

Radiological anatomy of a cranial malformation in a conjoined dicephalus twin

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SUMMARY

Conjoined twins capture interest due to their unusual anatomy. They vary in size, internal anatomy and degree of organ sharing. A sound knowledge of this structural anatomy is therefore important in separation surgery.

Non-invasive techniques including 3D ultrasonography, computed tomography (CT) and magnetic resonance imaging (MRI) can expose the details of the anatomical complexity precluding twin separation. This study reports the cranial anatomy of a *dicephalus dipus tribrachii* twin as determined by CT-Scan and MRI evaluations. The CT scan gave the best bone delineation while the MRI scans gave the best soft tissue picture. The paper illustrates the usefulness of multiple techniques in delineating conjoined twin anatomy for the purposes of separation planning.

Key words: Conjoined twins – CT and MR imaging – Cranial malformation

INTRODUCTION

Early records of conjoined twins have been found on cave drawings, stone tablets, papyrus, writings and folklore (Hoyle, 1990). Despite their long history, the precise aetiology

remains elusive (Hoyle, 1990; Cywes et al., 1997; Cywes et al., 1982). The reported incidence ranges between 1:2,800 and 1:200,000 deliveries (Cywes et al., 1982; Machin, 1993; Viljoen et al., 1983; Bhattay et al., 1975). The incidence is 1:14,000 in Central and Southern Africa (Cywes et al., 1982; Viljoen et al., 1983; Bhattay et al., 1975). Only a few cases of conjoined twins have been reported in East Africa (Shija and Ngiloi, 2000).

The commonest variety of conjoinment is thoracopagus twins accounting for 32.7% of cases (Machin, 1993). Dicephalus Dipus conjoined twins form a rare variant, and are usually stillborn or die immediately after birth. Since most of studies focus on the obstetric problems, the anatomical description of dicephalus twins is often incomplete.

MATERIALS AND METHODS

The stillborn twins were received at the department of Human Anatomy from a peripheral hospital where the mother had presented. The obstetric history was scant but previous antenatal visits and scans had reportedly indicated normal twins. No further details could be obtained.

The twins were perfused with 10% formaldehyde and 0.4% phenol via the left

femoral artery. The preservative was also injected at various body points namely: the cheeks, posterior head, neck, pectoral region, upper arm, forearm, abdomen, gluteal region, thigh and the legs for completion of fixation. Final preservation was by immersion of the specimen in 10% formalin throughout the duration of the study.

Computed Tomography and Magnetic Resonance Imaging were used to evaluate the craniocerebral anatomy. A three-dimensional data set was acquired with a Spiral CT scanner AR. ST. (Siemens Medical Systems, Erlangen, Germany) with a section thickness of 1 mm. MR imaging was performed with a 0.35-T MR imager (Siemens Medical Systems, Erlangen, Germany), using a head coil.

RESULTS

The twins had two heads with well-formed ears, eyes, nostrils, mouths and normal hair distribution (Fig. 1). The right head was smaller (head circumference 29.5 cm vs. 33.5 cm) (Fig. 1).

The fontanelles were normal on palpation. However, there was a posterior skull defect on the right head (Fig. 2).

Computed Tomography Findings

The tomogram confirmed the gross findings. The cerebral hemispheres, ventricular system, brainstem and cerebellum were all visualised as normal in the left head. There was, however, significant air both in the sub-



Fig 1a.- Whole body view of the twins.



Fig 1b.- Normal ears on the right head.



Fig. 2.- Note the posterior skull defect.

arachnoid and intra-ventricular spaces. The orbit and nasal passages were also normal.

There was a large cystic mass in the posterior cranial fossa of the right head communicating with the 4th ventricle. The cerebral peduncles were splayed by the enlargement of the 4th ventricle. The cerebellar vermis was not visualised. The lateral and 3rd ventricles were also dilated (Fig. 3). A vault deficiency was present posteriorly over the cystic mass. The cerebral sulci were poorly formed.

Magnetic Resonance Imaging Findings

The left head had a well developed skull with no vault deficiencies. The brain tissue showed well formed gyri and sulci. The gray and white matter differentiation was normal but intra cranial air was noted bifrontally (an artefact due to embalming) (Fig. 4). The lateral ventricles were visible and normal. The 3rd and 4th ventricles could also be visualised in the lower scans. Slices of the mid brain were normal. The cerebral aqueduct and pre pontine cistern was visible. The cerebellum was normal in both appearance and size while the

foramen magnum with the spinal cord could be seen and appeared normal.

The skull was defective over the right hind brain area (Fig. 5). A large 4th ventricular cyst completely occupied the posterior cranial cavity.

The cerebellar hemispheres were hypoplastic. The cerebral tissue was not well formed with the gyri and sulci completely absent in some areas. Grey-white matter differentiation was poor. The lateral ventricles were expanded and the foramen magnum was enlarged.

