Neuroradiological findings in adult cranially conjoined twins

Case report

OLAV JANSEN, M.D., V. A. MEHRABI, M.D., AND KLAUS SARTOR, M.D.

Department of Neuroradiology, University of Heidelberg Medical School, Heidelberg, Germany; and Department of General and Pediatric Surgery, University of Tehran, Tehran, Iran

The authors demonstrate the radiological anatomy and review the accepted embryological theories in a case of total craniopagus. These 24-year-old female cranially conjoined twins were studied with computerized tomography (CT) and CT angiography, magnetic resonance (MR) imaging and MR angiography as well as selective arterial digital subtraction (DS) angiography to clarify whether surgical separation was possible. The neuroradiological findings are discussed, taking into consideration both the embryological and surgical literature.

The malformation was classified as a total parietooccipitotemporal craniopagus. Whereas CT angiography and MR imaging including MR angiography demonstrated a common superior sagittal sinus, only selective arterial DS angiography revealed a significant arterial and venous cross-flow between the two adjacent temporal lobes. Selective intraarterial DS angiography is required in the neuroradiological evaluation of complex malformations, even when the anatomy of brain and skull can be well demonstrated with high-quality MR and CT studies.

KEY WORDS • craniopagus • magnetic resonance imaging • neuroradiology

Case Report

These 24-year-old female twins from Iran were admitted for neuroradiological diagnostic investigation to clarify whether surgical separation was possible. During childhood and adolescence the two girls had adapted satisfactorily to their situation and developed well. No other congenital abnormalities or significant neurological deficits were present. The intellectual potential of each of the twins was normal; both finished high school and recently had started to study law. Psychological evaluation revealed two distinct personalities: whereas the right twin (A), whose body was rotated more frontward, appeared extroverted, the left twin (B) appeared introverted. Twin A was right handed and Twin B was left handed.

Neuroradiological Methods

Cranial CT Scanning. These studies were performed by volume acquisition using the spiral mode (PQ 2000; Picker, Cleveland, OH). The dataset was used to reconstruct high-resolution axial images (slice thickness 3 mm) as well as three-dimensional (3-D) images.
Cranial MR Imaging. These studies were performed on a 1.5-tesla unit (Edge; Picker). Axial and coronal images with a slice thickness of 5 mm and a 0.5-mm gap were obtained using standard T₁-weighted spin-echo sequences (780 TR, 20 TE, 1 excitation). The T₂-weighted images, with a slice thickness of 5 mm and a 0.5-mm gap, were acquired in the axial plane with a standard spin-echo sequence (2484 TR, 90 TE, 1 excitation). Additional contiguous axial images with a slice thickness of 1.3 mm were obtained using a 3-D spoiled-gradient echo sequence (30 TR, 4.4 TE, 1 excitation; 30° flip angle). Further contiguous axial images with a slice thickness of 1.4 mm were obtained using a 3-D time-of-flight angiographic sequence (35 TR, 4.8 TE, 1 excitation; 23° flip angle). These images were then used for arterial MR angiography reconstruction accomplished by means of a maximal intensity projection technique. Venous MR angiography was performed using an axial 3-D phase-contrast sequence (70 TR, 7.9 TE, 1 excitation; 15° flip angle) generating 2.1-mm-thick contiguous slices, which were then used for MR angiography reconstruction performed by means of maximal intensity projection technique.

Cerebral Angiography. These studies were performed on a monoplane DS angiography unit (Diagnost Arc; Philips, Eindhoven, The Netherlands). A No. 5 French sheath was placed in the left common femoral arteries of Twins A and B. The left internal carotid artery (ICA) of Twin A and the right ICA of Twin B were both catheterized with a No. 5 French catheter. Angiograms were obtained using various projections after both single and simultaneous ICA contrast injections. Angiograms of the posterior circulation had been performed previously in another hospital and showed no evidence of cerebellar cross-flow.

