Rachipagus: A Report of Two Cases - Thoracic and Lumbar

Samuila Sanoussi, Sani Rachid[1], Chaibou Maman Sani[2], Bawa Mahamane[2], Guemou Addo

Departments of Neurosurgery, [1]General Surgery and [2]Anesthesia, National Hospital of Niamey, Niger

ABSTRACT

We present two cases of rachipagus in two male infants and review the literature on this anomaly. These infants were from consanguineous marriages and cases of twins were reported in their families. In the first case it was a limb attached to the lower lumbar region with a rudimentary posterior arch. At the junction there was a lipomeningocele. Anatomical dissection of the limb identified the bones of the lower limb. In the second case, the parasites were joints of the upper limb that were attached to the chest by rudimentary posterior arches. In both cases there was only one spinal canal and a single spinal cord. Except the spina bifida in the first case no other malformation was diagnosed. The parasites were successfully excised. The two patients are well at one year of follow-up. Rachipagus is a rare embryogenic malformation with a good prognosis in the absence of associated congenital anomalies.

Key words: Niger Republic, parasitic twins, rachipagus, siamese

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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.[1] Two cases of Siamese twins in connection with taking clomiphene and two with griseofulvin have been reported.[1] Spina bifida is associated with some Siamese twins. Some authors[1] have suggested an abnormal inactivation of the X chromosome. No ethnic factor is reported in the literature.[1] 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 ischions (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. The term rachipagus is derived from Greek words, with spinal pagus meaning set. The union is most often at the posterior arches of the spine. Rachipagus is an extremely rare pathological entity.[2] The first case reported was that of Deslonchamps, in 1851,[3] cited by Taruffi C 1882[4] and Schwalbe in 1907.[5] More recently a meta-analysis of 1200 cases of merged twins reported another case.[6] The other rachipagus (21 cases) of this series of 1200 cases are rather defined as rachipagus parasite. We present two additional cases of parasite rachipagus on the chest and lumbar levels at The National Hospital of Niamey, Niger.

CASE REPORTS

Case 1: [Figures 1 – 3]

A three-month-old male infant with a superrnumerary (extra) leg attached to the lumbar region was admitted in the neurosurgery services of the Niamey National Hospital, Niger. This boy was from a consanguineous marriage, after a pregnancy of eight months, and he was delivered per vaginum. The mother was 17 years old and a history of twin pregnancy was reported in the father's family. The infant had a good general condition, and the weight and height were normal. He had no motor or sphincteric deficit. The back outgrowth looked to be a limb attached to the lower lumbar region with a proximal lipoma. We could identify the knee and foot with the toes attached. This limb did not move spontaneously or after stimulation, but the child cried when the parasite was pinched. There was an appendix located in the upper part of the junction area resembling the male sex organ. Radiographs of the parasite limb and the spine of the child showed the bone structures of the femur, tibia, and fibula, with a junction area at L3-L4. There was only one spinal canal. The posterior arch was absent from L2 to L5, with a duplication or agenesis of the posterior articular. The parasitic limb was excised at the base of which an extradural lipoma and a voluminous meningocele were found. The intra-dural content included a neural plate and normal roots with a single spinal cord. Excision of the lipoma, repair of the meningocele, and musculofascial reconstruction were easy, and the postoperative follow-up has been uneventful. Autopsy dissection of the parasite showed a dysmorphic ischium merged to the femur by a rudimentary capsule. Tibia and fibula were identifiable, but dysmorphic. The appendix at the top of the fusion zone proved to be a

Address for correspondence: *Dr. Samuila Sanoussi, Department of Neurosurgery, National Hospital of Niamey, BP: 10239 Niamey, Niger. E-mail: s-sanoussi@hotmail.com*

penis. No glandular tissue or other neural tube structures were found on histological examination. The follow-up (one year) was uneventful.

