A RARE CASE OF DISCORDANT ANOMALY IN A DICHORIONIC DIAMNIOTIC TWIN PREGNANCY: TESSIER CLEFT WITH SEVERE HYDROCEPHALUS IN A LOW RESOURCE SETTING

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ABSTRACT

Introduction: **Congenital anomalies in twin pregnancies pose significant challenges for prenatal diagnosis and management, especially in low-resource settings. Discordant anomalies, especially those involving the central nervous system (CNS) and craniofacial structures, are rare but associated with high perinatal morbidity and mortality rates.**

Case Presentation: **We report a rare case of Severe Tessier cleft in a discordant twin of a 27-year-old Gravida 5 Para 0+4 woman who booked for antenatal care at 31 weeks and 5 days' gestation with no prior fetal anomaly scans. She had a dichorionic diamniotic twin pregnancy, with ultrasound revealing one of the fetuses affected by severe hydrocephalus.**

She presented at 37 weeks and 6 days of gestation and had an elective cesarean section. The first twin, a healthy female, had a birth weight of 3.2 kg and good APGAR scores. The second twin, a male with multiple craniofacial and skeletal congenital anomalies. The APGAR scores were 6 at the first minute and 7 at the fifth minute, but died at about the third hour of life.

Conclusion: **This case underscores the importance of prenatal anomaly assessment and care. Multidisciplinary care, including in-depth counseling and timely intervention, is crucial to optimizing maternal and neonatal outcomes.**

Keywords: Congenital hydrocephalus, Tessier clefts, Twin deformity, Congenital anomaly, Discordant twins, Case report

INTRODUCTION

Congenital structural anomalies are more common in a twin setting, and usually, only one twin is affected.[1] As such, structural anomalies have been observed to occur in about 1 in 17 dichorionic, 1 in 10 monochorionic, and 1 in 5 monoamniotic twin pregnancies.[2] In contrast, chromosomal anomalies are probably somewhat less common in a twin setting, because aneuploidy is less likely to survive in a twin setting.[3] Congenital anomalies in one fetus of a dichorionic diamniotic (DCDA) twin pregnancy pose significant diagnostic and management challenges.

Discordant anomalies, especially those involving the central nervous system (CNS) and craniofacial structures, are rare but associated with high perinatal morbidity and mortality rates.[4] Tessier clefts represent a spectrum of craniofacial anomalies that extend beyond typical cleft lip and palate classifications, often involving complex deformities of the orbits, maxilla, and mandible[5,6], and hydrocephalus is one of the associated findings in this condition.[7] When severe, the presence of hydrocephalus signifies underlying syndromic or structural defects that are associated with poor prognoses,[8,9] especially when it occurs in the background of other serious craniofacial malformations. The actual prevalence of fetal craniofacial anomalies is likely underestimated, as many early gestational fetal deaths are often disregarded without investigation. Additionally, the frequency of abortions involving these fetuses remains unknown.[8] According to Tessier (1990), the clefts were noted to be rare in live births and found to occur with an incidence of 1.43 to 4.85 in 100,000 births.[10] The recent advancements in prenatal screening and diagnostic techniques have improved the early detection of congenital anomalies in pregnant women. Also, the sensitivity of ultrasound in detecting craniofacial anomalies has improved, but still varies significantly and is best detected at fetal anomaly ultrasound scanning. However, in late pregnancy, most studies indicate that hydrocephalus is one of the most easily identified congenital anomalies through ultrasonographic examination.[11]

The incidence of severe craniofacial anomalies and hydrocephalus in a twin gestation with only one of the fetuses affected is rare, as the etiology is unknown in most cases. Although Tessier clefts are uncommon, however, the occurrence of the more common orofacial clefts has been linked to factors such as alcohol and teratogenic antibiotic use, e.g, tetracycline, trimethoprim-sulfamethoxazole, Aminoglycosides during early pregnancy, maternal exposure to typhus and typhoid infections, and the absence of iron and folic acid supplementation during pregnancy.[12–14] We present a case of selective twin deformity (hydrocephalus and severe craniofacial anomalies (Tessier cleft) in a dizygotic twin to highlight the occurrence of this severe anomaly selectively in twin gestation and the management challenges associated with care.

