

Surgical treatment of a rare form of limbless paracephalic parasitic twin with an omphalocele

Gazi AYDIN, Selahattin TOKTAŞ, Ayhan ARGUN, Adem ARSLAN

From the departments of pediatric surgery and pathology, Dicle University Hospital, Diyarbakır, Turkey

Summary

The authors report a rare case of asymmetric double monstrosity. The boy had an omphalocele and a parasite consisting of a head, neck and bulk of tissues attached to the epigastrium. At the operation the headed limbless parasite was removed

successfully and the omphalocele was repaired. The postoperative course was uneventful.

Index words: Limbless paracephalic parasitic twin, headed limbless parasite asymmetric double monstrosity

Introduction

Parasitic twins are rare in medical literature and few cases have been surgically treated since 1968, mostly caudal duplications (2,3,4,5). Only two of these cases had epigastric insertion with a set of accessory lower attached to the sternum. We report the case of an infant born with a hemiacardiac, headed limbless parasite.

Case Report

The male infant was born in the 38th week of gestation following an uneventful pregnancy. The labor was spontaneous and delivery uncomplicated. The mother had taken oral contraceptives before pregnancy. The baby was the eighth living child of a 38 year old mother and father. There was no history of familial congenital malformations and maternal illness. The baby was admitted to the hospital on the second day of birth and weighed 3850 gm. He had passed meconium on the first day.

A parasitic twin was attached to the epigastrium which consisted of a head, neck and a bulk of tissues (Fig. 1 and 2). It was motionless but easily movable. He was cyanotic, oedematous and hypothermic. There was a pulsation on the left side of the neck with a rate of 94 per second. The neck was anteriorly continuous with a skin including two nipples. Both auriculas were not present in a otherwise normal looking face and a hydrocephal-



Fig. 1-2. Appearance of the newborn showing the headed parasite and the omphalocele before surgical intervention

ic head. Both lips were present with median mandibular cleft but the oral cavity was hypoplastic. Both palpebra were present but the parasite was anoptalmic. The x-ray examination showed a normal bony structure of skull with a hypoplastic columna vertebralis. 20 piece of vertebra were count-

Address: Dr. Gazi Aydın, Dicle Üniversitesi Tıp Fakültesi Çocuk Cerrahisi Anabilim Dalı, Diyarbakır, Türkiye

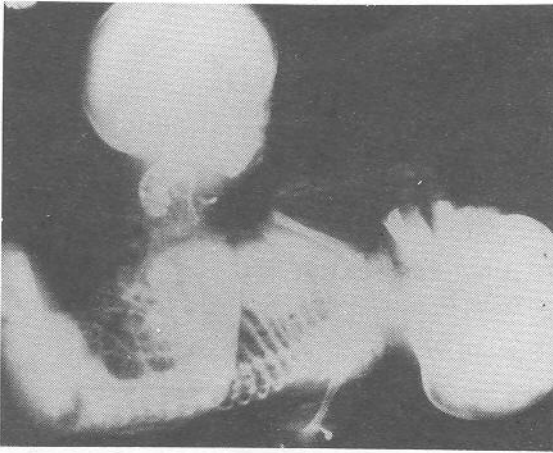


Fig. 3. Total-body x-ray showing skeletal segments.

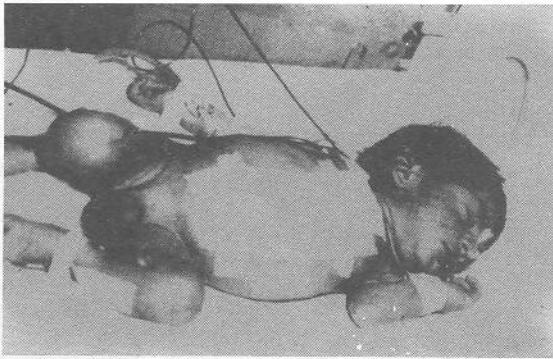


Fig. 4. Appearance of the infant after surgical correction.

ed. (Figure 3).

The boy was otherwise normal except for an omphalocele at 6 cm. diameter.

Surgical Findings

After a thorough clinical evaluation the parasitic twin was separated from the baby under general anesthesia on the second day of his admission. A circumferential incision was made in the skin 3 cm above the junction of the nutrient vessels. The parasite had a single artery from an umbilical artery of the normal twin leading to the parasite heart and leaving it by single vein of the host. The parasite had xypoid like cartilage attached to the xphoid of the baby. The artery and the vein connecting the parasite to the host were ligated and transected. The head was removed and the defect in the skin was sutured vertically without tension. Omphalocele was primarily closed by approximating the rectus muscles (Figure 4).

There was a primitive heart in the neck tissues. The heart continued to beat for about 1-2 hours after separation of the parasite. The postoperative period was uncomplicated and the baby left the hospital on the 7th postoperative day.

Histopathological Findings

Total weight of parasite was about 950 gm. The circumference of the head was 37 cm and the brain was only 3 mm thickness with the rest of the cranial contents consisting of water. Anophthalmia, severely hypoplastic external ears, cavum nasi with agenesis of nasopharynx and oropharynx was shown.

The heart was at the top of a simple cardiac tube arrested embryologically in the 4th week. A segment of intestine 7 cm in length and a piece of pancreatic tissue were seen dorsal to the heart. All these findings were in accordance with the arrest development between 3-7 weeks of embryologic life (6).

Discussion

Our patient can be classified as a case of parasitic limbless paracephalic twin. It is very rare malformation and as far as we know that there is no previously reported case of epigastric insertion of parasite consisting a head neck and fairly developed vertebral columns. It has been said that the insertion of the parasite is usually found in the hypogastric or umbilical regions (3). There are two cases of insertion above umbilicus in the literature with a set of accessory lower limbs. They were mostly associated with urological and intestinal malformations, omphalocele, cardiac anomalies and oesophageal atresia (1,2). Our case was limbless and had an omphalocele but demonstrated no other malformations.

References

1. Nasta R, Scibilia G, Corrao A, Lacono M: Surgical treatment of a asymmetric double monstrosity with oesophageal atresia, omphalocele and interventricular defect. *J Pediatr Surg* 21: 60-62, 1986.
2. Olcay I, Zorludemir U, Kıvanç F: Asymmetric double monstrosity. *Z Kinderchir* 44: 174-175, 1989.
3. Rowe ML, Rawitch MM, Ranninger K: Operative correction of caudal duplication (dipygus). *J Pediatr Surg* 63:840-48, 1968.
4. Simpson JS, Gibson DA, Cook GT: Surgical correction of caudal duplication (dipygus). *J Pediatr Surg* 8:935-38, 1973.
5. Spitz L, Rickwood AMK, Pilling D: Dipygus (caudal duplication). *J Pediatr Surg* 14: 557-60, 1979.
6. Willis RA: Malformations; Their mode of genesis illustrated by their gross forms in: *The borderland of embryology and pathology*, London Butterworths, chapter 4, pp132-180, 1962.