Abnormal Findings

Skull. The CT scans, which better demonstrate osseous anomalies, revealed a partially common cranium (Fig. 1). Although the frontal bones were completely developed in each twin, the temporal and parietal bones were found to be absent. The occipital bones were only half developed but connected via a separate sutureal bone.
bone in the midline, resulting in a common posterior fossa (Fig. 1 lower). This kind of fusion is classified as a total parietooccipitotemporal craniopagus.

There was also a severe deformity of the skull base, with both shortening and posterior rotation of the medial skull base structures. However, all elements of the bony skull base existed in each twin, including all skull base foramina. The osseous fusion included the mastoid processes, which resulted in an overall fusion length of 17 cm.

**Dura Mater.** Dura mater separating the two brains was visible only in cranial areas above the level of the ventricular roofs; the larger proportion of the two brains was thus not separated by dura. Corresponding to this dural hypoplasia between the two brains, the falx cerebri was also absent in each. Whereas the lateral parts of the tentorium could be seen clearly in each twin on coronal T₁-weighted MR images, the medial parts of the tentorium were absent in each, resulting in a common tentorial notch (Fig. 2 upper).

**Cerebrum and Cerebellum.** All parts of the brain existed in both women and could be identified on the MR images. The frontal lobes were well developed. However, the parietal and temporal lobes of the adjacent hemispheres appeared severely deformed and hypoplastic, producing a triangular configuration of each hemisphere in the axial plane (Fig. 3).

The temporal lobes were most severely deformed, with cystic dilation of the temporal horn and rotation of the sylvian fissures from the transverse to the coronal plane, which was combined with rotation of the entire temporal lobes in the coronal plane (Fig. 2 lower). Whereas the adjacent frontal and parietal lobes were clearly separated by the subarachnoid space, there was gyral interdigitation of the temporal lobes, with a reduction of the subarachnoid space in between (Figs. 2 lower and 3).

**Veins.** On MR angiography and even on spin-echo images the proximity of the two superior sagittal sinuses, which begin at the level of the venous confluence, was evident. On inspection of transverse T₁- and T₂-weighted images, a common sagittal sinus and venous confluence

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**Fig. 3.** Axial T₂-weighted MR image obtained at the level of the lateral ventricles showing the interdigitation of the temporal lobes (small arrows), the triangular shape of the adjacent hemispheres, and the common venous confluence (venous star, large arrow).

**Fig. 4.** Maximal intensity projection reconstruction of a phase-contrast angiography sequence of the veins in the coronal view showing the common sagittal sinus extending from the common venous confluence to the medial transverse sinuses. The flow phenomenon within the common sinus simulates an intravascular septum (arrow).

**Fig. 5.** Upper Left and Right: Simultaneous DS angiograms of both medial ICAs showing a large arterial communication between the left MCA of Twin A and the right MCA bifurcation of Twin B (upper left, arrow). The venous phase (upper right) clearly shows the common sagittal sinus of both twins and significant venous drainage via the right insular vein and cavernous sinus of Twin B (arrows). Lower: Digital subtraction angiograms of the right ICA of Twin B showing the large arterial connection (arrow) between both MCAs in the early arterial phase (lower left). Multiple pial connections (arrowheads) are seen from the right MCA branches of Twin B to the left temporal lobe of Twin A in the late arterial phase (lower right).
were suspected, the latter having the appearance of a venous star (Fig. 3). Findings on MR angiography supported the diagnosis of a common sagittal sinus, but in the 3-D reconstruction of the angiographic sequence there seemed to be a septum remaining within this common sinus (Fig. 4). Ultimately, using angiographic studies we proved the existence of a common sagittal sinus that extended from the common venous confluence to the partition into two medial transverse sinuses, and we ruled out any septal structure within the common sinus (Fig. 5 upper right). We also demonstrated on angiographic studies that the venous drainage of the left temporal lobe of Twin A was directed into the insular veins of Twin B and from there to the cavernous sinus of Twin B (Figs. 5 upper right and 6). However, only the source images of MR angiography showed the existence in each twin of a hypoplastic lateral transverse sinus originating from the common sagittal sinus.