CASE PRESENTATION

A 27-year-old Gravida 5 Para 0 +4 woman presented for booking at a gestational age of 31 weeks and 5 days at a Private Hospital in Ibadan, Oyo State, Nigeria. She did not have any complaints at the initial presentation, and she has never been registered at any other facility. She had no anomaly scan done in the index pregnancy. Her antenatal booking blood investigations were essentially normal. However, Obstetric ultrasound scans done in the third trimester revealed a dichorionic diamniotic twin with hydrocephalus and multiple anomalies (fused meningocele, dilated lateral ventricles, porencephalic cyst, absent 3rd ventricles) in the leading twin, as shown in Figure 1. Following this, she was counselled on the need to have an elective cesarean section at term. She was commenced on hematinics, and a 3D/4D ultrasound scan was requested to determine the details of the detected anomalies. Additionally, she had no chronic medical conditions and no pregnancy-associated complications. She has had four previous spontaneous miscarriages, though the causes were not evaluated before the index pregnancy. She takes alcohol occasionally, but she does not smoke cigarettes or take any recreational drugs.

The patient was not compliant with her antenatal clinic attendance. After, her initial antenatal care (ANC) visit, she presented at gestational age of 37 weeks and 6 days, when she was admitted and subsequently had a Caesarean section. Findings at surgery include: A well-formed lower uterine segment, Twin 1 was a female with a birth weight of 3.2kg and APGAR score of 8 at 1 and 9 at 5 with normal morphology as shown in figure 2A while Twin 2 was a male with hydrocephalus and multiple craniofacial anomalies (absent eyes bilaterally, absent nose, fused upper lips), birth weight was 3.4kg and APGAR score was 6 at 1 and 7 at 5 as shown in figure 2B.

The parents were informed of the deformities and the probable prognosis of the deformity in the 2nd twin, including the possibility of not being compatible with life. The need for referral to a tertiary healthcare

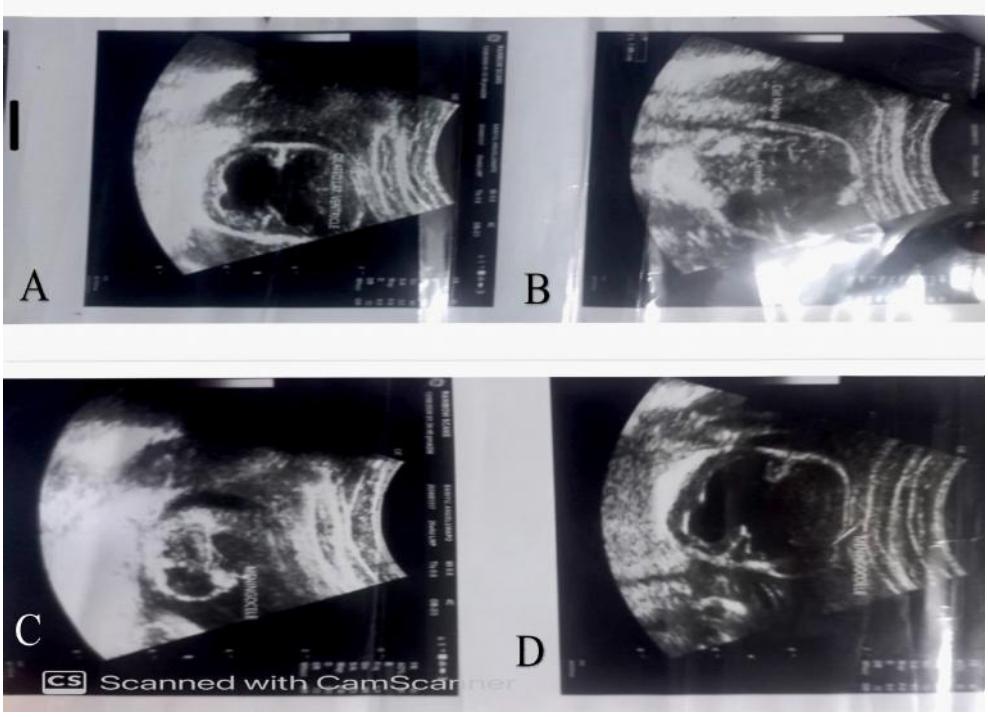

Figure 1: Antenatal fetal Ultrasound scan of the fetal head of the twin with anomaly showing A.Coronal view of the dilated lateral ventricles. B. Abnormally large Cerebellum and Cisterna magna. C. longitudinal plane of meningocele and D. transverse plane of meningocele

Figure 2: Image of the normal twin (A)and twin with malformations (B).

centre was also discussed with the parent. However, the baby died about three hours after birth. No postmortem examination was performed at the parents' request. Genetic testing was declined, and grief counseling was arranged for the parents.

