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Epigastric Heteropagus Conjoined Twin – A Case Report

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***Abstract*: Background: Conjoined twins are babies attached to each other at some point. They occur in 1 in about every 200,000 live births and may be described as Symmetric (Diplopagus) or Asymmetric (Heteropagus). Heteropagus refers to conjoined twins where an apparently normal twin- the autosite- has attached to it an incomplete member- the parasite- and are exceedingly rare with a worldwide incidence of about 1 in 1-2 million births. The form of these heteropagus twins in which the parasite is attached to the epigastric region of the host referred to as epigastricheteropagus is even rarer. Case report: Here we report a day-old female neonate delivered by Spontaneous Vertex Delivery in a Primary Health Centre to a 20year old P_{3+0} 3A with a parasite attached at the epigastric region. The parasite comprised a trunk to which two identifiable lower limbs were attached, with a single stalk attached to its upper pole from which two hands with identifiable digits arose. There was also a coexisting omphalocoele in an otherwise normal autosite. Abdominal USS and CT scan showed a single liver with no major connection between the babies, Intravenous Urogram confirmed the presence of two kidneys normal in position and excretion with normal collecting system within the autosite and none in the parasite. Doppler USS of the stalk eliminated major vascular connection between the two. Separation was successfully done at 2 months of life and patient did well postoperatively. Child is currently five (5) years old and growth and development proceeding normally. Conclusion: Epigastric heteropagus twins require multidisciplinary approach; careful planning and separation plan individualized according to the need of emergent separation and degree of organ fusion.**

***Keywords*: heteropagus, conjoint, twins, separation, child.**

1. Introduction

Since prehistoric times conjoined twins have continued to fascinate mankind. The first report of this rare aberration of human reproduction was by Ambrose Piere in the 16th century [1]. With a worldwide incidence of 1 in every 50,000 to 100,0000 births [2], the term conjoined twins refer to babies who are joined at some point, and areeither Symmetric (Diplopagus) or Asymmetric (Heteropagus) [2]-[4]. Heteroghagus a term derived from the Greek words 'pagus' which means 'that which is fixed' refers to a rare form of conjoined twin in which twins are asymmetrically attached with 1 twin the auto site nearly normal anatomically while the parasite is attached in a non-duplicate fashion to any part of the body or even within the autosite [2]-[4]. The former may be referred to as exoparasiticheteropagus twin and the latter endoparasitic heteropagus [1]. Endoparasiticheteropagus in which the parasite is attached to the epigastric region of the autosite is called epigastricheteropagusand occurs rarely amongst the 1 in 500,000-2million live births that are heteropagus conjoined twin [3], [6], [7] with only 71 cases reported in English literature worldwide from 1946 to 2015 [8]

The major challenge to the management of this anomaly is occasioned by its rarity and thus low individual surgeon exposure coupled with the fact that Epigastric Heteropagus twins may present with more complex manifestation than typical conjoined twins.

We present here a case of epigastricheteropagus conjoined twin admitted on the first day of life, who was successfully managed.

2. Case Report

A 1-day-old, full-term female born to a 20-year-old P_{3+0} 3A via SVD at a Primary health facility after a prolonged labor. No previous history of twining in mother, no family history of same. No history of assisted reproductive techniques. Pregnancy was booked with 3 visits but no antenatal ultrasound scan done. No history of cigarette smoking or ingestion of alcohol or un-prescribed medication. Father is 30yrs old peasant farmer. Baby is able to feed well and pass stool and urine.

Examination revealed an autosite that was active and pink, moving all limbs actively with no features of dysmorphism. Combined weight was 3.1kg.S ystemic examination was normal except for a co-existing omphalocoele major with an intact covering.

Parasite was attached to the epigastric region of the autosite and comprised two lower limbs and a single stalk on its upper pole giving rise to two hands with digits. There were well developed hip joint and knee joint with is also flexion

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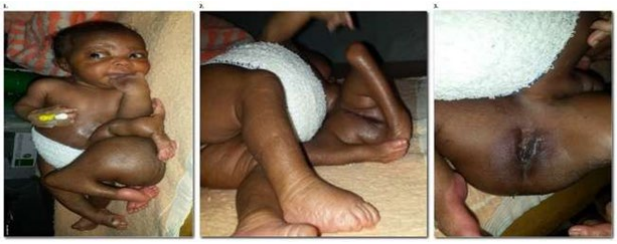

Fig. 1. The picture of the baby with the parasiric twin (fully formed lower limbs and genitalia) connected to her

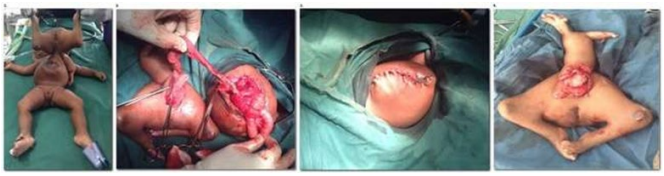

Fig. 2. Intra-Operative Pictures showing the parasitic twin before and after it wa completely excised from the baby

contracture to about 60[0] in both joints. The limbs however show no active movements, and no response to tactile stimulus. There were also identifiable female external genitalia. (Fig 1).

Abdominal USS and CT scan showed a single liver with no major connection between the babies, Intravenous urogram confirmed the presence of two kidneys normal in position and excretion with normal collecting system within the autosite and none in the parasite. Doppler USS of the stalk eliminated major vascular connection between the autosite and parasite.

Omphalocoele was managed non-operatively of done and it healed well.

