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Delivery of a Dicephalic Siamese Parapagus in the Community Clinic: Case Report and Review of the Literature

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Abstract

Twins are one of the congenital anomalies described as phenomenal or mysterious. It is a rare and specific complication of monochorionic monoamniotic pregnancies resulting either from the late and incomplete splitting of one embryo into two distinct twins, or from the early secondary fusion of two initially separate embryos. These anomalies are often amenable to early diagnosis by ultrasound in the first trimester, which is still lacking in our community due to the uneven resources available. The aim of our clinical case study was to remind obstetric clinicians and the medical imaging sector that this very rare disease exists, hence the importance of obstetric ultrasound in the first trimester as an essential diagnostic tool to enable a management strategy to be put in place to avoid any surprises that could jeopardise the vital prognosis of the pregnant woman, particularly during parturition.

Keywords

Siamese, Ultrasound, Central African Republic

1. Introduction

Siamese is an exceptional obstetric malformation, occurring once in every 50,000 to 60,000 births [1] [2], with a perinatal mortality rate of 60%. It results from

incomplete separation of the embryonic disc before the third week of pregnancy, the aetiology of which remains poorly understood. In fact, two theories explain its aetiopathogenesis: the theory of incomplete fusion of a single embryo and that of fusion between two embryos [3]. Conjoined twins are classified according to the most important site of union, with the suffix pagus meaning fixed. Several classifications have been described [4]. Prenatal diagnosis of conjoined twins was first reported by Wilson *et al.* [5], who demonstrated the crucial role of ultrasound not only in diagnosis but also in establishing the degree of conjunction. Prenatal diagnosis of Siamese twins, the main aim of which is to assess the viability of the twins, offers a wide range of management options depending on the presumed prognosis. These include termination of pregnancy, continued pregnancy and delivery with subsequent separation, or even selective fetal reduction in higher-order multiple pregnancies. Despite a detailed assessment of prenatal lesions, this entity remains a real surgical challenge, as it is not always possible, not always easy, and the prognosis is uncertain, even with experienced teams. In the Central African Republic, a country with limited resources, this case of siamese poses a real management problem, as it is often diagnosed late and compromises the woman's life. The aim of our analysis, through our clinical case and a review of the updated literature, is to study the value of early antenatal diagnosis while detailing the various difficulties encountered in the management of this type of anomaly and to insist on multidisciplinary collaboration in a tertiary hospital structure in the hope of improving the mother and newborn baby prognosis.

2. Observation

This was a 20-year-old parturient, 2nd gesture and 2nd parity, spontaneous pregnancy partially monitored with only three contacts; no obstetric ultrasound had been performed. She was referred to the department for retention of the last head following an initially unsuccessful unassisted home birth. Her family then took her to the local Level I maternity hospital. When the foetus could not be expelled despite attempts at obstetric manoeuvres, the parturient was referred to the maternity ward of the community university hospital for treatment. On admission, the fetus was in good general condition, BP 120/76 mmHg, pulse 105/min FR 22 cycle/min, 3 CU/10mn, all fetal limbs were at the vulva and the back was forward, while fetal heart sounds were absent. On vaginal touch, the fetal head was noted to be above the superior strait at the −4/5th level. On the basis of the above, we confirmed the referral diagnosis and sought to understand the aetiology. We carried out an obstetric ultrasound in the delivery room, which revealed two fetal cephalic poles with biparietal perimeters measuring 87 mm and 90 mm, respectively, without revealing the other fetal podalic pole or the plane of the back, which was unusual, leading us to conclude that there was a twin pregnancy malformation with probable fusion of the cephalic poles. Hence the indication for a maternal rescue caesarean section, which resulted in the laborious extraction of a stillborn Siamese twin weighing 3400 g female, followed by delivery of a single placenta and

closure of the placenta section by section. Figure 1 is an image of our case.

Figure 1. Raise figure and put image of case twins joint.