DISCUSSION

The twins described in this study possessed two heads, three arms and were fused at the level of the antero-lateral thorax and abdomen. This pattern of duplication and fusion meant that the set could be described as parapagus dicephalus tribrachii twins. Dicephalus twinning is an extremely rare type of conjoined twins (Daskalakis et al., 2004), accounting for only 11-13% of all conjoined twins. As in most other previous accounts

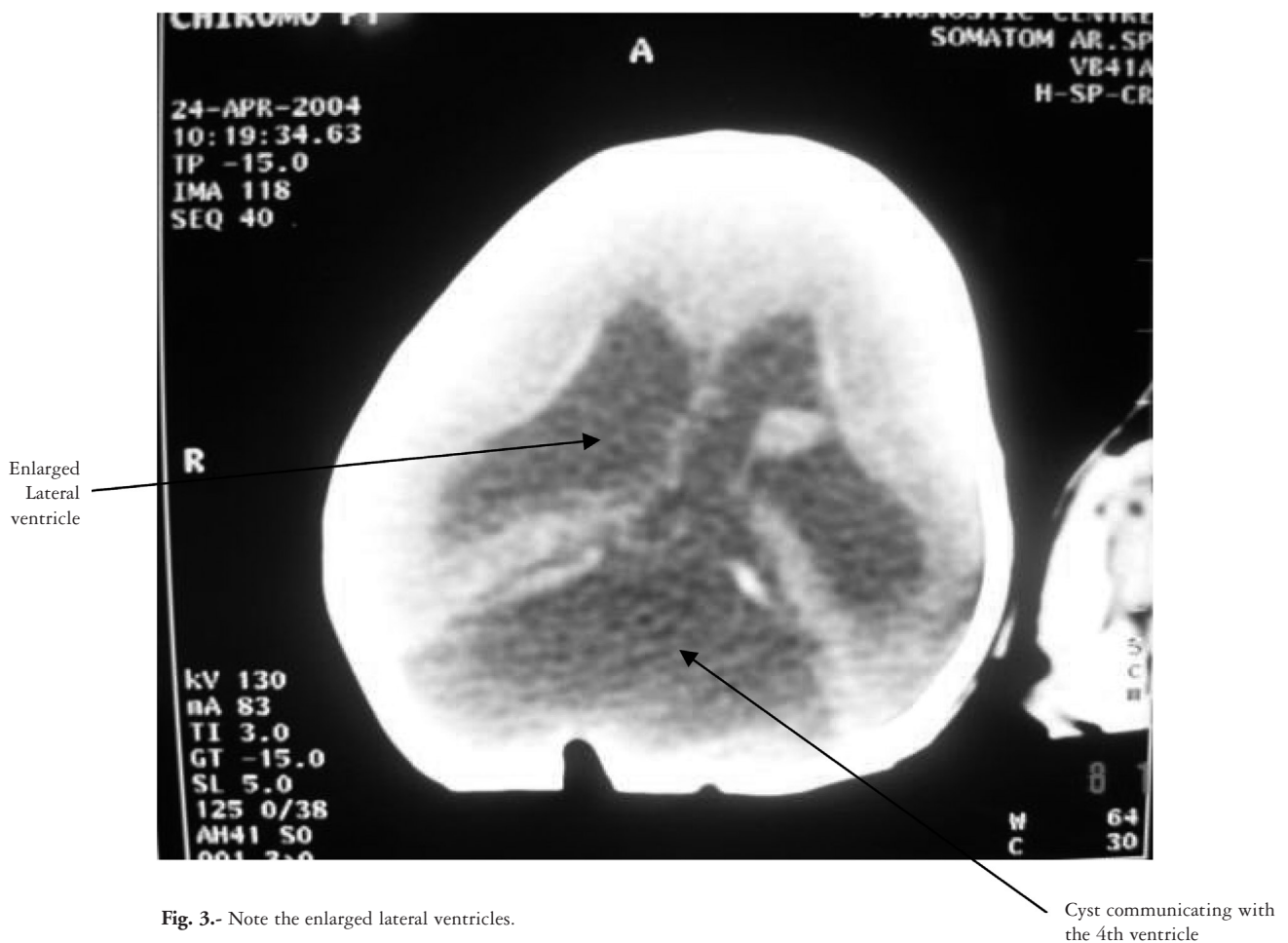


Fig. 3.- Note the enlarged lateral ventricles.

Cyst communicating with the 4th ventricle



Fig. 4.- MRI scan of the left head.

