Pediatric Surgery International (2022) 38:169–181
https://doi.org/10.1007/s00383-021-05006-w

ORIGINAL ARTICLE

A cohort of five cases with asymmetric conjoined twining and literature review

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Accepted: 26 August 2021 / Published online: 31 August 2021
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Abstract

Purpose Asymmetric conjoined twining (ACT) is a form of conjoined twining which is a rare malformation of monochorionic monoamniotic twin pregnancy. Most publications were single case reports. We reported a cohort of five cases with ACT from a single tertiary medical center and reviewed the case reports of ACT over the last decade to enrich the clinical research of this disease and summarized the clinical features of the disease.

Methods We reviewed five cases of ACT admitted in Tianjin Children's Hospital from 17 March, 2008, through 7 March 2017. The cohort was analysed from general information, imaging manifestations, separation surgery, histopathological findings, outcome and follow-up. We searched the English literatures on case reports of ACT over the past decade from the PubMed database and presented details about the clinical characteristics, treatment, and prognosis of all cases.

Results There were four males and one female in our cohort. Among the five cases, two parasites were located in epigastrium, two in rachis, and one in retroperitoneum (fetus in fetu, FIF). All of the parasites were separated successfully by operation in five cases and were confirmed to be ACT by histopathology reports. Four patients made an uneventful recovery except for one case of wound infection. All of them were doing well in follow-up. In the literature review, we found 41 cases of exoparasitic heteropagus twining (EHT) and 63 cases of FIF.

Conclusions ACT is very rare and usually diagnosed by prenatal ultrasonography (US). Computed tomography (CT) and magnetic resonance imaging (MRI) examinations are essential imaging examinations before separation surgery to delineate the anatomical relationship between the autosite and the parasite. In general, the separation surgery of ACT is less complicated and the prognosis is better compared with the symmetric conjoined twining (SCT).

Keywords Asymmetric conjoined twining · Separation surgery · Autosite · Parasite

Introduction

Conjoined twining is a rare form of twin pregnancy, including symmetric conjoined twining (SCT) and asymmetric conjoined twining (ACT). ACT is extremely rare, including exoparasitic heteropagus twining (EHT) and endoparasitic heteropagus twining, which also named fetus in fetu (FIF). The earliest case was proposed by Friedrich Meckel in 1800 [1]. It is usually detected by prenatal ultrasonography (US). Computed tomography (CT) and magnetic resonance imaging (MRI) examinations should be performed before separation surgery to determine the location, extent, organ distribution and fusion of the junction between the autosite and

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the parasite, as well as the distribution of the blood vessels between the twins. Here we reviewed five cases of ACT. Among them, two parasites were located in epigastrium, two in rachis and one in retroperitoneum (FIF). All of them were separated by operation successfully. Besides, a review of literature was presented for the sake of a further understanding of ACT.

Patients and methods

Patients

There were five cases of ACT admitted in Tianjin Children's Hospital from 17 March 2008 to 7 March 2017.

The general information of all patients, such as age, sex, relevant family history, history of gestation, gestational weeks at birth, mode of delivery, method and timing of diagnosis, deformity type, physical examinations were collected. Written informed consents were obtained from parents of the five patients and the study was approved by the ethics committee of Tianjin Children's Hospital (Tianjin, China).

Imaging examinations

It was very important to perform the related imaging examinations before separation surgery to delineate the anatomical relationship between the autosite and the parasite. CT and MRI examinations clarified the visceral and vascular communication between the parasite and the autosite. Echocardiography (UCG) revealed cardiac malformations in autosite. US and UCG were performed in all five cases, CT and MRI in four cases (Case 1, 3, 4 and 5).

Separation surgeries

After defining the anatomical relationship between the autosite and the parasite through physical examinations and necessary imaging examinations, all patients underwent separation surgeries.

Literature review

We searched the English literatures on case reports of ACT over the last decade from the PubMed database and collected the general information, clinical characteristics, treatment, and prognosis of all cases.

Results

Patients' details

The age, sex, relevant family history, history of gestation, gestational weeks at birth, mode of delivery, method and timing of diagnosis, deformity type, physical examinations of the five cases were shown in Table 1. There were four males and one female. Three of them were admitted to the Department of Neonatal Surgery (Case 1, 2 and 5), and two were admitted to the Department of Pediatric Neurosurgery (Case 3 and 4). Among these cases, two parasites were located in epigastrium (Case 1 and 2), two in rachis (Case 3 and 4) and one in retroperitoneum (FIF) (Case 5). Only one case was diagnosed before birth (Case 5). The physical examinations of them were listed in Table 1. The malformations of cases 1, 2, 3 and 4 were shown in Figs. 1a, b, 2a, 3a, b and 4a–c, respectively. The parasites' extremities of the three cases (Case 1, 2 and 4) could not move spontaneously or respond to external stimulation. The scrotum and penis of parasite presented in two cases, one of which showed no automatic micturition (Case 1), and the other could urinate autonomously (Case 2). In case 4, there was an anus and a perineum at the junction of the parasite's limb and the autosite's buttock, but neither urination nor defecation was observed. In case 3, an irregular mass on the back of the autosite could be seen (Fig. 3). The located skin on the left side of the junction was red and dry without exudation. Besides, an abnormal bony processes could be found on the right side of the basement.