Arteries. Carotid studies revealed a large arterial communication between the proximal left middle cerebral artery (MCA) of Twin A and the MCA bifurcation of Twin B, with a spontaneous flow from left to right (Fig. 5 upper and lower left). Angiographic studies of the left carotid artery in Twin A showed a reduction in the capillary blush of the midhemispheric cortex. This area was vascularized by small connecting arteries stemming from the right MCA branches of Twin B and resulting in an important arterial cross-flow in the blood supply to the left temporal lobe in Twin A (Fig. 5 lower right).

Discussion

Cranially conjoined twins are regarded as one of the rarest human malformations. Because of the high mortality rate of babies born with this anomaly and early attempts at a surgical separation in those who survive, the reports of radiological imaging of craniopagus are scarce.1,3,4,6-9

It is believed that conjoined twins develop as a result of incomplete fission of a single developing embryo and that this abnormality is established at the end of the 2nd week of gestation.2 The fusion in total craniopagus is probably based on the absence of an area of cutaneous ectoderm (scalp) and ectomeninx (skull and dura mater). At the margins of this defective scalp, skull and dura mater are fused.2 Failure of the dura to develop is not limited to the area of fusion but also extends to the falx cerebri and the tentorium cerebelli. This was clearly demonstrated on MR imaging in the twins presented here. As for the possibility of surgical separation, the state of the superior sagittal sinus is more important. In cases of total vertical craniopagus the superior sagittal sinus fails to develop. Instead, a common circumferential sinus joins the lateral sinuses of the twins anteriorly and posteriorly. In these twins, who were examples of a total parietooccipitotemporal craniopagus, the superior sagittal sinus is normally developed in the frontal and central portion. The common venous confluence is shifted cranial to the level of the lateral ventricles. From the confluence to the medial transverse sinuses, the twins in this report had the same venous drainage via a common sagittal sinus. This common sagittal sinus is the most important neuroradiological finding in terms of the possibility of surgical separation of these twins and has always been described in total craniopagus. In comparison to the vertical type of fusion there is only a partial fusion of the sagittal sinus in the twins presented here.3,6-8

Aside from the common sagittal sinus, the gyral interdigitation of the temporal lobes is of particular interest. Whereas the adjacent gyri that do not show interdigitation have a normal pia-arachnoid covering, it has been found intraoperatively that in interdigitated areas these meningeal layers appear to be absent, resulting in cerebral continuity.7 Arterial and venous connections between cranially conjoined twins have been described in these areas and were also seen in the present case: DS angiography demonstrated a significant arterial and venous cross-flow from Twin B to the temporal lobe of Twin A. The large communicating artery from the MCA of Twin A to the MCA bifurcation of Twin B was suspected to be the main temporal branch of the left MCA of Twin A. Gyral interdigitation and cross-flow of the temporal lobes lead us to presume that the leptomeninges are absent in this area.

Whereas the absence of scalp, skull, and dura mater at the site of fusion is the primary developmental failure, all the remaining abnormalities, such as deformity of the skull base, cerebral deformity, and cerebral displacement, result from mechanical pressure. The growth of one brain and skull base interferes with that of the other. These forces cause severe deformity, resulting in a triangular shape of the adjacent hemispheres and rotation and shortening of the adjacent portions of the skull base.

The quality of current imaging methods, especially that of MR imaging, offers the opportunity for a detailed clarification of the pathoanatomy in rare human abnormalities such as craniopagus. Although MR imaging shows superbly the degree of development and the deformity of brain and dura mater, CT scanning is better in showing the bone structures of interest. Magnetic resonance angiography helps in evaluating the cerebral circulation but must be complemented by selective angiographic studies whenever small vessels and arterial connections are to be studied in detail.8
References


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Address reprint requests to: Olav Jansen, M.D., Department of Neuroradiology, University of Heidelberg Medical School, Im Neuenheimer Feld 400, D-69120 Heidelberg, Germany. email: Olav_Jansen@krzmail.ukl.uni-heidelberg.de.