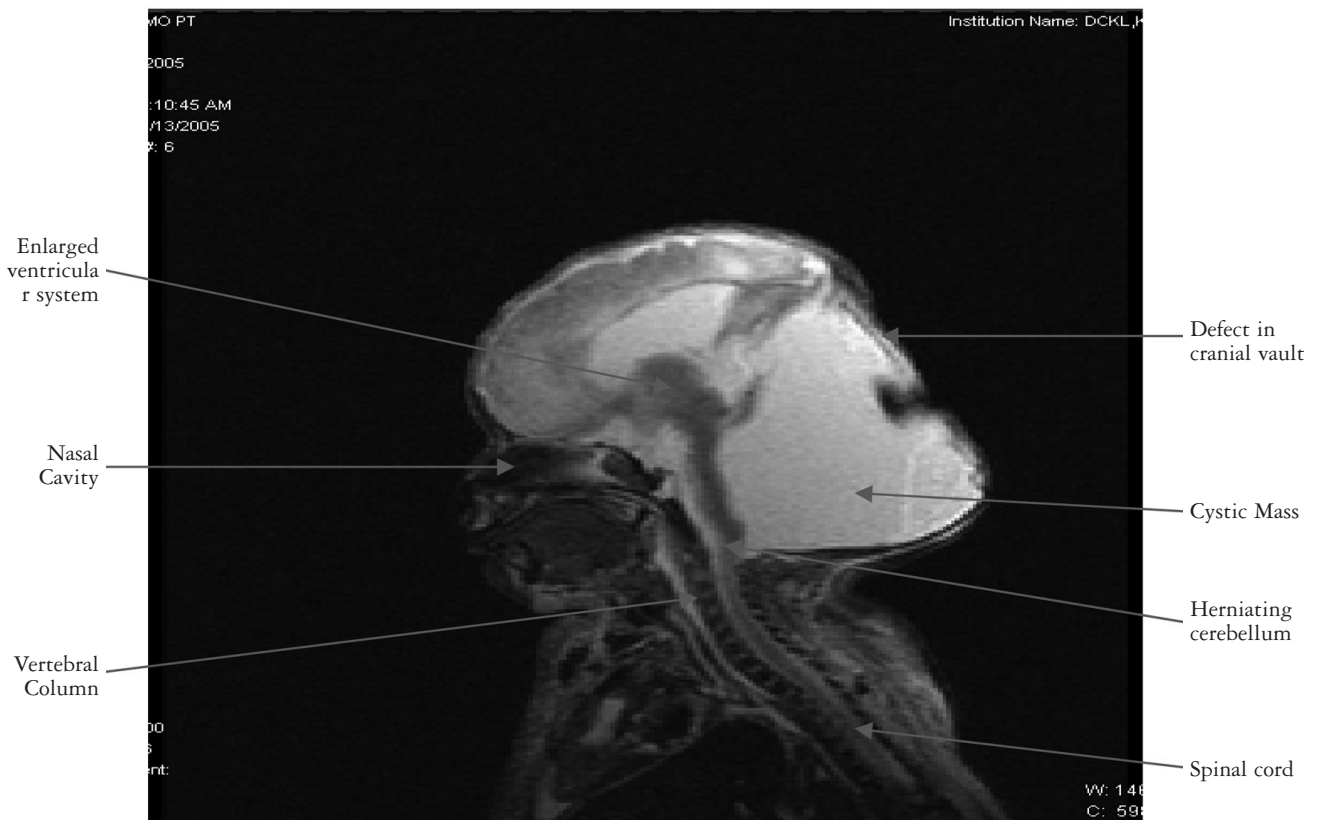


Fig. 5.- MRI scan of the right head.

(Golladay et al., 1982), the twins in this study did not survive beyond birth. They were however unique in surviving till full term since survival to full term in most cases is unusual (Golladay et al., 1982).

CT was used to evaluate osseous details and MR images to establish the degree of brain involvement. The skull and brain of the smaller twin had multiple anomalies. We contend that these nervous system anomalies formed part of a package inconsistent with survival. In cases of surviving twins with lesser degrees of conjoined anatomy, the brains may be imaged further by use of MR arteriography and MR venography to evaluate cerebral circulation (Respondek-Liberska, 2004).

The diagnosis of the conjoined twins studied here was made at delivery. This is the situation that obtains in many countries (Durin et al., 2005). In China prenatal diagnoses are made in only 31.13% of cases of conjoined twins (Liang et al., 1999). Antenatal ultrasound investigations may not be performed due to economic constraints (Colombo and Sterpa, 2004). Indeed, there have been reports of diagnosis after prolonged labour at laparotomies (Aiyedun, 2002).

Where prenatal diagnoses of such abnormal pregnancies are made, delivery may be optimized. In-utero diagnosis of conjoined twins by say, ultrasonography, allows the opportunity for physicians and parents early in the pregnancy to make decisions concerning pregnancy termination (Daskalakis et al., 2004, Votteler and Lipsky, 2005). In selected cases, possibility of a separation procedure after birth with acceptable outcome may be judged (Sen et al., 2003).

In conclusion, a combination of CT and MR images has provided good anatomic details, including the patterns of bony and soft tissues. Investigations in living twins may involve the use of contrast media, a true limitation in the post-mortem assessment here. Accurate diagnosis of anatomical abnormalities is key for surgical planning and prognostic information. Each set of twins is unique,

with the specific pattern of separate and conjoined anatomy at variance with descriptions in general texts.

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