Separation was done 2nd month of life with intra operative finding of a loop of small intestine running within the connecting stalk and ending blindly distally on the ventral surface of the autosite liver and proximally (in the parasite). Omphalocoele was not repaired to avoid abdominal compartment syndrome. (Fig 2).

Patient did well post operatively and was discharge on the 14th after separation.

The ventral hernia that resulted from the omphalocele was repaired at one (1) year of age.

Follow up has been uneventful, with growth and development progressing normally.

3. Discussion

The worldwide incidence of conjoined twin is estimate at 1 in 50,000 to 200,000 live births [2], [5], [7], [10]-[12], with 10% of these being heteropagus twins [4].

Incidence however varies from 1 in 88,000 in Sweden to 1 in 97,500 to in the USA and in China 3 in 100,000 [5].

The earliest reported case of conjoined twin in Nigeria in was in Sokoto in 1935[14]. A total of 18 cases have so far been reported in Nigeria as at 2014 [14].

The incidence of heteropagus twins on the other hand has been estimated at 1 in 2 million an estimated 34% of heteropagus conjoined twins are joined at the epigastric region[14], with 71 cases of epigastricheteropagus reported in English literature worldwide from 1946 to 2015 [8].

The exact etiology of conjoined twins is complicated and not very clear. Although monozygotic twining can be easily induced ex vivo by administration of various teratogenic agents, the exact mechanism of spontaneous twining remains unknown. Various theories have been propounded, to explain how these monozygotic mono-amniotic-monochorionic twins arise [12], [13]. Fission theory proposes that conjoined twins are basically due to incomplete division of the inner cell mass at around 14days gestation.[1], [2], [5], [6], [12], [13] the fusion theory on the other hand submits that conjoined twins result from secondary union of two monovular embryonic discs [4], [5]. Selective ischemic damage of the fetus in utero due to unbalanced distribution of placental blood, leading to the death of partial resorption of one of the twins has also been propounded to explain the origin of conjoined twins [2]-[6].

Although the incidence of conjoined twin is more common in females than in males with a F:M [3:1]; [2], [5], [9] in the case of epigastricheteropagus twins, male preponderance has been reported with 78-80% of patients being male,[4], [5], [8] some reports submit the M:F of Epigastric Heteropagus Conjoined twin at 5:1; our report however features a female autosite (and parasite).

In the case presented, the parasite comprised a torso with well-developed lower limbs and a single stalk giving rise to two hands with all limbs devoid of skeletal muscle, this is in keeping earlier reports which demonstrated that the defective twin is usually composed of extremities but may contain viscera, but only rarely a beating heart and even more rarely a brain [4], [15]. The least common supernumerary structure organs as the heart and the neural tube featured in literature. Oversensitivity of the brain, heart and lungs to ischemia has been proposed as a possible reason for this occurrence [4], [15].

The limbs in the parasite in this case report lacked spontaneous movement and unresponsive to tactile stimulus skeletal muscle this is in keeping with previous reports [5], the failure of myoblast differentiation and subsequent skeletal muscle atrophy due to absence of proper innervations has been proposed in an attempt to explain this occurrence [4], [5].

In this case report, the twins were attached by a stalk containing a strand of cartilage, findings consistent with previously case reports, what was however rare is that the stalk also contained a blind ending loop of small intestine attached to the autosite liver. This is a rarity as though communication with the autosite abdominal cavity is common 80%[8], the stalk rarely

contains viscera [4]. No literature has reported epigastricheteropagus twins with vascular or bony connection [2].

Internal organs of the autosite were normal in this case, similar to earlier reports. However, the autosite may have associated abnormalities as well, most commonly congenital heart defects especially ventricular septal defects [4]. Omphalocoele as seen in this case are also common [5], occurring in up to 50% of epigastricheteropagus twins. The interference of the connecting cartilage bridge with closure of the abdominal wall during the later stages of gestation has been proposed as a cause of the occurrence of the Omphalocoele.[5]

Other anomalies that have been reported in the autosite include abdominal wall hernias, congestive heart failure, intestinal obstruction, cleft lip, chonalatresia, imperforate anus and rarely spinal cord deformities [3]-[5], [9].

Regarding timing of separation, in the case reported, separation was done at 2months of age, well within the time frame suggested by Spitz and Kielyof between 2-4 months of age. However, emergency separation would have been considered at whatever age in the presence of stillborn twin, intestinal obstruction, rupture of Omphalocoele, heart failure, obstructive uropathy or respiratory failure.[9]There have however been reports of successful separation in the much earlier in life [5], [7].

Regarding prognosis, despite the unusual clinical appearance epigastricheteropagus twins have a good prognosis [4], [5]. Outcome however depends on the extent of organs shared early diagnosis, intensive prenatal management and proper route of delivery are critical for successful separation [3], [4], [6].

4. Conclusion

Epigastric heteropagus twins may present in a variety of ways with variable levels of communications and associated anomalies thus their management requires a multidisciplinary approach, requiring careful planning and experience with time if separation individualized and separation plan individualized according to the need of emergent separation and degree of organ fusion.

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A. Compliance with Ethical Standard

We affirm that full compliance with ethical standard was observed throughout.

B. Conflict of Interest

The authors declare that they have no conflict of interest.

C. Ethical Approval

Ethical approval was obtained from the ethical board of Abubakar Tafawa-Balewa University Teaching hospital, Bauchi to conduct the study.

D. Informed Consent

Was obtained from the patient's parent.

E. Author Contribution

All authors have been directly involved with the various aspects of the study. We attest to the fact that all authors have participated in the research, read the manuscript, attest to the validity and legitimacy of the data.

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