Imaging details

The imaging details were shown in Table 2. US and UCG were performed in all five cases, CT and MRI in four cases (Case 1, 3, 4 and 5) (Figs. 1c, d, 3c–e, 4d, e, 5a, b). In case 5, a heterogeneous mass with clear boundary and intact capsule was seen in the right upper abdomen of the child, including multiple long bones and a probable vertebral body formation, suggesting that it might be a FIF.

Intraoperative findings

The details of surgeries, such as time of operation, surgical findings and medical operations, were shown in Table 3. All patients underwent single-stage surgery performed by a multidisciplinary team. The time of separation surgery ranged from 9th day to 6th month of life.

Table 1 Patients' information

Case no.	Age, sex	Relevant family history	History of gestation	Gestational weeks at birth	Mode of delivery	Method and timing of diagnosis	Location	Physical examination
1	12 h, Male	No	G1P1	39^{+4}	Caesarean section	After birth	Epigastrium	Hypoplastic lower extremities, upper extremities, scrotum and penis attached to the epigastrium of the autosite
								An omphalocele of size 3 cm × 3 cm in the autosite
2	–, M	–	–	–	–	–	Omphalopagus	Incomplete lower extremities, upper extremities, scrotum and penis attached to the upper abdomen of the autosite
								A large omphalocele of size 5cm×5cm in the autosite
3	23 days, M	No	G2P2	Full-term (No details)	Vaginal delivery	After birth	Rachis	An irregular mass on the back of the patient, showing the shape of the penis and scrotum in hypospadias
4	6 mon, F	No	G2P2	Full-term	Vaginal delivery	After birth	Rachis	Vegetative limb on the autosite's left gluteal region, with seven toes in the extra foot
				(No details)				An anus and perineum at the junction of the limb and the autosite's buttock
5	1 mon, M	No	G1P1	36^{+1}	Caesarean section	At the 25 weeks of his mother's gestation by prenatal US	Retroperitoneum	No obvious abnormality

Case 2 was an abandoned baby

M male, *F* female, *GnPm* gravida n, para m, *FIF* fetus in fetu

All of the parasites were separated successfully from the autosite (Figs. 1e, 2b, c, 3f, 4f, 5c, d).

Histopathological findings

Histopathological examinations were performed in five cases and all of them were confirmed as ACT. The histopathological details were shown in Table 4.

Outcome and follow-up

Case 1 showed wound infection after operation, and wound healed gradually after regular dressing change. The other four cases made an uneventful recovery without wound dehiscence, urinary and fecal incontinence, or heart failure. The postoperative wounds of cases 1 and 3 were shown in Figs. 1f and 3g, respectively. In our five cases, the shortest follow-up lasted for 3 months and the longest

