

Dorsolumbar Parasitic Rachipagus Twin, Case Report

Tariq Elemam Awad, MD., Karam Elsayem, MD., Mohamed Youssef Elqazaz, MD.

Neurosurgery and General Surgery Departments, Faculty of Medicine, Suez Canal University, Ismaillia, Egypt.

Abstract

Background Data: Siamese twins are joined by a part of their body at birth and the causes of the merger are often unknown. When the merger is on the spine it is rachipagus. It is an extremely rare and strange condition, and there are only a few documented cases in the literature. We present one additional case of parasite rachipagus on the dorsolumbar level.

Purpose: Our aim is to describe a girl with a parasitic twin attached to her lower back, combined with a spina bifida and a lipomyelomeningocele.

Study Design: Case report and review of the literature

Patient and Method: In September 2014, a female baby from North Sinai was referred to Suez Canal University Hospital. She was 1 month old and had an extra well developed parasitic twin attached to her back. She was surgically treated in the Neurosurgery and Pediatric Surgery Departments.

Results: A rare example of rachipagus conjoint parasitic twinning in a newborn girl is described. A lipomatous mass, a rudimentary intestinal loop with an attached atrophic pelvis and hind limb were found adherent to the dorsal vertebral arches of the autosite in the thoracolumbar region. Surgical excision of the parasitic twin, excision of the dorsal lipoma, repair of the cord and wound repair were done

Conclusion: Parasitic rachipagus is a rare embryogenic malformation with a good surgical prognosis on the autosite in the absence of associated congenital anomalies. (2016ESJ111)

Keywords: Rachipagus, Autosite, Parasite, Conjoint twinning

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Introduction

Siamese twins are joined by a part of their body at birth and the causes of the merger are often unknown. Classification of Siamese twins is based on the site of the union. The thoraco-omphalopagus are attached at the chest and abdomen (74% of the cases), pygopagus are joined at the buttocks (18% of cases), ischiopagus are attached at ischium (6% of the cases), and craniopagus at the level of the head (2% of cases). When the merger is on the spine it is rachipagus.⁷ Rachipagus is an extremely rare pathological entity. The union is most often at the posterior arches of the spine. Most of times, one of the twin embryos become atrophic and parts of it will survive attached to the normal embryo (the autosite) at the junction area. The atrophic embryo is called the parasitic twin.¹⁴ The rarity of rachipagus is obvious from the fact that in 1995, out of a review of 1,200 conjoined twins, Spencer¹⁷ could find only one typical example of rachipagus and an additional 20 parasites tentatively classified as rachipagus parasites. Any structure can be found in the parasite. Bones of the limbs are frequently present. More rarely heart and neural tube structures can be found in the parasite. In some lumbar parasites, the glandular tissue, intestine or anal orifice can be observed.¹⁵ We present one additional case of parasite rachipagus on the dorsolumbar level.

Case Report:

A female baby from North Sinai was born by Cesarean section after an uncomplicated full-term pregnancy, weighing 3250 g. The patient did well; she was referred to Suez Canal University Hospital. Antenatal course was uneventful and no history of maternal medication or harmful drug use or exposure to radiation during pregnancy was reported.

On physical examination, a well-formed parasitic twin was attached to the back, in the Dorso-lumbar region. The parasitic mass consists

of a well-formed foot with five toes, small tibia and fibula, knee joint, femur, atrophic ischial bone and a buttock like appearance. (Figure 1) Clinically, the autosite had no neurological deficit or evidence of any other congenital abnormality.

