



Case Report

A Rare Type of Asymmetric Conjoined Twins: Epigastric Heteropagus and Omphalocele

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Abstract

Background

Parasitic Conjoined Twins (PCT), an extremely rare condition with a severely defective fetus attached externally to a normal twin (autosite), occur in just 1-2 million live births. Epigastric heteropagus twins (EHT) involve attachment to the autosite's epigastrium. This study presents a case of EHT, highlighting the importance of timely separation surgery and a multidisciplinary approach for successful management. **Case presentation:** A 23 days-old, full-term 3.5 kg male baby, the healthy infant had an omphalocele just below the attachment of the parasitic twin and the parasitic bowel was slightly prolapsing through it. The parasite was attached to the epigastrium of the autosite and it possessed two rudimentary upper limbs, two well-developed lower limbs with exhibited pain and tactile sensitivity without spontaneous movement, and a well-developed scrotum and penis, but absent testis and absent anus. The parasite was passing urine. An echocardiogram of the autosite revealed a Tetralogy of Fallot (TOF)-patent foramen of ovale (PFO). Surgery to separate the twins was performed on 23th day of life with no short-term complications. **Discussion:** Epigastric heteropagus twins (EHT) are rare type of monozygotic monochorionic asymmetrical conjoined twins. Delayed Surgery is reported to increase successful separation as it allows enough time to delineate the internal anatomy, planning and assemble multidisciplinary teams for appropriate preoperative, intraoperative and postoperative management. **Conclusion:** We report successful separation of epigastric heteropagus autosite with an omphalocele.

Keywords: Asymmetric Conjoined twining; Autosite; Parasites; Epigastric Heteropagus.

Introduction

Parasitic Conjoined Twins (PCT) constitute an exceptionally rare and remarkable anomaly. In which a profoundly defective fetus or fetal components are externally attached to a relatively normal twin, known as the autosite. This extraordinary occurrence is documented in just 1-2 out of every million live births. In the unique case of Epigastric Heteropagus, the parasitic twin is specifically attached to the epigastrium of the autosite.

The primary aim of this study is to present a case of Epigastric Heteropagus and underscore the critical importance of timely intervention and a multidisciplinary approach for the successful management of such cases.

Case presentation

We are presenting the case of a 23-day-old male infant delivered via caesarean section. The antenatal diagnosis revealed Parasitic Conjoined Twins with an omphalocele in the third trimester. The infant was born at full term with a birth weight of 3.5 kg and arrived in good health with Apgar scores of 7/10 and

9/10 at the first and fifth minutes, respectively. The omphalocele was located just below the attachment site of the parasitic twin and showed slight protrusion of the parasitic bowel.

The parasitic twin was firmly attached to the epigastrium of the auto site and had two rudimentary upper limbs and well-developed lower limbs with tactile sensitivity. During dressing changes of the lower limbs of the parasite, who had an ulceration at the knee, the auto site would cry, suggesting probable sensitivity in the lower limbs; however, it did not exhibit spontaneous movement. Notably, the parasitic twin had a well-formed scrotum and penis but lacked testes and an anus, although it could urinate. Routine hematological and laboratory tests produced normal results. An echocardiogram of the auto site showed Tetralogy of Fallot (TOF) along with a patent foramen ovale (PFO).

Further examination using magnetic resonance imaging (MRI) revealed the presence of a solitary pelvic kidney and a few bowel loops within the parasitic twin. Consequently, separation surgery was performed on the 23rd day of life, during which the omphalocele of the auto site was managed using the tanning process.

The surgical procedure involved making a circular incision extending from the lower chest to the umbilicus, providing access to the common peritoneal cavity. The parasitic twin was connected to the sternum of the auto site via a cartilage tract and received its primary vascular supply from the falciform ligament of the auto site. Within the pelvic region of the parasitic twin, we identified a functional single kidney, urinary bladder, testis, and portions of the small and large intestine, although the absence of an anus was noted. The bowel opened onto the sac of the omphalocele with minor prolapse. We primarily closed the abdominal wound of the auto site with slight tension and performed an umbilicoplasty.