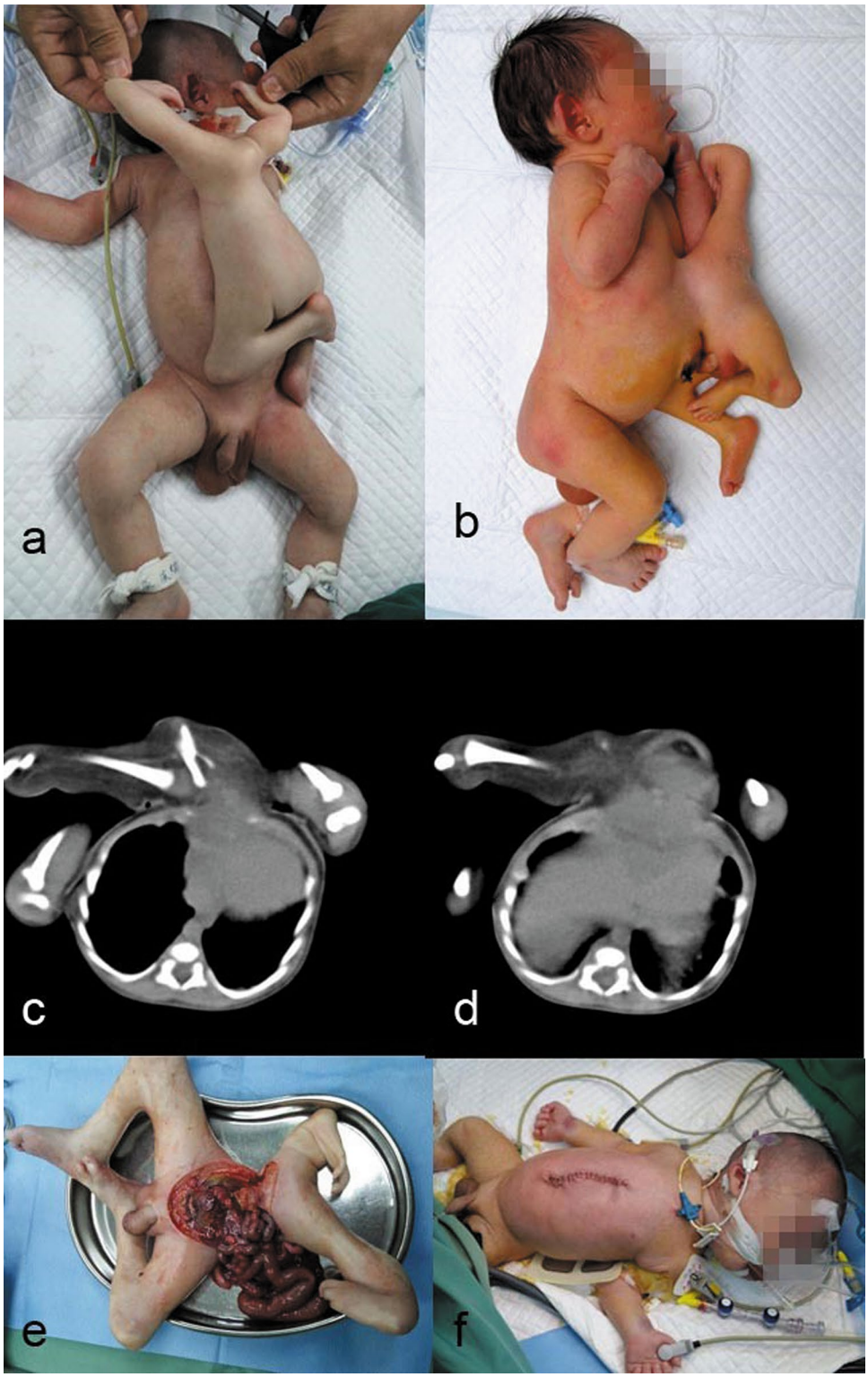

Fig. 1 Picture of case 1. **a**, **b** The malformations of the patient; **c**, **d** CT images of the patient; **e** the parasite surgically separated from the autosite; **f** the postoperative wound of the autosite

lasted for 2.5 years. All of the five patients were doing well, enjoying a normal quality of life. The outcome and follow-up were shown in Table 5.