MRI brain, CT chest and abdomen, abdominal ultrasound, Echocardiography and MRI of the spine were performed. All radiographs showed no abnormality. In MRI of the spine the spinal cord is herniating in the bifid spine area at the level D12-L3. This part of the cord is attached dorsally to a lipomatous mass (dorsal lipoma) and they are surrounded by meningeal coverings. The parasitic mass is then attached. (Figure 2)

For excision of the parasite, the patient was operated in a prone position, and an elliptical incision was made at the base of the swelling, with superior and inferior extensions. Dissection was relatively easy. A well-demarcated avascular plane could be found between the spinal canal of the autosite and the parasite, except for a prominent vessel going from autosite to parasite, the origin of which could not be identified at surgery. The lipomatous element of the parasite was attached to the outside of the dura of the autosite. When the dura was opened, the partially unfolded posterior cord in the region of the parasite's attachment was found adherent to the overlying dorsal lipoma. The dorsal lipoma was excised and the cord was repaired into a tube using 7/0 Prolene sutures. At this stage no detethering of the cord was attempted. The dura was repaired. The soft tissue defect following the removal of the parasite was closed using tissue mobilization.

The resected specimen grossly showed ischium (at the base of the mass, near the site of the parasite's attachment) articulating with a single femur, that in turn articulated with a single fibula and tibia. The attached foot consisted of tarsals, metatarsals and toes. In addition to plenty of lipomatous tissue, abortive formation of small bowel loop with well-

developed mesentery was found at the bottom of the mass. Pathological examination revealed no neoplastic tissues. (Figure 3)

The postoperative period was uneventful, and the patient was discharged on the 5th postoperative day. Suture removal was done on the 14th postoperative day. The girl is doing

well after 1 year and is walking. (Figure 4) She is on close neurologic follow-up, though no neurologic deficits have been seen. Follow up MRI dorsolumbar spine showed no residual lipomatous tissue but the spinal cord is tethered. This is no clinical signs of cord tethering but close follow up is mandatory. (Figure 5)

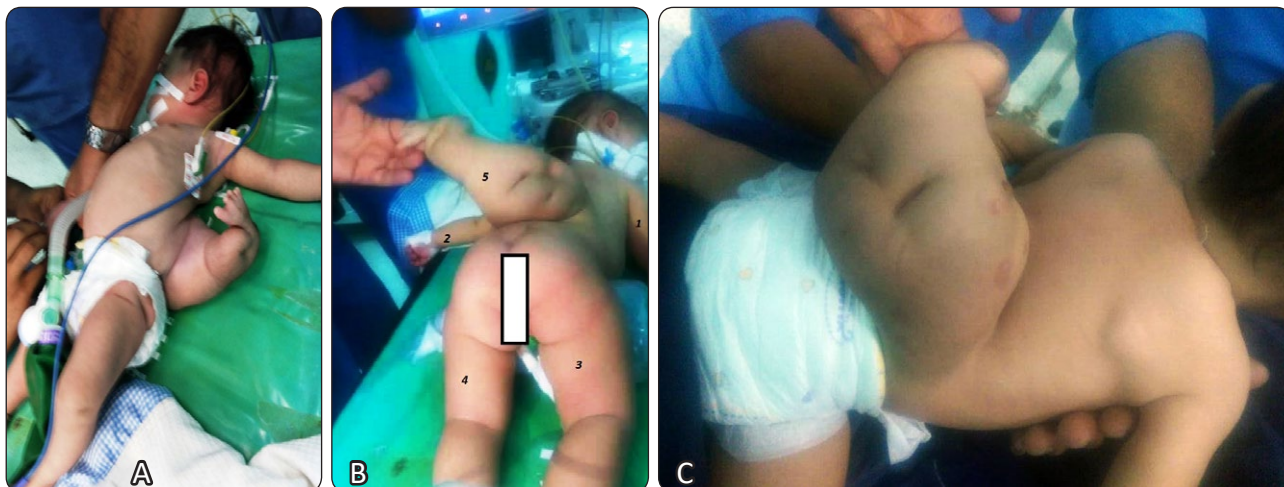


Figure 1. A well-formed parasitic rachipagus twin attached to the dorsolumbar spine. A: supine B and C prone position. The parasite consists of a well formed foot, leg, knee, thigh, pelvic part and a buttock like appearance.

