Parasitic Rachipagus Conjoined Twins With Spina Bifida, Diplomyelia, Scoliosis, Tethered Cord Syndrome, and Ventricular Septal Defect
—Case Report—

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Abstract

A 17-year-old girl presented with a rare case of parasitic rachipagus conjoined twins associated with spina bifida, diplomyelia, scoliosis, tethered cord syndrome, and ventricular septal defect. Physical examination found a well developed breast and an apophysis on the back of the patient, and neuroimaging demonstrated scoliosis, spina bifida from T8 to L5, butterfly-shaped vertebra of T6, abnormal bone behind T7, diplomyelia, and tethered cord. Successful surgical excision of the parasitic mass was performed. Histological examination discovered uterine tube, Muller’s duct, cartilage, and nerve tissue besides the mammary gland. Rachipagus conjoined twins are extremely rare, but should be considered if well developed abnormal tissue is found in the dorsal midline of the spine. However, the possibility of malformations in other organs in the autosite should be considered. Careful preoperative examination and refined microsurgery may provide good outcome for the patient.

Key words: rachipagus, diplomyelia, spina bifida, tethered cord syndrome

Introduction

Rachipagus, from the Greek rachi- (spine) and -pagus (fixed), is defined as conjoined twins joined dorsally in the vertebral column and was first named by Spencer.9) Rachipagus is the most rare type of all conjoined twins with no more than 30 cases reported. One of the embryos is always dead or defective, resulting in a parasitic twin. The most common parasitic organ is accessory limb. We treated a patient presenting with a mature breast attached to the back in the midline over the thoracolumbar region, who underwent successful surgical excision of the parasitic twin.

Case Report

A 17-year-old female was admitted to the hospital with a breast on the back which enlarged with age. This patient was deserted after birth with a small mass of about 2-cm diameter, and she was brought up in an orphanage. A nipple was present on the top of the mass, but she had not sought medical treatment until the mass developed to a mature breast of about 10-cm diameter, which affected her appearance. The breast had developed well without discharge or pain. When she was 15 years old, a cardiac murmur was found occasionally during medical examination. Further ultrasonic cardiography identified an 11-mm defect between the bilateral ventricles. The ventricular septal defect was repaired by thoracotomy and the murmur disappeared after the operation. She had no history of weakness, hypesthesia, or change of urination and bowel motions.

Physical examination was remarkable for a thoracolumbar mature breast measuring about 10 × 9 × 7.5 cm in the midline of the dorsal area between T10 and L1 (Fig. 1). The breast had developed well with a normal nipple. An apophysis with a few pelages were located on the left side of the breast. Scoliosis was obvious to the right side. The left buttock was a little bigger than the right. The power, tone, and coordination of the bilateral upper and lower limbs were normal and symmetrical. There was no difference in superficial and deep sensation between the bilateral upper and lower limbs. Sensation of the skin around the anus was symmetrical, and the perianal reflex was not abnormal. Knee jerks were present. Kernig's signs and Brudzinski's signs were negative, and no pathological reflexes were identified.

Thoracolumbar radiography, computed tomography, and magnetic resonance (MR) imaging showed that the vertebral column deviated to the right with the core at T7, the left lower part of the T5 vertebral body was flattened, the T6 vertebral body was collapsed in the central part and shaped like a butterfly, spina bifida was present from T8 to L5, and the left vertebral plate was fused partially from T8.

Received November 22, 2010; Accepted March 23, 2011
Parasitic Rachipagus Conjoined Twins

Fig. 1 Photograph showing mature breast in the midline of the dorsal area between T10 and L1 with apophysis located on the left.

Fig. 2 Thoracolumbar radiograph showing that the vertebral column deviated to the right with deficiency of the vertebral plate between T8 and L3.

Fig. 3 Reconstructed computed tomography image showing an abnormal bone shaped like a scapula protruding from the left side of the 8th costal bone.

Fig. 4 Coronal T2-weighted magnetic resonance image showing that the spinal cord was separated by an excrecent bone from T8.