3. Comment

Conjoined twins are rare, with a prevalence of between 1 in 50,000 pregnancies in utero and 1 in 250,000 live births [1] [2]. Its rarity is due to the rarity of twin pregnancies (approximately 1% to 2% of pregnancies), the rarity of monozygotism (approximately 35% of twin pregnancies), the rarity of embryo division after the 14th-day post-fertilisation or partial fusion of the two primitive lines leading to conjoined twins. Conjoined twins are generally incompatible with life, meaning that 65% of cases are stillborn, while of those born alive, 35% die within the first 24 hours [6]. Only 25% survive to an age where surgical separation can be considered [7]. The aetiopathogeny of conjoined twins has always been, and remains to this day, a subject of debate, with many hypotheses being considered and others completely abandoned:

- Syphilis was implicated in the first half of the twentieth century.
- Drunkenness during coitus.
- Husband's alcoholism [8].

Others are still being discussed. However, none of them has really been proven.

- Several authors have noted "epidemics of conjoined twins" in Jerusalem [9], South Africa, and Sweden. These authors suspected a seasonal effect on the production of conjoined twins through changes in oxygen concentration and temperature.
- A report by Rosa *et al.* (1987) mentioned exposure during pregnancy to Griseofulvin and low-dose radiation [10] [11].

There are currently 2 main theories concerning the mechanism of formation of conjoined twins:

- The hypothesis of late incomplete fission of a single embryo is the most widely accepted.
- Another hypothesis that is increasingly evoked is a fusion between two initially separate mono-ovular embryonic discs. Others accept that the two theories can coexist. Several classifications have been described according to the site of union, common organs, and symmetry. Saint-Hilaire established one in 1832, based on the description of the external site of union and symmetry. The result

is eight types described in the literature: ventral unions occur 87% of the time and are classified as follows: cephalopagus (11%), thoracopagus (19%), omphalopagus (18%), ischiopagus (11%) and parapagus (28%). Dorsal unions occur in 13% of Siamese twins and are classified as craniopages (5%), rachipages (2%) and pygopages (6%) [2] [3]. Another classification described takes into account the shared body site as follows: anterior or thoracopagus, posterior or pygopagus, cephalic or craniopagus and caudal or ischiophagus, the post-natal prognosis is linked to cardiac damage; or neurological and urinary anomalies [12] [13]. For ultrasound analysis, Duhamel's classification, established in 1966, seems the most appropriate. He synthesised these different classifications, taking into account embryogenesis data. He differentiated between JCs with parallel axes, which are "side by side", tetra-pages, which are "face to face," and crucipages with opposite axes to which omphalopages are brought closer. In 1976, Wilson carried out the first ultrasound scan of JC at 35 weeks' gestation, and since then, antenatal diagnosis has been made at an increasingly early stage. In 1989, Gruter discovered thoracopic twins at 16 weeks' gestation, and then in 1997, Hill made the diagnosis of JC at 7 weeks' gestation. [2] [6] [4]. Prenatal diagnosis can be made as early as the first trimester, but this must be confirmed by ultrasound at 22 weeks gestation when the organs are more developed and the diagnosis becomes easier. Ultrasound identification of one of the following classic signs may suggest the diagnosis [14] [15]: Ultrasound signs in the first trimester of pregnancy:

- There is a single identifiable gestational sac with a single trophoblastic crown, as in any monozygotic twin pregnancy.
- There is only one amniotic sac: this should not be confused with the extra-embryonic coelom. The amniotic membrane becomes permanently attached to the chorionic membrane between the 11th and 13th AUG. As long as the extra-embryonic coelom is visible, its echogenicity is greater than that of the amniotic fluid.
- There is only one vitelline vesicle located extra-amniotic, and its identification confirms that the fluid zone around the amniotic membrane is indeed the coeloma. In JC cases, there are never two vitelline vesicles, unlike in monozygotic monoamniotic twin pregnancies.
- A single embryonic mass is visible: it is compact when there is a lot of coalescence, and often has a bifid appearance. However, a superficial examination may overlook the anomaly, especially when there is only one cephalic pole, particularly in the case of cephalopagous JC. On the other hand, when there is only minimal compression, two masses may be present.

4. Conclusion

Siamese twins are one of the rarest congenital malformations and still present a major challenge in terms of antenatal and neonatal treatment. Ante-natal diagnosis is more often established in the first trimester, but in our country, with limited

resources and despite low household incomes, it is sometimes delayed and only observed at the time of delivery and in the face of the various obstetric complications that ensue. Incorporating compulsory obstetric ultrasound into the criteria for the holistic management of pregnant women could avoid the surprises and complications associated with the dystocic delivery of Siamese twins, in addition to training providers in the obstetric sector.

Ethical Consideration

The patient's informed consent had been obtained.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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