

Case Report

Parasitic Limb Attached to the Back: A Rare Case of Rachipagus Parasitic Conjoined Twinning

Danfulani Mohammed¹, Gele I. Haruna¹, Ma'aji M. Sadiu¹, Saidu A. Sule¹,
Muhammad A. Musa²

¹Department of Radiology, Faculty of Clinical Sciences,

²Department of Anatomy, Faculty of Basic Medical Sciences,
College of Health Sciences, Usmanu Danfodiyo University, Sokoto, Nigeria.

Corresponding Author: Muhammad A. Musa

ABSTRACT

Parasite rachipagus twinning-one of the rarest of the conjoined twins. We present a 3 weeks old male neonate who presented at our facility with history of an accessory limb attached to the back on the lower part of the neck since birth. No attachment in other parts of the body and no history of motor or sphincteric deficit. This case was surgically managed with good prognosis presented.

Keywords: parasite rachipagus, neonate, accessory limb

INTRODUCTION

Conjoined twins is a rare clinical entity with an estimated incidence varying from 1 in 50 000 to 1 in 100 000 live births. It results from monozygotic twinning. [1] Heteropagus or parasitic twinning refers to a rare type of conjoined twinning in which an incomplete small parasitic twin is attached to a normal host on whom it is dependant. The normal host is known as the autosite. This rare form accounts for 5% of conjoined twins. The site of attachment of the parasite to the autosite varies. [2] In the spinal form of parasitic twin (rachipagus) an ectopic or accessory limb is attached to the back of a normal baby or only a mass of viable tissue may remain. [3]

Rachipagus is an extremely rare pathological entity. The first case was reported by Desonchamps in 1851. [4] Literature only indicates few reports of cases in Africa. [4,5] A rare case of parasitic rachipagus at the cervico-thoracic level is here presented.

CASE REPORT

We present a 3 weeks old male neonate who presented to the Paediatric Department of Usmanu Danfodiyo University Teaching Hospital with history of an accessory limb attached to the back on the lower part of the neck since birth (Figure 1). No attachment in other parts of the body. No history of motor or sphincteric deficit. No history of fever or convulsion. He is the first child of the parent in a monogamous setting. He was delivered through unsupervised spontaneous vaginal delivery at home at term. The mother was 20 years old. Although the mother did not receive antenatal care in a hospital, the antenatal period was said to be uneventful. There was family history of twins on the mother's side; however there was no family history of congenital anomaly in the past.

On examination the child was in general good condition. He weighed 4.1kg, was afebrile, not pale and anicteric. There was a parasitic full limb (upper) attached to

the back at the lower cervical region in the midline. The limb had an arm, forearm and hand. Seven fingers were noted on the hand with syndactyly of the second and third digits. The parasitic limb did not show spontaneous movement but was sensitive to pain. There was soft tissue bulkiness at the site of attachment of the parasite to the autosite. No meningocele was seen. The remaining systemic examinations were essentially normal. Baseline laboratory investigations were normal. A clinical assessment of parasitic limb was made and he was referred to Radiology Department for radiography of the parasitic limb, neck and chest.



FIGURE 1: Photograph of the baby showing a parasitic hand attached to the back (arrow). Note the polydactyly of the fingers as well.

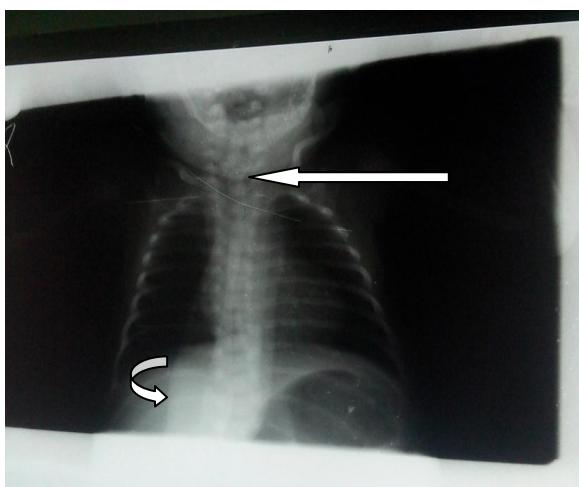


FIGURE 2: Frontal chest radiograph showing a soft tissue shadow projected over the chest and upper abdomen centrally due to a parasitic limb posteriorly (curved arrow). The normal upper limbs are also demonstrated. Note the spina bifida at C7 and T1 levels (straight arrow).