Fig. 5 Sagittal T2-weighted magnetic resonance image showing that the end of the spinal cord was located at L3.

to T11 (Fig. 2). An abnormal bone shaped like a scapula protruded from the left side of the 8th costal bone (Fig. 3). The spinal cord was separated by an excrecent bone from T8 (Fig. 4). The end of the spinal cord was located at L3 and the filum terminale was enlarged (Fig. 5). Brain MR imaging showed no abnormalities, with no hydrocephalus, Arnold-Chiari malformation, or other malformations. Ultrasonography and dynamic examination of the urinary

Neurol Med Chir (Tokyo) 51, October, 2011
system were not abnormal. Karyotype analysis indicated normal chromosome, 46,XX.

Surgery was performed to remove the ectopic breast and bone, and repair the diplomyelia. The patient was positioned prone and a fusiform skin incision was made around the base of the breast. After liberation and total resection of the breast from the base, we found the deeper structure resembled a crater, in the middle of which was a lump of soft tissue well supplied by a neurovascular bundle connected to the vertebral canal. The soft tissue was resected. Exploration of the caudal part of the soft tissue found a bony spur which divided the dura into two parts and might be responsible for the diplomyelia. We opened the dura and resected the bony spur. Then we made another incision around the process of the ectopic bone, and separated and totally resected this bone. The ectopic bone resembled an aplasia scapula (Fig. 6). After excision of the accessory breast and bone, the surrounding skin flaps were used to provide adequate wound coverage.

Blood loss during the operation was about 800 ml, but the subsequent recovery was uneventful. Histological examination showed normal breast development, and the soft tissue beneath the breast contained skeletal muscle, cartilage, nerves, paramesonephric duct cyst, and fallopian tube. The diagnosis was parasitic rachipagus. The tethered cord was not released, so 2 weeks later, the patient was treated for a second time to cut off the filum terminale. She recovered well and was discharged 7 days later. No change occurred in neurological and urinary dynamic examinations during the 1-year follow up.

**Discussion**

The incidence of conjoined twins is approximately 1 in 50,000 births. The conjoined twins can be divided into 8 different types according to the conjoined part: thoracopagus, omphalopagus, pygopagus, ischiopagus, craniopagus, parapagus, cephalopagus, and rachipagus.10) Rachipagus denotes conjoined twins joined dorsally to the vertebral column, presumably united by the vertebral arches, with a common spinal canal flanked by the separate vertebral bodies of the individual twins. Most cases do not consist of two complete infants, instead there is usually a dorsal parasite on an autosite, which demonstrates embryologic derangement of the vertebral column and/or spinal cord. Rachipagus conjoined twins are extremely rare. A review of more than 1200 cases of conjoined twins found one classic example of 2 complete infants with dorsal union of the entire head and trunk. Radiography showed clearly visible bony union of the vertebral arches from T6 to L3.9) Subsequently, we found 20 cases of dorsal parasitic twinning. Reports about rachipagus total no more than 30 cases.

Two theories for the embryogenesis of rachipagus conjoined twins have been proposed, fission theory and fusion theory.11) The fission hypothesis is similar to the theory for the development of monoamniotic monozygotic twins in which stimulus is encountered at approximately developmental days 13- and 14, and the embryo may divide incompletely, leading to the formation of conjoined twins.9) Although the fission hypothesis can explain some phenomena, the most accepted theory is the fusion hypothesis. The normal embryology involves primarily the neural folds as they become elevated above the surface of the embryonic disc and fuse together to form the neural tube. Disintegration of the ectoderm on the folds allows the neural plate to separate from the overlying surface and close into a tube. At the same time, the interrupted exterior layer of ectoderm unites to form an intact covering over the tube. During the embryogenesis of rachipagus, two monovular embryonic discs occupy opposite aspects of a common amniotic cavity, and continuing enlargement of these embryos would allow dorsal contact in the area of the neural folds. If the skin is intact, the embryos will not fuse. However, in rachipagus, the ectoderm is disrupted to allow the neural tube to close, and this disruption allows the neural folds of two embryos to join resulting in rachipagus. If the embryos develop equally, rachipagus with two complete infants will result. However, under most conditions, one of the embryos will fail to survive, leaving parasitic body parts overlying or attached to the vertebral column of the autosite, always vascularized and often innervated by the autosite.9,11,12) A neurovascular bundle may be present between the parasitic body and autosite.1,2,7) In our case, we also found a neurovascular bundle connecting the ectopic breast to the vertebral column.