Postoperatively, the infant did not require ventilation and had an uncomplicated recovery. Oral feeding began on the third day, and the infant was discharged on the tenth day. Currently, at three months of age, the infant is under the care of a cardiac surgeon for the management of Tetralogy of Fallot. Histological analysis of the resected organs confirmed the presence of both small and large intestines. Additionally, an oval-shaped mass located in the

pelvic cavity corresponded to testicular tissue, alongside a third morphologically distinct left kidney, adrenal gland, ureter, and bladder.

Discussion

Conjoined twins, whether symmetrical or asymmetrical, represent an exceedingly rare. Congenital abnormality. Asymmetrical conjoined twins, often referred to as parasitic or heteropagus twins, are characterized by the parasitic twin's attachment to specific regions of the auto site [1,2]. The incidence of conjoined twins is estimated at 1 in 50,000 to 100,000 births with heteropagus twins being even rarer, occurring at a rate of 0.05 to 0.1 in 100,000 births [3,4]. Epigastric heteropagus twins (EHT) form a unique subgroup of monozygotic monochromic asymmetrical conjoined twins. They typically result from a blastogenesis error, which occurs due to incomplete division of a single zygote after 14 days post-fertilization [5]. Of approximately 27 documented cases, 54% have been male [6]. Consistent with our case Delayed surgery has been reported to increase the chances of successful separation, as it allows ample time for internal anatomical assessment and the coordination of multidisciplinary teams for effective preoperative, intraoperative, and postoperative management [7]. In our case, the parasitic twin exhibited limbs and external genitalia, which aligns with findings in numerous reported cases. The primary blood supply to the parasite was derived from the falciform ligament, consistent with most reported cases [8].

The use of MRI is essential as it provides critical anatomical insights, facilitating optimal separation planning by the surgical team [9]. Emergency separation is warranted when one twin is deceased or near death and endangers the other twin's life, while elective surgery is preferred when the twins are stable, allowing adequate time for fusion evaluation through imaging. Typical anatomical features developed in the parasitic twin include rudimentary limbs, a pelvis, external genitalia, a genitourinary system, and a portion of the bowel.

Omphalocele is the most common associated anomaly, often followed by cardiac anomalies in the auto site [10]. Both of which were present in our case. (Figures 1-3)



Figure 1: The malformation of the patient (A) at birth (B) at the time of surgery at 21 days of life.

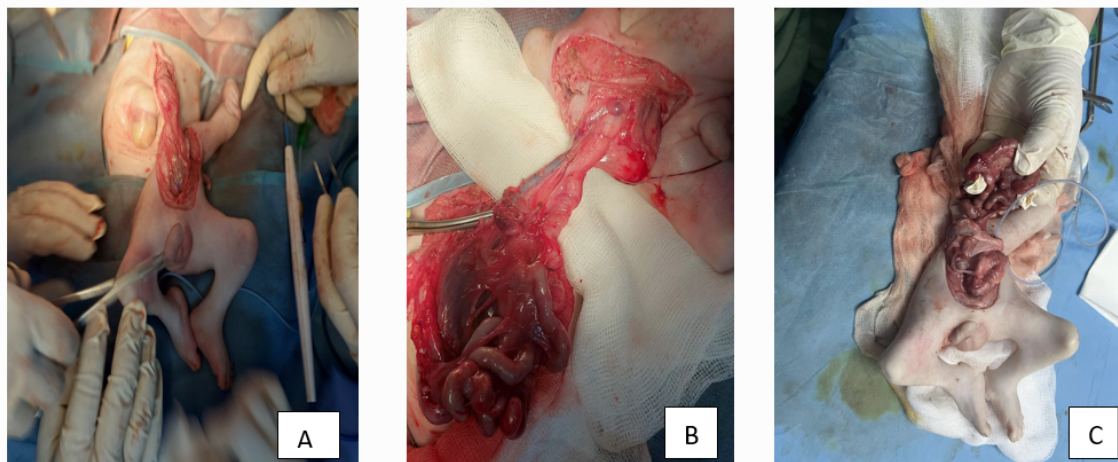


Figure 2: The parasite surgically separated from the auto site; (A) A well-formed scrotum and penis but lacked testes and an anus, although it could urinate; (B) vascular supply from the falciform ligament of the auto site; (C) pelvic region of the parasitic twin, we identified a functional single kidney, urinary bladder, testis, and portions of the small and large intestine.