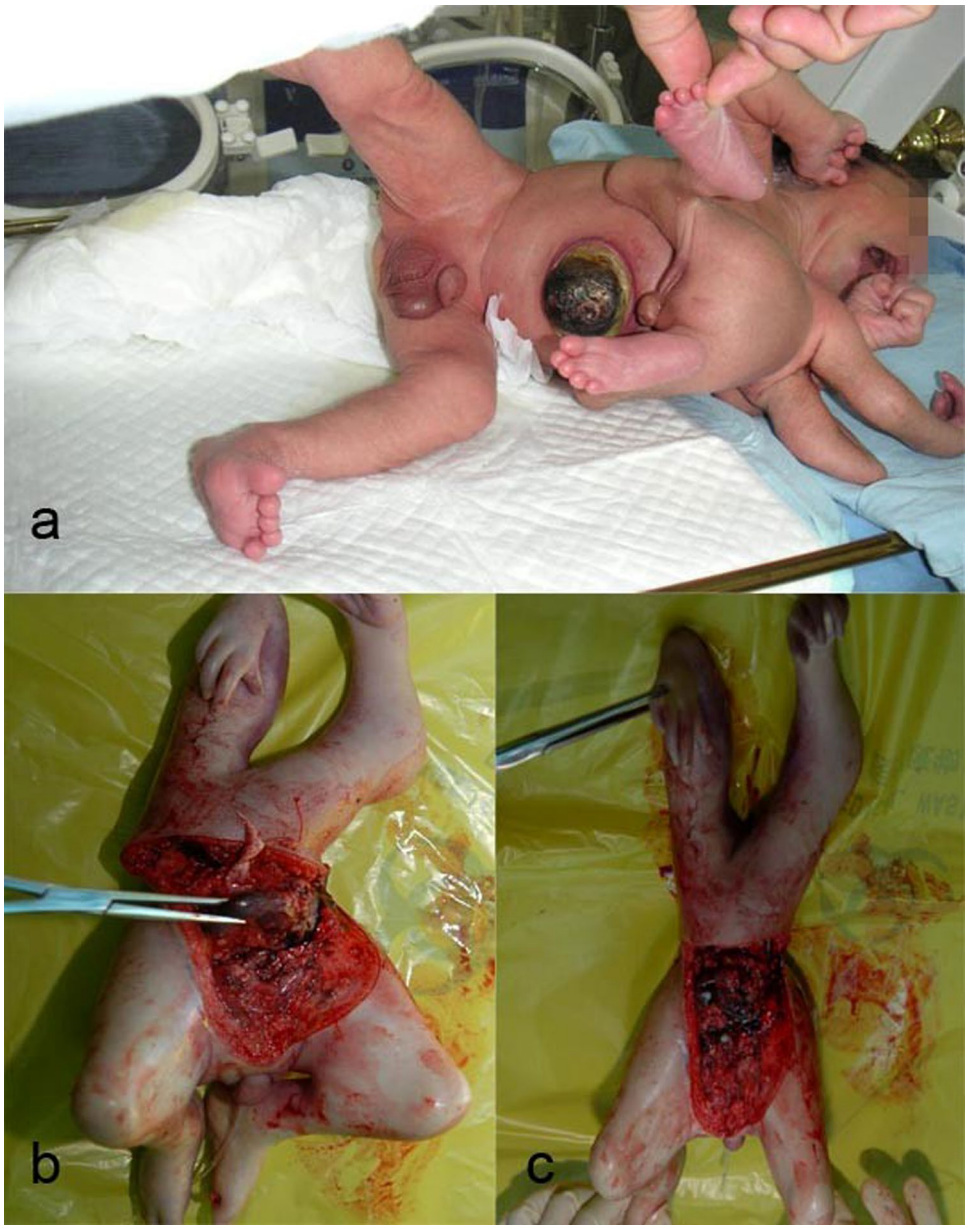

Fig. 2 Picture of case 2. **a** The malformations of the patient; **b**, **c** the parasite surgically separated from the autosite

Literature review

The details about the general information, clinical characteristics, treatment, and prognosis of all cases were presented in Supplementary Table A (EHT) [2–38] and Supplementary Table B (FIF) [39–88].

In the literature review, we found 41 cases of EHT and 63 cases of FIF. In EHT, 17 were females, 24 were males. In FIF, 32 were females, 28 were males and 3 were not mentioned. In terms of parasitic sites, the most common site of EHT was abdomen, especially repigastrium, followed by rachis, while the most common site of FIF was retroperitoneum. When it came to treatment (mentioned in the literature), most cases received surgical treatment and most of them had a good prognosis, whether EHT or FIF. Specifically, in the 41 cases of EHT, 33 cases underwent separation surgery, of which 25 cases were fine after operation, 4 cases had postoperative complications such as wound infection and wound dehiscence, 2 cases left congenital heart disease and 2 cases died after operation; 7 cases did not receive surgical treatment. The treatment of 1 cases was not mentioned in EHT. In the 63 cases of FIF, 48 cases underwent separation surgery, of which 42 cases were fine after operation, 1case left epilepsy and delayed psychomotor development, 1 case left urinary retention and 4 cases died after operation; 4 cases did not receive surgical treatment. The treatments of 11 cases were not mentioned in FIF.

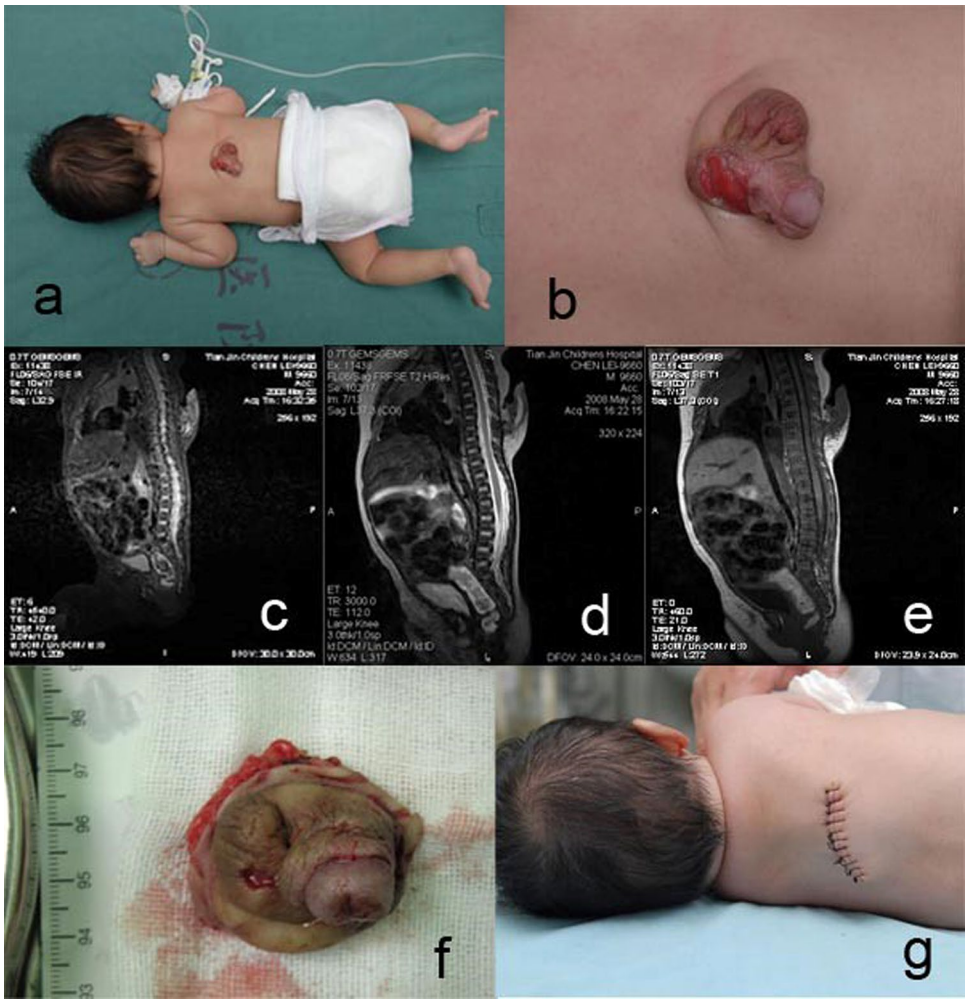

Fig. 3 Picture of case 3. **a**, **b** the malformation of the patient; **c**–**e** MRI images of the patient; **f** the parasite surgically separated from the autosite; g. the postoperative wound of the autosite