Most rachipagus conjoined twins are female, and the conjoined part is located on the dorsal midline more often than on either side, especially the right side. Rachipagus in the thoracic and lumbar areas are more common than in the cervical area. If the dorsal union is located only in the sacrum and/or coccyx, the twins are called pygopagus but not rachipagus. The tissue type varies in the rachipagus parasite, but the most common is a limb.9) Usually, the parasite limb is the upper extremity in cervical rachipagus, and the lower extremity in thoracic or lumbar rachipagus.9) In addition, cartilage, nipple, eyeball, lung, gland, kidney, genital organ, urinary bladder, ureter tube, anus, scrotum, and intestinal canal are also found in parasites.1,2,5,7,8,11,12) Cardiac and central nervous system tissue are rare in parasites, although some nervous tissue from the rachipagus autosite may dominate the parasite.9)

Rachipagus autosite is commonly associated with other malformations in rachipagus conjoined twins. These malformations mainly contain nervous system malformations, such as vertebral body malformation, spina bifida, tethered cord syndrome, and cystic cranioschisis. Rachipag-
pagus twins conjoined in the occiput, cervical, or thoracic areas could also be combined with anencephaly or hydrocephalus. Other malformations may also be found in the rachipagus autosite such as patent ductus arteriosus, atrial septal defect, clubfoot, gallbladder hypoplasia, rectovaginal fistula, syndactyly, and horseshoe kidney.\(^7\)\(^9\)\(^10\)\(^11\)\(^12\)

Based on the typical clinical manifestation and pathologic characteristics, rachipagus conjoined twins are unlikely to be misdiagnosed, but must sometimes be discriminated from fetus in fetu and teratoma. Fetus in fetu is also a rare developmental malformation first described by Meckel and the standard of diagnosis proposed by Willis in 1953. However, there are few reports on fetus in fetu.\(^4\)

Fetus in fetu has some common characteristics, such as vertebral axis and hyperdeveloped organ structure, and the vertebral axis is an important standard of diagnosis. Fetus in fetu mainly appears in the abdominal cavity or cavitas pelvis, but may also occur in the intracalvarium, oral cavity, and thoracic cavity.\(^5\)\(^9\) Fetus in fetu is a hypoplasic parasitic fetus, so could involve similar pathology to rachipagus. The main differential point between rachipagus conjoined twins and fetus in fetu is that fetus in fetu has vertebral axis and does not occur in the dorsal part of the spinal column. Teratoma is caused by abnormal aggregation and differentiation of pluripotent cells, so does not form a normal organ, and can be distinguished from rachipagus conjoined twins. However, rachipagus conjoined twins associated with teratoma has been reported.

The treatment of rachipagus conjoined twins is simple. No case of both rachipagus twins surviving has been reported, so the surgery does not involve separation of the twins, reservation of the neural function, or transplantation of the skin flap. The surgery is only required to resect the parasite. Other combined malformations should also be resolved, such as myelomeningocele or tethered cord.\(^8\)

If the patient has spina bifida, spine reconstruction should be considered.\(^4\) All previous patients achieved good outcome except that 3 patients before 1935 died after surgery, one patient had temporary paralysis of a leg,\(^9\) and one patient suffered urinary incontinence after operation of tethered cord syndrome.\(^9\)

Neuroimaging and pathological examination confirmed that our patient harbored rachipagus parasite. Our case demonstrated many specific characteristics. Firstly, all previous patients were less than 7 years old, and most were infants and neonates, so our case is the oldest till now. Secondly, the clinical manifestation of parasite is accessory limb in most cases, whereas our case had well developed breast as the main manifestation. Thirdly, the reported cardiac malformations of the autosite include atrial septal defect, patent ductus arteriosus, and exoccardia, whereas autosite combined with ventricular septal defect was found in our patient. Lastly, autosite with meningocele or tethered cord is reported, whereas our case presented with scoliosis in the autosite. Scoliosis has been reported in several cases of pygopagus,\(^3\) so our case indicates that rachipagus can also combine with scoliosis.

Rachipagus conjoined twins are extremely rare, but should be considered if well developed abnormal tissue is found in the dorsal midline of the spine. However, the possibility of malformations in other organs in the autosite should be considered. Careful preoperative examination and refined microsurgery may provide good outcome for the patient.

References


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