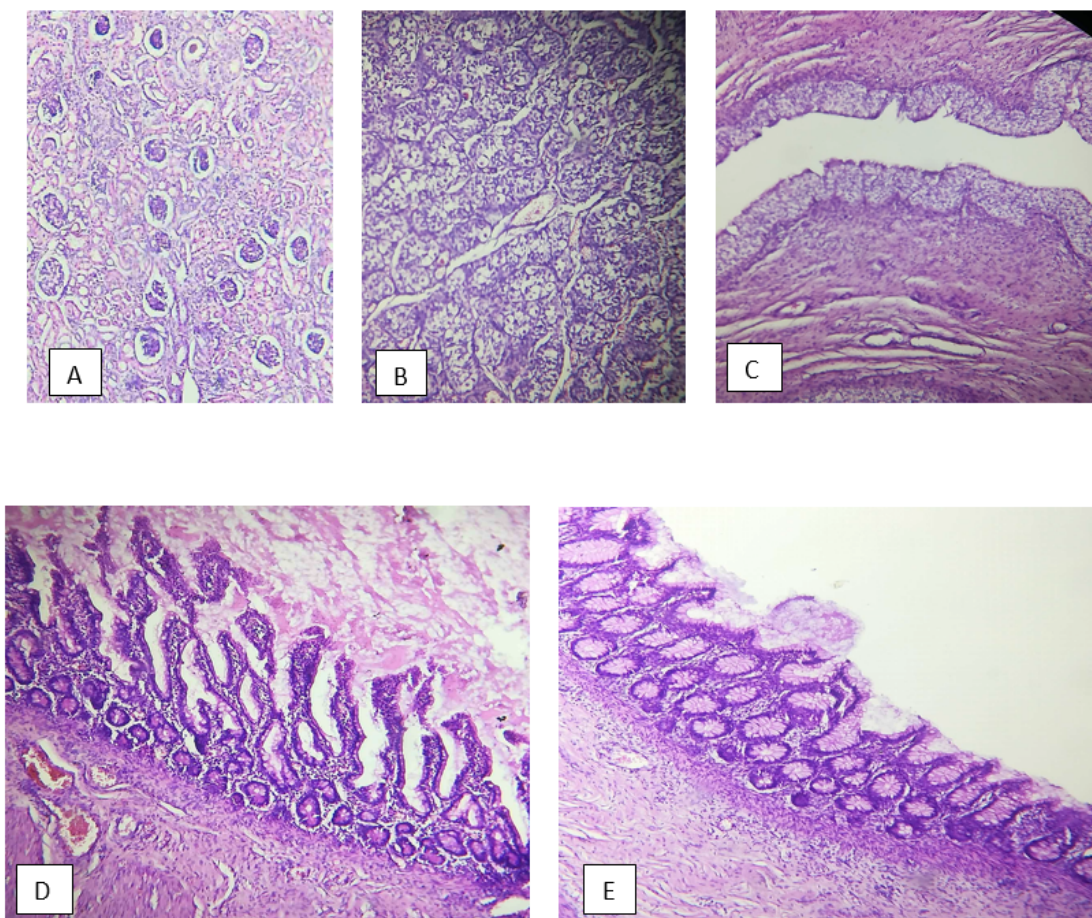


Figure 3: Histological analysis; (A) Renal parenchyma; (B) Fetal-type testicular parenchyma; (C) Bladder wall; (D) Small intestinal wall; (E) Colonic wall.

Conclusion

We report a successful separation of an epigastric heteropagus auto site. Delayed surgery has been shown to enhance the likelihood of a successful separation, allowing sufficient time for precise anatomical delineation and the coordination of multidisciplinary teams for effective preoperative planning.

Consent to Publication statement

Author(s) declared taking informed written consent for the publication of clinical photographs/material (if any used), from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient, however it cannot be guaranteed.

Author Contribution

Author(s) declared to fulfill authorship criteria as devised by ICMJE and approved the final version. Authorship declaration form indicating individual contribution, submitted by the author(s), is available with the editorial office.

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