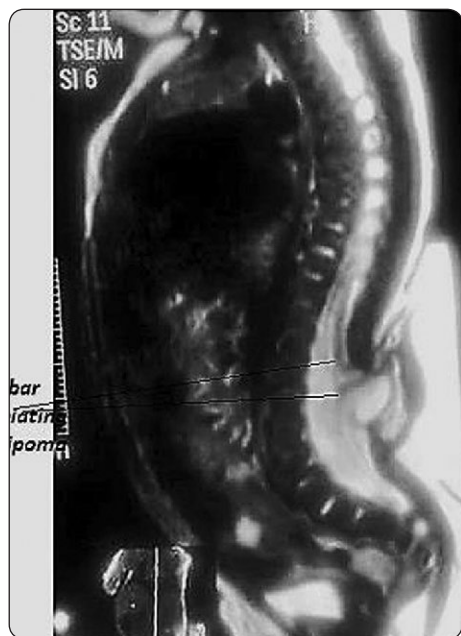


Figure 2. Preoperative MRI of the spine of the autosite T2 weighted image showing herniation of the dorsolumbar spinal cord into the spina bifida and attached to a dorsal lipoma.



Figure 3. (A, B) the parasite consists of a well formed foot, leg, knee, thigh, pelvic part and a buttock like appearance. (C) The autosite after excision of the parasitic twin.



Figure 4. Follow up when the patient is one year old. A: the girl is neurologically intact. B: Wound healing properly.

Figure 5. Postoperative MRI of the spine of the autosite T1 weighted image showing the dorsolumbar spinal cord at the spina bifida with no residual fat. The spine cord is tethered but the girl is neurologically intact.

Discussion

Heteropagus is a term used to describe the development of an asymmetrical form of twinning when one of the twins monopolizes the placental blood at the expense of other fetus with consequent ischemic atrophy of the latter.⁷ Some parts of the damaged fetus are attached to the partner's body, continuing to grow and developing like a parasitic organism.^{7,10} Conjoined twinning are classified in terms of the attachment site of the body: thoracopagus (thorax), ischiopagus (pelvis), cephalopagus (face), omphalopagus (abdomen), and craniopagus (cranium).^{11,14} Rachipagus describes a parasitic twin joined dorsally at the vertebral column. If the conjoined region is located at the lumbosacral spine, it is called pygopagus. Sometimes a parasitic fetus may completely develop and connect to partner's body via

laminae of the thoracic vertebrae. Sometimes, depending on the severity of the pathology, only a mass of viable tissue may remain.^{3,4,14}

The most widely accepted theory explaining the embryogenesis of this abnormal condition is the fusion hypothesis. In the early fetal life, embryonic discs of monozygotic monoamniotic twins are located in the same amniotic cavity. At the third or fourth gestational weeks, the neural folds of the two different embryos can merge if the skin covering the neural tube gets damaged. If the two embryos develop fair and equally, two complete but conjoined fetuses arise.²¹ But, in most cases, one of the twins dies and some parts of its body can remain attached to the vertebral column of the other fetus.^{7,21} This attached part composed of primitive embryonic tissue prevents closure of the neural tube during later development, resulting in spina bifida, or other neural tube defects.^{8,14} Thus, the most

anticipated anomaly of the living fetus is the closure defect of the dorsal vertebral column. Location of the spinal lipoma at the same level and the origin of the parasite support this explanation. Important abnormalities involving other organ systems are rare in the living fetus. In patients with rachipagus, a lipoma at the base of the parasitic mass with an intraspinal extension has frequently noticed.^{10,14,21} In the case presented here, the mass was firmly attached to the posterior elements of the dorsal D12 vertebra to the 3rd lumbar vertebrae, and the intraspinal lipoma was found at the same level.

Based on the typical appearances and histological results, diagnosis of rachipagus is made easily but rarely the differential diagnosis may be required from teratoma and fetus in fetu.^{8,18} Teratoma is defined as a true tumor arising from the uncontrolled growth of pluripotent stem cells.^{13,18} Sometimes, teratoma may contain mature tissues such as respiratory epithelium, fat tissue, hair follicles, fingers, teeth, and jawbones. However, these bones are scattered throughout the tumor and never resemble the real limb. If the fetus is located inside the body of its twin, this process occurs in the other twin's body cavities; this condition is termed fetus in fetu.^{8,13,19} Sometimes, a malformed, abnormal fetus enveloped inside its twin may remain totally silent until later ages, or it can cause an intra-abdominal or sometimes an intrathoracic mass. However there have been a few reports of parasitic twin location in the head, sacrum, scrotum, and even in the oral cavity. Presence of vertebral axis and appropriate arrangement of other organs or limbs in its relation are the criteria to distinguish a fetus in fetu from a rachipagus and highly differentiated teratoma.^{1,5,6,20} We did not find any vertebral structures in our patient's specimen. The appearances, the external-midline location of the parasitic mass, and the presence of intestinal mucosal loop suggested the presence of conjoined twinning. However,