Discussion

ACT, also known as heteropagus conjoined twining, incomplete conjoined twining, parasitic twining, is a rare congenital developmental malformation. ACT means a relatively mature fetus parasitizing a severely developmentally deficient fetus in any portion of the body. We reviewed the case reports of EHT and FIF over the past decade (Supplementary Table A and Supplementary Table B), except for cases of termination before 20 weeks of pregnancy. The incidence is 0.5–1 in 1000000 births [89]. There is a male predominance in ACT as opposed to female predominance in SCT [17, 90]. In literature review, there were 24 males and 17 females in EHT while 28 males and 32 females in FIF. In our five cases, four cases were male and one was female. Case 3 had been published previously as an 11-day-old infant with an accessory penis and scrotum on the posterior thoracic region [91].

The mechanism of conjoined twining has not been clear yet. Two possible theories have been proposed, namely the fission theory and the fusion theory. Both theories indicate that ACT occurs around two weeks after fertilization. The fission theory claims that a single zygote cannot divide completely [92], while the fusion theory proposes that the fusion of two embryos occurs prior to implantation [93, 94]. At present, most people support the fission theory. In the literature, DNA analysis of one case showed that the ACT was dizygotic, confirming the possibility of fusion theory [95]. Another ischemic theory has also been proposed, in which the parasite results from ischemia and resorption of the autosite [92, 96].

ACT is classified into EHT and FIF. Spencer et al. suggested that FIF should have at least one of the following characteristics: (1) a completely encapsulated mass, (2) partially or completely covered by normal skin, (3) one or more clearly identifiable anatomical structures, (4) attached to the autosite by only a few relatively large blood vessels and (5) either close to the attachment site of conjoined twins, or connected with the neural tube or gastrointestinal tract [90]. In our case 5, parasite originated from the retroperitoneum of the autosite was wrapped in a complete capsule with both lower extremities and feet. Imaging examinations showed a heterogeneous mass with multiple long bones and