The radiographs showed the parasitic limb in the midline at the back attached to the posterior arches of C7 and T1 vertebrae through a bony girdle (Figures 2 and 3). The parasitic limb consisted of humerus, radius and ulnar bones. There were seven digits on the hand consistent with polydactyly (Figure 4). There was also syndactyly of the second and third digits (Figure 3). Soft tissue fullness was noted over the site of attachment of the limb to the host. Spina bifida occulta was demonstrated at C7 and T1 levels on the autosite (Figure 2). Transfontanelle and abdominal ultrasound revealed normal findings. Computed tomography (CT) and Magnetic resonance imaging were not done as the parents could not afford the costs. An assessment of a parasitic limb attached to the spine (parasite rachipagus) was made. He had total excision of the parasitic limb. No spinal involvement of the autosite was found at surgery. The baby was discharged one week after surgery. The follow up visits was uneventful.



FIGURE 3: Radiograph of the parasitic limb showing humerus, radius, ulna and polydactyly of the digits.

DISCUSSION

Parasitic or heteropagus conjoined twins are exceedingly rare. It is a type of conjoined twinning in which an incomplete small parasitic twin is attached to a normal host on which it is dependant. The normal host is the autosite. [2,6] The parasite follows the disappearance of one of the embryos with the survival of certain additional

structures attached to the normal embryo near or at the junction area. [4] The mechanism that leads to fusion of the twins begins very early in embryonic life at day 6 post fertilization (blastogenesis). [4,6] In conjoined twinning the site of the fusion can be at the chest (thoracopagus), umbilicus (omphalopagus), rump (pyopagus), hip (ischiopagus), cranium (craniopagus), side (parapagus), head (cephalopagus) or spine (rachipagus). The thoracopagus is the most common form while the rachipagus is among the rarest form of conjoined twinning. [1] This case is a parasitic rachipagus attached to the spine at the level of C7/T1. [1]

Symmetrical conjoined twins are monozygotic, monoamniotic and monochorionic. They are always of the same sex with male: female ratio of 1:3. The sex difference is less pronounced in heteropagus (asymmetrical) twins with a reported female preponderance of 51% to 54% males. [1,7] The autosite in this reported case was of male gender.

There have been reports of associated congenital anomalies in conjoined twins such as anencephaly, cleft lip, lumbosacral meningocele, cardiac, gastrointestinal and urogenital anomalies. Rachipagus parasitic twinning is usually associated with vertebral and central nervous system abnormalities at or near the site of union. [2,6] The autosite in this case showed spina bifida occulta at C7 and T1.

The diagnosis of parasitic twins can be made during the antenatal period by ultrasonography. Antenatal diagnosis plays a role in the identification of the size and location of the parasite so as to know whether termination of the pregnancy can be considered or when the pregnancy is allowed to be carried to term to plan for the mode of delivery. [1,8] In this case the mother did not have antenatal ultrasound and the baby was delivered par vaginam at home. During the post natal period, the evaluation and diagnosis can be made by imaging modalities such as plain radiography, ultrasonography (USS),

computed tomography (CT) or magnetic resonance imaging.

Radiography reveals the bony and soft tissues of both the parasite and the host and evaluates for other congenital anomaly. It also helps in evaluation of the site of attachment of the parasite to the host. [4] In rachipagus, the union is most often at the posterior arches of the spine⁴ as demonstrated in this case (Figure 1). Ultrasonography (transabdominal, transfontanelle and echocardiography) identifies other congenital anomalies such as central nervous system, cardiovascular gastrointestinal tract and urogenital anomalies. There were no such anomalies demonstrated in the index patient on ultrasound. CT and MRI define extent of parasitic organs that may reside within the autosite. [4]

In heteropagus twinning, any structure can be found in the parasite. Bones of the limbs are frequently present as in the index case (Figure 3). More rarely heart and neural tube structures can be found in the parasite. [2]

The differential diagnoses of this condition are teratoma, and fetus in fetus. [4,9] These can be easily differentiated from rachipagus parasite based on its already described features in addition to the fact that the limbs are most frequently found in parasite rachipagus than in teratoma or in fetus in fetus. [4,9]

The treatment of conjoined twinning is separation surgery. The outcome is more favorable with the asymmetric (parasitic) than the symmetric conjoined twins because of less vascular and visceral connection in the parasitic twin. [1] The autosite, in the absence of a major cardiac defect usually has a good prognosis following a successful separation surgery [9] as in the case here presented.

CONCLUSION

Parasitic rachipagus twinning is among the rarest of the conjoined twins. The prognosis is usually good on the autosite in the absence of associated congenital

anomalies. A rare case of rachipagus parasitic conjoined twinning that was surgically managed with good prognosis was presented.

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