Case 2: [Figure 4]

This was a male infant born of a consanguineous marriage after eight months of pregnancy. He was seen in the Neurosurgery Clinic at the age of 18 months. He presented with a double outline of the upper limb grafted in the inter-scapular region in the midline. The general clinical and neurological examinations were normal. There was no spontaneous motor reflex in the parasite limb, but only a sensation of pain. Radiographs showed bones of the upper limbs with two phalanges. These bones were dysmorphic and merged to the dorsal spine, with spina bifida from T5 to T8. The bony part of each parasite corresponded, at the spine merger, to the autosite articular bones. These joints were absent in some places or duplicated in other segments. The dural sheath and the spinal cord were unique and apparently normal. Excision of the parasitic limb was accomplished without difficulty and the surgery was simple. Anatomical dissection of the parasite showed structures reminiscent of the scapula bone, humerus, radius, and ulna. No glandular tissue or other neural tube structures were found at the histological examination. The follow-up (one year) was uneventful.

DISCUSSION

Siamese twins occur in cases of twin pregnancies of the mono chorionic, mono amniotic type. They owe their name to Chang and Eng Bunker (1811-1874), who were merged twins, from Siam.[1] The classification of twins or Siamese twins is according to the site of union. The term rachipagus is derived from Greek words, with pagus meaning set. The