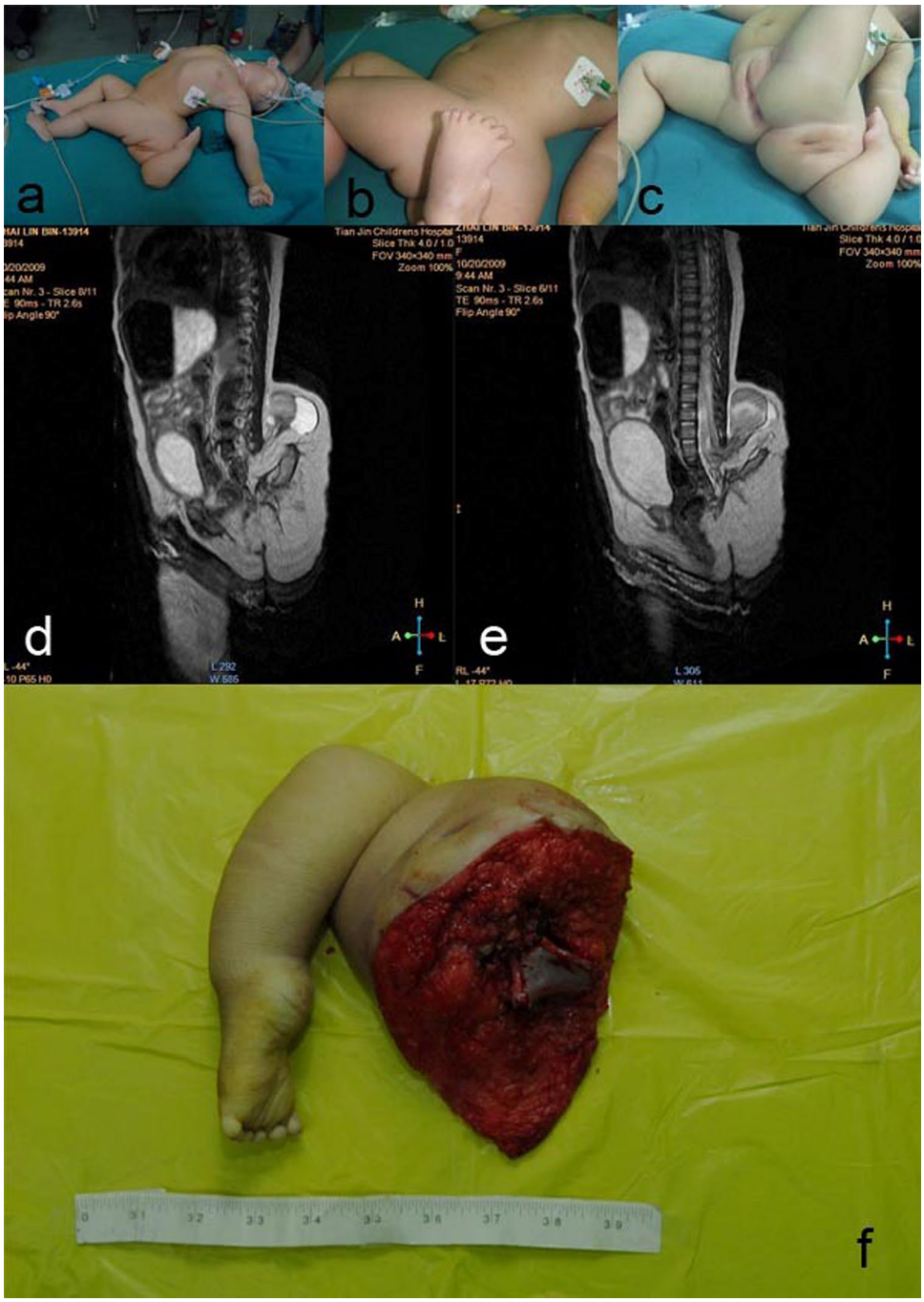

Fig. 4 Picture of case 4. **a**–**c** The malformation of the patient; **d**, **e** MRI images of the patient; **f** the parasite surgically separated from the autosite

a probable vertebral body formation. The histopathological examinations demonstrated cartilage and bone-like tissue which were suggestive of rudimentary vertebrae and long bones of lower limbs. As a result, the final diagnosis of FIF was made. The common parasitic sites of EHT include epigastrium (omphalopagus), rachis (rachipagus), thorax (thoracopagus), abdomen and ischium (ischiopagus). A few parasites can be located in extracranial region (cephalopagus) and perineum as well. However, there are also reports of atypical EHT, such as atypical ischiopagus [97]. As for FIF, the parasite can be located in anywhere of the autosite. Retroperitoneum, abdominal cavity and thoracic cavity are more usual. In the literature review, the most common site of EHT was abdomen, especially repigastrium, followed by rachis, while the most common site of FIF was retroperitoneum. In our five cases, two parasites were located in epigastrium, two in rachis and one in retroperitoneum (FIF).

Clinical features are associated with the type of ACT. The most common feature of EHT is supranumerary limbs, especially in thoracopagus and omphalopagus. The most

Table 2 Imaging details of the patients

Case no.	US	CT and MRI	UCG
1	Bilateral testicular hydrocele	The parasite communicated with the autosite's abdominal cavity at the diaphragmatic level, and part of the autosite's liver protruded outward into the parasite. Defects could be seen in the soft tissue, part of the sternum and costal cartilage of the anterior chest wall. A branch from the brachiocephalic trunk of the autosite entered the parasite	ASD, VSD PDA, PHT
2	N	–	ASD, PDA
3	N	Subcutaneous soft tissue masses in the dorsal region, which did not communicate with the structure of the spinal canal	N
4	N	Lumbosacral spinal dysraphism, sacral dysplasia, tethered cord, myelomeningocele, expansion of central spinal canal from ninth thoracic to fourth lumbar vertebral level, lipoma of filum terminale	ASD
5	A heterogeneous mixed echo mass in the right upper abdomen measuring about 6.7 cm × 4.8 cm × 5.7 cm with calcifications and osseous elements resembling limb bones and vertebral bodies	A heterogeneous mass with multiple long bones and a probable vertebral body formation The surrounding tissues, such as liver, common bile duct, portal vein, pancreas, intestinal tube and inferior vena cava, were squeezed by the mass and had some displacement	ASD

ASD atrial septal defect, *VSD* ventricular septal defect, *PDA* patent ductus arteriosus, *PHT* pulmonary hypertension, *N* nomal

