Case Report

Successful separation of craniopagus conjoined twins

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ABSTRACT

Craniopagus conjoined twins represent a rare phenomenon of congenital malformation/ dysmorphism. The clinical pathology of this complex entity is reviewed and placed in perspective. Confusion surrounds the severity of craniopagus conjoined twins especially in relation to the difficulty of separation and subsequent outcome. Successful separation of craniopagus twins remains a rarity, however modern neurosurgical techniques have created opportunities for successful separation and brought hope for a normal survival of these children who in the past were often left as historical footnotes or put on display as oddities of nature. We report on a craniopagus conjoined twins from Sudan who had been successfully separated.

Key Words:

Craniopagus; Conjoined; Twins; Sudan.

INTRODUCTION

Conjoined twinning is one of the most challenging human malformations. Treating conjoined twins can be a daunting challenge for the surgeon. Moreover, these cases often raise religious, moral, ethical and legal issues [1,2].

Craniopagus twins (CPT) i.e. twins joined at the head are an uncommon malformation found once in 2.5 million live births and represent only 2–6% of conjoined twins [3]. Approximately 40% of conjoined twins are stillborn and an additional one-third die within 24 hours, usually from congenital organ anomalies, leaving perhaps 25% to be considered for surgical separation [4,5]. Nevertheless, worldwide a handful of CPT separation attempts occur yearly. In the last half-century, with the many advances in medicine including brain imaging, neuroanaesthesia and neurosurgical techniques, a successful outcome is possible

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following separation of total CPT. In CPT, the face and foramen magnum are not primarily involved, the skulls are usually joined in roughly homologous regions but asymmetries are common, and both vertical and non-vertical or angular forms are found [6,7].

Stone and Goodrich [8] have recently sub classified craniopagus into four varieties, depending on whether a significantly shared dural venous sinus system (total versus. partial) is present, and whether the inter-twin longitudinal angulation is below 140 degrees.

We report on craniopagus female twins from Sudan who had been successfully separated. To our knowledge this is the first case report of craniopagus twins from Sudan.

CASE REPORT

The diagnosis of CPT was made during routine antenatal ultrasound scan in the first trimester. The mother was a 26-year-old primigravida. The delivery was by an elective Caesarian section at 36 weeks gestation on the 22nd of September 2010. The two babies cried immediately after birth with Apgar scores of nine at one and five minutes for both. Birth weight was 6.5 kg. The twins were transferred to the special care baby unit for further assessment. Clinical examination revealed that both babies were well, sharing one cranium vertically with completely separate bodies on each side (Figure 1). There were no dysmorphic features and systemic examination was normal for both. Initially the babies were kept on intravenous 10% Dextrose and later started on breast-feeding.

Investigations including complete blood counts, liver and renal function tests, cranial and abdominal ultrasound were normal. Echocardiography revealed normal anatomy for both hearts. A 3D cranial computed tomography (CT) with reconstruction showed craniopagus conjoined twins (vertical type 3) sharing a single bony cranium with defective parietal bones. Magnetic resonance imaging (MRI) showed cerebrospinal fluid (CSF) cleft communicating between the ventricular systems through the posterior parietal region. Magnetic resonance venography (Figure 2) revealed evidence of venous sharing through the superior saggital sinus at its posterior occipital extent. Magnetic resonance angiography (MRA) showed no obvious arterial sharing. Both babies passed urine and meconium normally. The twins were discharged home and arrangements were made for surgery in UK.

The parents contacted the charity organization, Facing The World (http://facingtheworld.net/), through the BBC which

Figure 1 - Craniopagus conjoined twins at birth.

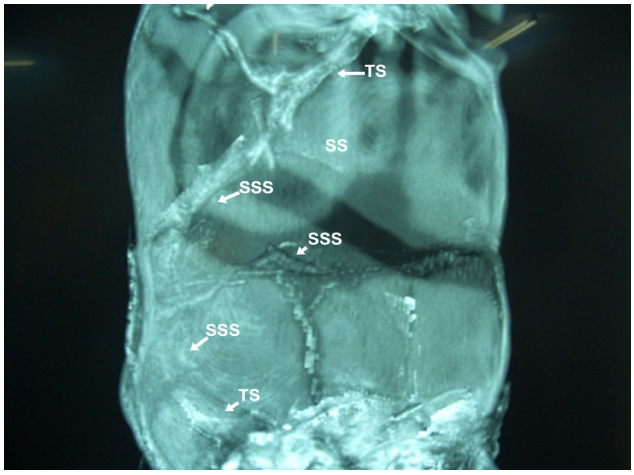

Figure 2 - Magnetic resonance venography (MRV) showing evidence of venous sharing through the superior saggital sinus at its posterior occipital extent. (SSS=superior sagittal sinus; TS= transverse sinus; SS= sagittal sinus).

made all the necessary arrangements to transfer the twins to the United Kingdom accompanied by their parents.

The surgeons at Great Ormond Street Hospital in London agreed to do this extremely hazardous surgery. The surgery took place in four stages, two operations in May, one in July and the final one was in August 2011.Tissue expanders were inserted in July 2011. The post operative course was uneventful and both girls were able to go to the general ward within a few days with no residual neurological deficit. Figure 3 shows the twins after separation at the age of 11 months interacting and playing.

Figure 3 - The craniopagus conjoined Sudanese twins (Rital and Ritaj) at the age of 11 months after successful separation.

DISCUSSION

A number of reviews have detailed the complex anatomical and surgical problems in separating total forms of craniopagus conjoined twins, notably shared dural venous sinuses and the subsequent negative implications towards survival and quality of life [9,10].

Experience with large numbers of conjoined twins is limited to a few centers in the world (13 sets described by O'Neill et al [11] from the Children's Hospital of Philadelphia, 14 sets reported by Mackenzie et al [12] from the same hospital, 17 sets reported by Spitz and Kiely [13] from Great Ormond Street Hospital, London, and 46 sets reported by Rode et al [14] from Red Cross Children's Hospital, Cape Town, South Africa. Recently, surgical experience with conjoined twins has been reported in other parts of the world, such as China, Saudi Arabia, and New Zealand [15-18].

There are few examples of successful and unsuccessful separation of craniopagus twins, notably in adult life. In Singapore in 2003, skull and brain separation of 29-year-old craniopagus Iranian twins was unsuccessful, and the sisters died of exsanguinations on the operating table [17].

Craniopagus conjoined twins from Egypt were separated at the Children's Medical Center in Dallas, Texas. They were born in Egypt on June 2, 2001, and were separated on October 12, 2003, during a 34-hour operation [19]. The operation involved skin grafting and separation of the venous sinuses. Later, they underwent skull reconstructive surgery, received rehabilitation therapy, and, finally, left for Egypt on November 19, 2005. Another successfully staged separation of craniopagus twins took place from 2003-2004 at Montefiore Medical Center in New York [20].

Conjoined twins is one of the most intriguing and provocative anomaly that has challenged mankind over the millennia. During the past half century craniopagus twins have challenged the capability of neurosurgeons. With modern neuroimaging and neurosurgical techniques it is now possible to separate such twins successfully.

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