teratoma occurring in association with a fetus in fetu and rachipagus has been reported in the literature. Thus, such a clear distinction of these entities may not be true at all times.^{9,13}

Neural tube defects associated with ectopic mass have been reported in a few cases. Krishna et al,¹² reported accessory legs associated with spina bifida and rudimentary external genitalia. Sharma et al,¹⁶ reported two of three cases with spina bifida and hemivertebrae. Zhang et al,²¹ reported rachipagus conjoined twinning in a seventeen-year-old female with spina bifida, tethered cord, diplomyelia, scoliosis, and ventricular septal defect. They observed a well-developed breast attached to the back in the midline at low-thoracic region. We did not find any other osseous anomaly in the vertebral column. Unlike teratoma, 6%–10% of which are malignant, rachipagus is regarded as a benign condition.^{2,8} Treatment is surgical. Parasitic twin excision is sufficient for treatment, and it does not require complicated separation procedures.^{7,21}

In summary, a case of rachipagus is presented herein, which had a well-developed lower limb, was located at the midline of the dorsal region, and attached to the vertebral column. The literature of this amazing condition was reviewed. We concluded that this anomaly was limited to the infant's dorsum. Nevertheless, other midline pathologies including vertebral column, spinal column, and heart should be kept in mind. Thus, careful preoperative examination of the patient is essential for a good postoperative outcome.

Conclusion

Parasitic rachipagus is a rare embryogenic malformation with a good prognosis on the autosite in the absence of associated congenital anomalies.

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Address reprint
request to:

Tariq Elemam Awad, MD.

Neurosurgery Department, Faculty of Medicine, Suez Canal University, Ismailia,
Egypt - Email: Tariqelemam@yahoo.com

الملخص العربي

توأأم متطفل متصل بالعمود الفقري الصدري القطني: تقرير حالة

البيانات الخلفية: نستعرض في هذا البحث حالة نادرة الحدوث لطفلة تبلغ من العمر شهر حضرت لمستشفى جامعة قناة السويس بكتلة على شكل حوض ورجل ثالثة متصلة بالعمود الفقري الصدري القطني ووجود كتلة دهنية بالحبـل الشوكي للطفلة مع انفلاق للعمود الفقري في منطقة الاتصال

الغرض: توضيح الخصائص الأكلينيكية والنتائج الجراحية لحالة نادرة الحدوث لطفلة تبلغ من العمر شهر تعاني من كتلة على شكل حوض ورجل ثالثة متصلة بالعمود الفقري الصدري القطني ووجود كتلة دهنية بالحبـل الشوكي للطفلة مع انفلاق للعمود الفقري في منطقة الاتصال

تصميم الدراسة: دراسة لحالات أكلينيكية لحالة نادرة ومراجعة للدراسات العلمية

المرضي والطرق: تم إجراء الجراحة في سبتمبر ٢٠١٤ في قسمي جراحة المخ والأعصاب وجراحة الأطفال في مستشفى جامعة قناة السويس بالإسماعيلية

النتائج: تم استئصال الجزء المتطفل المتصل بالعمود الفقري مع استئصال للورم الدهني وإصلاح فتق الحبل الشوكي والأم الجافية وإغلاق الجرح في طبقات واستئصال الجلد الزائد

الاستنتاج: التوأأم المتطفل المتصل بالعمود الفقري هي حالات نادرة وراثية يمكن علاجها بنجاح في حالة خلو الجنين الأصلي من العيوب الخلقية