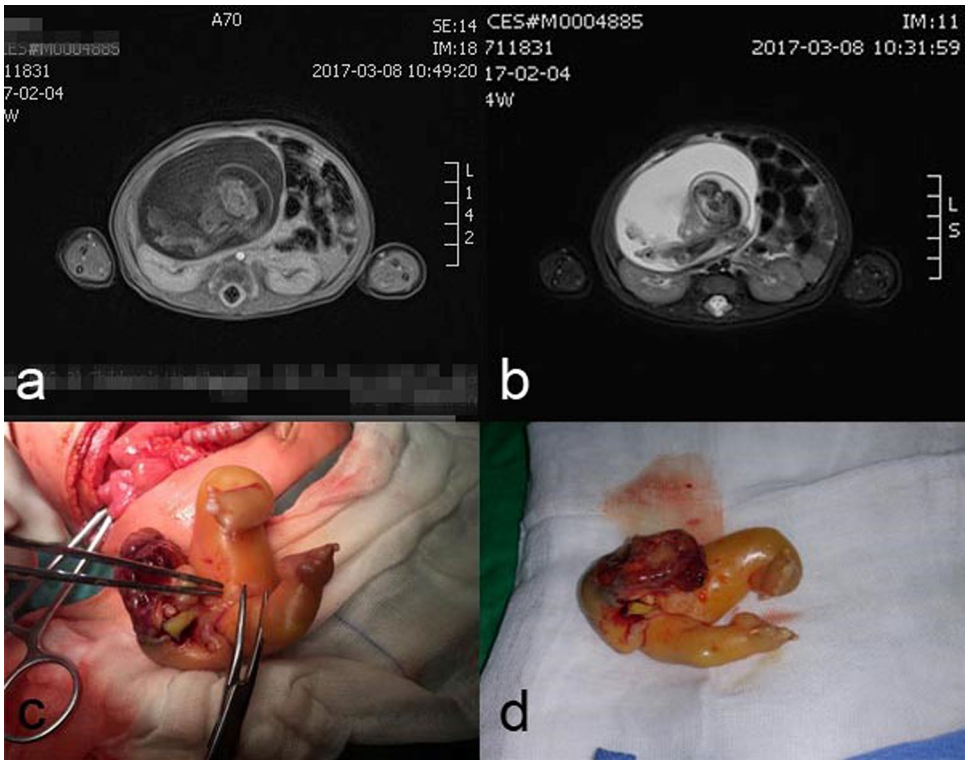

Fig. 5 Picture of case 5. **a**, **b** MRI images of the patient; **c**, **d** the parasite surgically separated from the autosite

limbs of parasites could not move spontaneously or respond to external stimulus. While two different cases have been reported in the literature. In one case, a pair of asymmetric conjoined twins had a common dual nerve supply, and stimulation of any part of the body resulted in movement of all limbs [98]. In another case, the extremities of the parasite exhibited spontaneous movement in the toes and apparent sensation [30]. In our five cases, three cases had

Table 3 The operation details of the patients

Case no.	Time of operation	Surgical findings	Medical operation
1	9th day	Adhesion between the intestine of the parasite and the liver of the autosite	Adhesiolysis
			Ligated the nourishing vessel
		The parasite's nourishing vessel originated from the brachiocephalic trunk of the autosite	Omphalocele repair
			Umbilical ureterectomy
			Abdominal wall plasty
2	4th month	Adhesion between the liver of the autosite and the peritoneum of the parasite	Catheterization (autosite and parasite)
			Adhesiolysis
		A larger liver	Ligated the nutrient vessel from the splenic artery of the autosite
		A stunted intestinal tube about 50 cm-long filled with foetal faeces terminated at the presacral of the parasite	
		A kidney of the parasite (2 cm × 1.5 cm) with a ureter attached to the bladder	
3	28th day	The parasite was connected with the vertebral lamina	Excised completely
4	6th month	The terminal spinal cord was degenerated and adhered to the lipoma	Disconnected the nourishing vessel and the nerves
			Dissociated the hip bone of the parasite
			Separated the adhesion between dura mater and spinal cord
			Resected most of the lipoma
			Artificial dural repair
5	34th day	Parasite derived from the retroperitoneum of the autosite	Dissociated the adhesive tissue
		A large amount of pale yellow amniotic-like fluid in the capsule, foetus-like masses with both lower extremities and feet	Completely removed the tumor and its capsule

Table 4 The histopathology findings of the patients

Case no.	Macropathology	Under the microscope
1	Maldeveloped trunk, hypoplastic extremities, intestines and male external genitalia. The scrotum was empty inside. The intestine length of the parasite was 90 cm, and a cyst of size 2 cm × 2 cm × 1.5 cm was observed below the intestinal tube	Smooth muscle cells and transitional epithelium, considered as a underdeveloped bladder. No skull, chest, spine or other organs
2	Lower extremities and one upper extremitie, kidney, ureter, few bowel loops	Some brain tissues and immature nerve tissues on the cephalic side. Bone, cartilage, and a little subcutaneous fat; one side of the fibrous capsule wall was covered by cuboidal epithelium, and the other side and the local capsule wall had adenoid structure
3	The shape of the penis and scrotum in hypospadias	Cavernous tissue, adipose tissue, arteries and veins, some muscle tissue and glands
4	The malformed limb	Muscle tissue and a few nerve tissue
5	Limbs, buttocks and some other organs	Kidney tissue, part of bladder and alimentary canal tissue; sectioning demonstrated cartilage and bone-like tissue suggestive of rudimentary vertebrae and long bones of lower limbs

supranumerary limbs, and none of them moved spontaneously or responded to external stimulus. Other manifestations such as abdominal wall defect and abdominal visceral fusion are usual in omphalopagus, while spinal defects are usually found in rachipagus. The clinical features of FIF are related to the location of parasite. For example, parasite located in retroperitoneum can be characterized by abdominal distension and low fever, while that in mouth causes oropharynx developmental deformities.