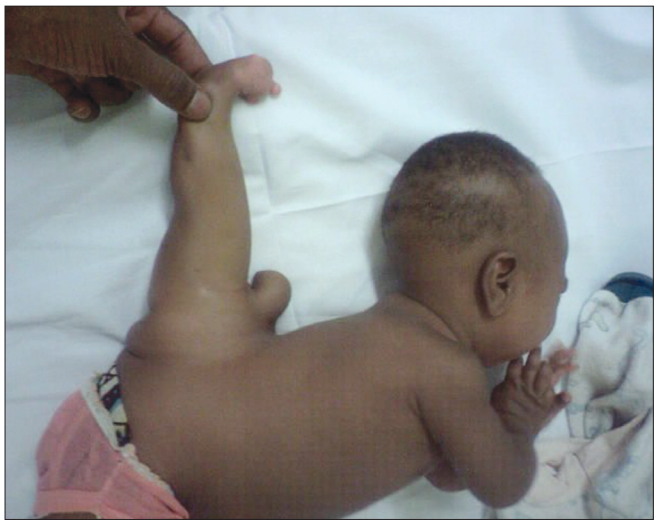

Figure 1: Lower extremity attached to the lumbar region before surgery

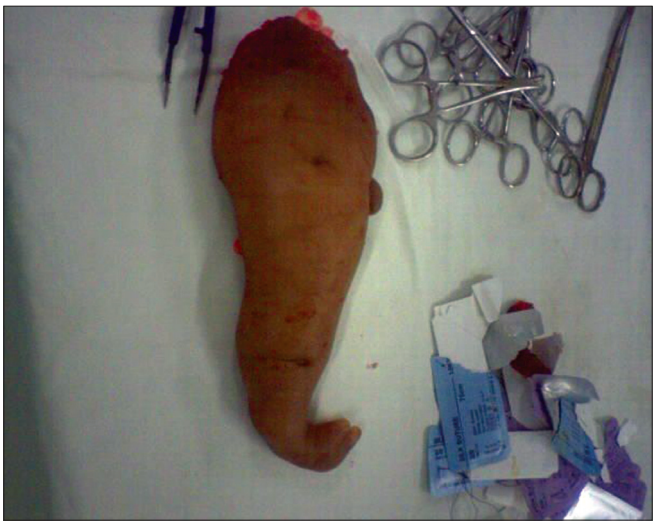

Figure 2: Lower extremity removed at surgery

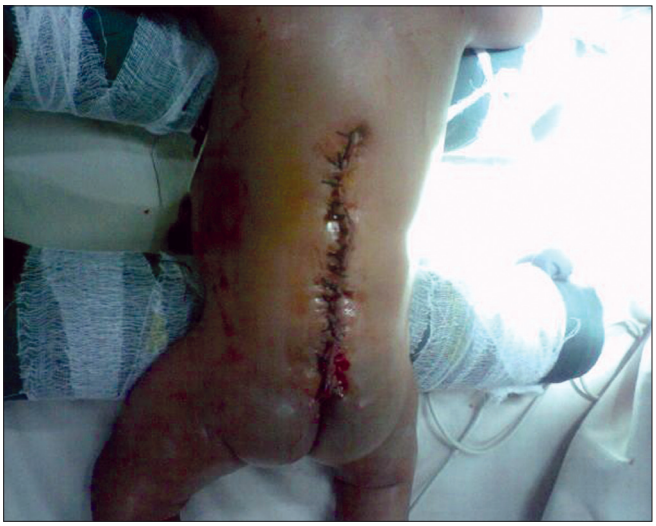

Figure 3: Same patient as Figure 1, immediately after excision

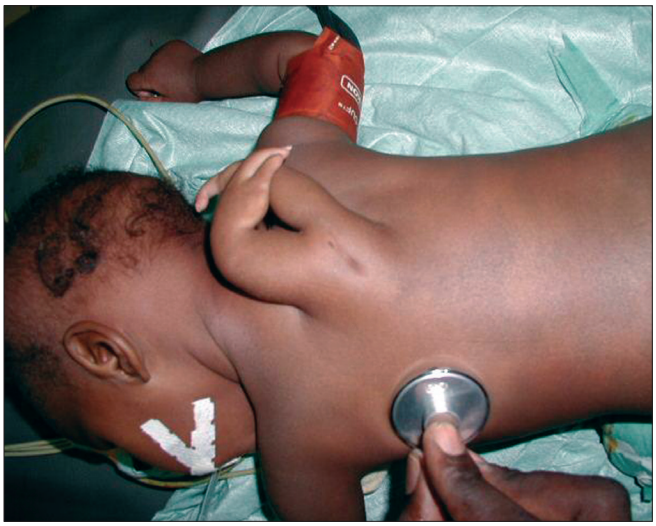

Figure 4: Upper limbs attached to the thoracic region

union is most often at the posterior arches of the spine. This is an extremely rare pathological entity.[2] Literature has reported few cases of rachipagus and parasite rachipagus.[3-6] The parasite follows the disappearance of one of the embryos with survival of certain additional structures attached to the normal embryo near or at the junction area. The two cases in this series are boys from consanguineous marriages with a family history of twin pregnancies. These cases were rachipagus parasite with the autosite healthy, without associated anomalies or neurological disorders. In one case the parasite was at the lower lumbar position and in the second case at the upper thoracic position. These locations of the parasite are reported in literature.[7,8] It is not uncommon to find sensory or motor innervations of the autosite.[7] In both cases we report that there was no spontaneous movement of the parasites, but the autosite was sensitive to painful parasite stimulation. The mechanism that leads to fusion of the twins begins very early in embryonic life, at D6, post fertilization at the blastocyst stage.[2] Rachipagus merger would ocur at the stage of the neural groove, before the closure of the neural tube, at specific areas called ectodermic depressions. Embryologic events during this period may form a single or double spinal canal with one or two spinal cords.[7] For Spencer,[7] fusion alone does not explain the embryogenesis of rachipagus. This is because during the dissection, the parasite contains abnormal anatomical structures and because neural groove fusion alone will only lead to a union of bones of the vertebral arches of the two embryos, with a unique spinal channel. Another mechanism may be associated with the merger. It is the mechanism of division-diversion-reunion, which explains the presence of two spinal cords and two vertebral columns in some malformations. The existence of a single channel containing a single spinal cord in both cases of this study supports the hypothesis of the fusion theory as the mechanism of occurrence of rachipagus. 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. Limbs are most frequently found in rachipagus parasites than in teratomas or in the fetus in fetus. This means that there is a thin line between these pathological entities.[8-10] In some cases a simple surgical resection of the parasite gives satisfactory results for autosites. Sometimes a multidisciplinary approach is needed because of associated anomalies.[7]

CONCLUSION

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

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