

Parasitic Rachipagus Conjoined Twins: Surgical Management and Follow-up of a Case in Kenya

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Summary

We present the case, surgical management and follow-up, of a parasitic rachipagus conjoined twin born to a 23 year old lady. The parasitic twin had hypoplastic upper and lower limbs attached to the dorsal thoracic and lumbar region respectively with poorly developed external genitalia. At surgery, a neurovascular bundle was found attached to the parasite upper limb. The parasite was successfully

excised. Subsequent follow up of the child has revealed a boy who despite the weakness of his left lower limb is able to participate in all activities of boys his age. Rachipagus conjoined twins are uncommon and literature only indicates a single report of two cases in Africa. Our case report adds to the number of cases of this condition that have been reported in Africa.

Key Words: Parasitic rachipagus, Conjoined twins

Introduction

Rachipagus, the rarest of all the conjoined twins, is thought to be formed by union in the midportion of the neural tubes of 2 dorsally oriented early embryos, the death of one (perhaps originally defective) embryo resulting in a parasitic twin (1). The first description of such a case was by Jones and Larkin in 1886 (2). Spencer analyzed 1200 conjoined twins and found 1 typical case of 2 complete infants and 20 dorsal parasitic twins (3). We present another example of this rare entity, a parasitic twin with the upper limb attached in the thoracic region and lower limb attached in the lumbar region of the autosite.

Case report

A 23 year old lady with 2 previous cesarean section deliveries and an unsupervised term pregnancy delivered a 3Kg male baby by cesarean section. There was no history of twinning in the family of both parents. The baby was healthy except for the deformity on the back due to a fully formed hind limb at the lumbar region with genitalia (Figure 1) and an underdeveloped upper limb in the thoracic region in the midline (Figure 2). Movement was present in the abnormal lower limb on tactile

stimulation. The autosite's right lower limb was noted to be more active than the left. There was congenital talipes equino varus (CTEV) of the left foot with that foot appearing smaller than the opposite one. There was a poorly developed male external genitalia attached to the thigh of the parasite.

Figure 1: Fully formed lower limb in the lumbar region



Figure 2: Underdeveloped upper limb thoracic region



The parents had been stigmatized by the appearance of the baby and they had to be counseled. The baby also had to be hidden from the general public to maintain privacy.

A plain radiograph of the spine performed revealed spina bifida in the lumbar region with a normal thoracic spine (Figure 3). An abdominal ultra sound performed revealed no abnormality. A decision to operate was made. The surgery was performed on the 9th day after birth with induction of anesthesia and intubation being performed with the patient in the lateral position, and then turned prone for the surgery. The thoracic limb was excised through an elliptical incision in the longitudinal axis while the lumbar limb was excised after raising a proximally based flap. A neurovascular bundle was found attached to the abnormal upper limb (Figure 4).

Figure 3: Lumbosacral Radiograph showing lumbar spina bifida



Figure 4: Neurovascular bundle attached to the upper limb



No bony attachment to the autosite was found in either of the limbs. The defect in dura in the lumbar region was also repaired. The limbs excised are shown in figures 5 and 6, with the post operative picture in figure 7.

Figure 5



Figure 6



The operation lasted 2.5 hours with a significant blood loss (40mls) warranting transfusion of 60mls of whole blood. The post operative recovery was good except for CSF leakage through the lumbar wound. This resolved with conservative management. The patient

with the mother were discharged and subsequently referred to the orthopedic team for correction of the CTEV of the left foot. Hydrocephalus did not develop in the post operative period.

Figure 7: Immediate post operative photograph



The patient has been on follow up and the eight year review in January 2012 revealed a healthy school going child who is of appropriate size for age and normal intelligence. The child participates in football matches at school. He walks with a limp. The surgical scars have matured well except for a swelling over the lumbar surgical site (Figure 8).

The left leg has reduced bulk, 19cm against 20cm for the right (measured at 5cm below the tibial tuberosity). The left foot is smaller than the right at 15cm, compared with 21cm for the right (Figure 9).

Figure 8: Swelling over the lumbar surgical site



Figure 9: lower limb discrepancy

There is no sensory deficit but the power in the left ankle flexors and extensors is grade 3. The parents are satisfied with the general condition of their child and do not want any further surgery.

Discussion

The incidence of conjoined twins is approximately 1 in 50,000 births; conjoined twinning with parasitic attachment is even rarer (4).

The conjoined twins can be divided into 8 different types according to the conjoined parts: The eight types of conjoined twins are cephalopagus, thoracopagus, omphalopagus, ischiopagus, parapagus, craniopagus, pygopagus and rachipagus (4, 5, 6)

Rachipagus is defined as conjoined twins joined dorsally in the vertebral column and was first mentioned by Spencer (3,7). Most rachipagus conjoined twins are female (7). The tissue type varies in the rachipagus parasite, but the most common is a single limb (3). Usually the parasitic limb is the upper extremity in cervical Rachipagus and the lower extremity in thoracic or lumbar rachipagus (3). The extent of the attachment in the parasitic twin is also varied in different case reports. Most parasitic twins had better developed lower than upper limbs. Our patient had a parasite with a poorly developed upper limb and a well developed lower limb with external genitalia. This may be because either the lower half of the body in the parasite is more resistant to resorption or the resorption process takes place in a cranio-caudal direction (1). It is not uncommon to find a neurovascular bundle going from the autosite to the parasite as in our case (4). There is a high incidence of craniovertebral anomalies and neural tube defects

among the autosites. Other malformations that may be associated with the autosite include patent ductus arteriosus, atrial septal defect, gallbladder hypoplasia and club foot (7) - our patient had spina bifida and neurological CTEV.

The differential diagnosis of rachipagus can be fetus-in-fetu or teratoma. Fetus in fetu is a hypoplastic parasitic fetus, which may appear in the body cavities of the autosite while a teratoma does not form any normal organ.

The treatment of this condition is usually staged with the first focusing on excision of the parasite and later the malformations that are associated with the autosite. We staged our surgery with the first surgery to remove the parasite and the second to correct the talipes equinovarus. The treatment is normally without complications, since there is always a tissue plane between the autosite and parasite. The complications that occur are related to treatment of the associated malformations. One patient is reported to have developed temporary paralysis of a leg and another suffered urinary incontinence after operation of a tethered cord syndrome (7). Our patient developed cerebrospinal fluid leakage which resolved spontaneously.

The growth and development of our patient has been normal except for the slight weakness of the left lower limb which makes him walk with a limp. This has not interfered with his activities as child.

Rachipagus conjoined twins are considered to be very rare and in Africa there is only one report by Sanoussi et al (8) who reported on rachipagus conjoined twins in Niger. It represents an example of deranged embryogenesis. If well developed abnormal tissues

are found on the dorsal midline of a newborn then it should be considered. Thorough physical examination and investigations for other malformations should be performed before any surgical procedure. The hope for survival of the autosite is excellent if there are no significant life threatening congenital anomalies.

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