She was thereafter discharged from the hospital on the 4th day post-surgery and followed up in the post-natal clinic. At the routine postnatal clinic, the mother had remained stable clinically, and her surviving infant had gained weight appropriately (weight was 4.6kg) and was adjudged by the mother to be feeding well. The mother was counselled on the possible risk factors to avoid in subsequent pregnancies. She was counselled to book for antenatal care early and be compliant in her next pregnancy. The risk of recurrence was discussed with her, and the importance of a fetal anomaly ultrasound scan was emphasized. She was discharged to the family planning clinic, and the baby was discharged to the well-baby clinic.

DISCUSSION

This case exemplifies the critical importance of early prenatal care and anomaly screening, particularly in multifetal gestations. DCDA twins have independent placental and amniotic environments, allowing for the possibility of discordant anomalies.[15] Severe hydrocephalus, particularly when associated with craniofacial defects like Tessier clefts, suggests complex embryologic disruptions, possibly syndromic or linked to neural crest cell migration abnormalities.[16,17] The origin of Tessier clefts remains unclear. It is believed that these groups of malformations may arise during a critical embryonic phase of stem cell proliferation and differentiation in the face and brain.[16,18]

The Tessier clefts are classified by an anatomical system that denotes clefts from the midline (0) to lateral facial planes (up to 14).[19] The combination of severe hydrocephalus and extensive craniofacial anomalies, as in the index case, is exceedingly rare and poses significant ethical, diagnostic, and management dilemmas. These malformations are usually detected before the 24th week of pregnancy. With the current advancements in ultrasonographic diagnostics, early recognition is considered standard practice.[20]

The involvement of environmental factors in the development of craniofacial anomalies is documented in the literature. Prenatal infections, including toxoplasmosis, rubella, syphilis, and cytomegalovirus, appear to have the strongest association.[21] In addition, maternal use of medications such as misoprostol, dextromethorphan, nalidixic acid, and cephalosporins in early pregnancy may elevate the risk of developing craniofacial anomalies.[22] Some central nervous system malformations, such as meningocele, dilated lateral ventricles, porencephalic cyst, and absent 3rd ventricle, have been closely associated with a lack of folic acid supplementation in pregnancy. Therefore, a possible explanation for this could have been non-compliance with her routine haematinics during her first trimester. This patient did not have preconceptional folic acid and was non-compliant with folic acid intake early in pregnancy. This underscores the importance of early and regular antenatal follow-up. One major problem with obstetric practice in sub-Saharan Africa remains illiteracy and a lack of women's empowerment, which makes quality health care inaccessible to women.

Also, late booking, as seen in this case, limits opportunities for early anomaly detection and informed parental decision-making, including potential pregnancy termination where legally and ethically permissible.[23] Routine first and second-trimester scans are essential for identifying major anomalies and improving outcomes through early referral and intervention.

In the assessment and counseling of women with twin pregnancies and discordance for a specific malformation, it is crucial to identify which fetus is affected. This is especially crucial in a dichorionic pregnancy, in which selective fetocide can be considered.[20,24] Although it is preferably performed before the age of viability.[25] Our patient presented for antenatal booking at a gestational age of 31 weeks and 5 days, when it would have been too late to perform such interventions.

Despite the severity of the anomalies in one fetus, the presence of a normal co-twin reinforces the need for tailored perinatal strategies that optimize outcomes for the healthy fetus while managing the risks posed by the affected twin.[26] In such cases, delivery timing must balance fetal maturity with the potential for complications like polyhydramnios, preterm labor, or intrauterine demise of the affected twin. The management of these complexities requires a multidisciplinary approach to the care of these women and their pregnancies. This multidisciplinary team is mostly accessible in tertiary hospitals. Unfortunately, most women dread referrals to the tertiary health facilities for various reasons, including bureaucratic bottlenecks and cost of care, among others, for which they are reluctant to be referred for tertiary care. Also, poor funding of referral hospitals in low-income settings like ours has resulted in the sub-optimal functioning of these hospitals and the poor outcomes for referred patients

In conclusion, Craniofacial congenital anomalies such as Tessier defects are severe congenital malformations, often diagnosed prenatally, although sometimes diagnosed later in pregnancy. The outcome is poor, with a high proportion of pregnancies terminated for severe fetal anomaly and a high perinatal mortality. Tessier defects and other associated syndromes appear to be multifactorial disorders, arising from the interaction of genetic predisposition and environmental factors. While their exact causes remain complex, several factors have been implicated. Evidence suggests that preventive measures, such as folate supplementation, may be effective for individuals at risk, especially in low-resource settings like ours, where macro- and micronutrient deficiency is higher. Early initiation of antenatal care components will ensure early diagnosis and prompt antepartum support and care.

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