ACT with other malformations are less common than SCT, especially in FIF. The most common malformation is omphalocele, followed by cardiac anomalies [32]. Other

Table 5 The outcome and follow-up of the patients

Case no.	Discharge time (after the operation)	Postoperative complications	Follow-up
1	16th day	Wound infection	2.5 years
2	23th day	No	3 months
3	13th day	No	2 years
4	18th day	No	2 years
5	14th day	No	1 year

malformations are rare, such as genitourinary system malformations and digestive system malformations. In our five cases, two cases had omphalocele. All of the patients underwent echocardiography and all of them found varying degrees of cardiac malformations. In addition, the soft tissue of anterior chest wall, sternum and costal cartilage defects were found in case 1. In the meantime, bilateral testicular hydrocele was also found. Case 4 showed lumbosacral spinal dysraphism, sacral dysplasia, tethered cord, myelomeningocele, expansion of central spinal canal from 9th thoracic to 4th lumbar vertebral level and lipoma of filum terminale.

ACT is usually identified by prenatal US. Sonographic findings in conjoined twins include inseparable fetal bodies and skin contours, no change in the relative positions of the fetuses and shared organs [99]. The earliest diagnosis was reported to be made at the 9th week of pregnancy [100]. But in our five cases, three mothers did not undergo US prenatally, so the malformations were not found before birth. Case 2 was an abandoned baby and the details could not be obtained. Only one patient was diagnosed by prenatal US. Prenatal three-dimensional ultrasound examination can provide a clearer image, which contributes to diagnosis and prenatal counseling. It was reported that two cases of ACT were diagnosed by prenatal three-dimensional ultrasound [13, 101]. It is very important to perform CT and MRI examinations before operation. These examinations are helpful to understand the connection site, organ distribution and fusion of twins, and to judge the blood supply of parasite. With the development of three-dimensional CT reconstruction technique, it is possible to present the structure of twins in three dimensions. It helps us to understand the twins' position relationship more intuitively, which is crucial for the design of operation schemes and the evaluation of prognosis.

Whether early termination of pregnancy is required, as well as the timing of termination need to be determined individually, according to the types and forms of ACT. Surgery can be performed during 1st week of patient's life to the age of 1 year [30]. That's because the patient's vital signs are relatively stable during this period. In addition, if the surgical age is too late, the growth and development of the autosite might be affected. In our five cases, the time of separation surgeries ranged from 9th day to 6th month of life. Cases 3, 4 and 5 were older than one week at admission, so the separation surgeries were performed after preoperative preparation. Case 1 was admitted to the hospital 12 h after birth. The operation was performed nine days later when the vital signs were stable and the preoperative preparation was ready. Case 2 was an abandoned infant whose timing of surgery was also influenced by several other factors. Compared with SCT, the operation procedure of ACT is less complicated because of the less extensive vascular and visceral connections. The surgical purpose of ACT is to remove the redundant parasite, while that of SCT is to separate the two surviving individuals. What's more, some ethical issues will arise when only one fetus can be preserved during the operation of SCT [102].

The prognosis of ACT is related to the location of connection and the degree of organ fusion between the parasite and the autosite, as well as other malformations. The presence and severity of cardiac malformations are the main determinant factors of the prognosis [15]. In general, the prognosis of ACT is better than that of SCT. In our study, except one case of wound infection, the remaining four patients recovered smoothly. All of them were doing well in follow-up. In the literature review, the prognosis (mentioned in the literature) of most cases were good, except for five cases of preoperative death, one case of intraoperative death, five cases of postoperative death, one case of early termination of pregnancy and two cases of death in utero. In the EHT, one case died in utero because of severe congenital cardiac anomaly. The other two cases with multiple cardiac anomalies died of cardiac arrest and acute respiratory disorder before surgery, respectively. One case died before doing any investigations because of septicemia. Two cases died postoperatively, one of them was due to respiratory distress and cardiac failure, and the other was not able to determine the exact cause of death. In FIF, one case died in utero at 37 weeks and the two cases who died before operation were all cases of parasite located in intracalvarium. One case of FIF died for cardiac arrest during the operation. Among the three cases of FIF who died after surgery, two cases died for serious postoperative complications, one case died for complex congenital heart disease.

Conclusion

ACT is very rare and usually diagnosed by prenatal US. CT and MRI examinations are essential before separation operation to delineate the anatomical relationship between the autosite and the parasite. In most cases, the surgical operation of asymmetric twins is less complicated and the prognosis is better compared with the SCT.

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1007/s00383-021-05006-w.

Acknowledgements Not applicable.

Author contributions All authors contributed to the study conception and design. BH, XZ were responsible to design and conduct the study. Material preparation, data collection and analysis were performed by CG, LP, JC and YF. The first draft of the manuscript was written by XZ. CC was in charge of critical revisions and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Funding This work was supported by the National Natural Science Foundation of China (grant number 81771589) and the Program of Tianjin Science and Technology Plan (grant number 18ZXDBSY00170).

Declarations

Conflict of interest The authors have no financial or proprietary interests in any material discussed in this article.

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