Saudi Med J. 2017; 38(3): 307-30.
14. Xin-Lu Meng, Yuan Wei, Yang-Yu Zhao. Conjoined Twins in a Spontaneous Monochorionic Triplet Pregnancy. Chin Med J (Engl) 2018; 131(20): 2492-2494.
15. Jabari S, Carbon R, Besendörfer M, Hartmann A, Rompel O, Hoerning A et al. Asymmetric Omphalopagus in a Triplet after In Vitro Fertilization: A Rare Case of Conjoined Twinning. Case Rep Pediatr, 2018 Aug 23; 2018: 9349606.
16. Mian A, Gabra NI, Sharma T, et al. Conjoined twins: from conception to separation, a review. Clin Anat, 2017; 30(3): 385–96.
17. Wataganara T, Ruangvutilert P, Sunsaneevithayakul P, et al. Three-dimensional ultrasound for prenatal assessment of conjoined twins: additional advantages? J Perinat Med, 2017; 45(6): 667–91.
18. Sepulveda, W.; Munoz, H.; Alcalde, J.L. Conjoined Twins in a Triplet Pregnancy: Early Prenatal Diagnosis with Three-Dimensional Ultrasound and Review of the Literature. Ultrasound Obstet. Gynecol, 2003; 22: 199–204.
19. T.C. Mackenzie, T.M. Crombleholme, M.P. Johnson, L. Schnaufer, A.W. Flake, H.L. Hedrick, et al. The natural history of prenatally diagnosed conjoined twins J Pediatr Surg, 2002; 37(3): 303–309. 21.
20. M. Hovorakova, R. Peterkova, Z. Likovsky, M. Peterka A case of conjoined twins cephalothoracopagus janiceps disymmetros Reprod Toxicol, 2008; 26: 178–182.
21. Kaufman MH. The embryology of conjoined twin. Childs Nerv Syst, 2004; 20(8–9): 508–25.
22. Brizot Mde L, Liao AW, Lopes LM, et al. Conjoined twins: prenatal diagnosis, delivery and postnatal outcome. Rev Bras Ginecol Obstet, 2011; 33(5): 211–8.
23. Sage E, Thomas A, Sundgren N. Conjoined Twins: Pre-Birth Management, Changes to NRP, and Transport. Semin Perinatol, 2018; 42(6): 321–8.