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Case report

Scintigraphic evaluation of craniopagus twins

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Abstract. Craniopagus twinning is a rare congenital abnormality, occurring at a frequency of 4–6 per 10 million births. A case is reported in which separation was successful for both twins. The importance of pre-operative radionuclide assessment of crucial organ function (liver, kidneys, heart, brain) and crossed circulation is stressed. The scintigraphic results were in keeping with radiographic, operative, and clinical findings. The routine use of radionuclide studies in the investigation of conjoined twins is recommended in order to delineate individual organ function, degree of fusion and measurement of cross-circulation.

Conjoined twins is an extremely rare congenital abnormality, occurring in one in 50 000 births [1, 2]. Craniopagus varies from 2–6% of conjoined twins [3, 4]. Surgical separation and subsequent survival depends largely on the extent to which organs and vasculature are shared. Thorough pre-operative evaluation is therefore necessary.

Radionuclides have been used previously in the pre-operative assessment of conjoined twins. Investigations have included liver, kidney and dynamic imaging of the heart [5]. To our knowledge functional brain imaging for craniopagus twins has not been reported.

In this case, the scintigraphic investigations including functional brain imaging were performed on a set of craniopagus twins. Scintigraphic findings correlated well with intraoperative and clinical findings, and were helpful in planning the surgical procedure.

Case report

J and L, male craniopagus twins, were delivered by an elective Caesarean section at 38 weeks gestation. Craniopagus twinning had been diagnosed by ultrasound at 28 weeks gestation. The babies were aged 8 months on admission and their combined weight was 14.7 kg, with a total length of 132 cm. The twins were joined by the occipital region of their skulls and were facing in opposite directions. The circumference of the junction was 24 cm. Neurological examination showed no apparent abnormalities, with normal reflexes, good motility and tonus in all limbs. The babies moved and cried independently, and had independent autonomic and sleep rhythms. Full blood count, urea, electrolytes, blood glucose and thyroid function tests were normal, with only minimal variations between the twins.

Kidney function was evaluated in both twins using $^{99}Tc^{m}$ diethylene triamine pentaacetic acid (DTPA) renography. Liver function, brain function and cross-circulation were measured following the injection of $^{99}Tc^{m}$ hexamethylpropylene amine oxime (HMPAO).

On day 1, 30 MBq of $^{99}Tc^{m}$ DTPA was injected into an antecubital vein of twin L, while a dynamic acquisition of 60 frames per second was in progress, with the detector positioned posterior to the patient. The gamma camera was equipped with low energy all purpose (LEAP) collimator using a 64×64 matrix. The field of view included the kidneys, large blood vessels, spleen, ureter and bladder. The first acquisition phase was immediately followed by a second phase, imaging kidney function. The perfusion frames were combined to 15 frames of 4 s and those of function were combined to 15 frames of 2 min. Frames were displayed and assessed for perfusion, uptake and excretion and a renogram was generated. On day 6 (after the first study) a similar renogram was completed on twin J.

On day 11, 200 MBq of $^{99}Tc^{m}$ HMPAO was injected into the left-hand vein of twin J. The progression of the tracer was followed from the heart of twin J, to the heart of twin L, and the peak to peak and mean transit times for both left ventricles were calculated. Nuclear angiography was followed by a whole body scan over both twins to determine the relative sharing of the blood circulation between them. 30 min post-injection the liver was imaged and excretion of $^{99}Tc^{m}$ HMPAO into the bowel was confirmed.

Following this, brain single photon emission

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computed tomography (SPECT) was completed using single-headed rotating gamma camera equipped with a low energy, high resolution collimator. Data were collected from 64 projections in the 140 keV photopeak (20% window) over 360° in 64×64 matrices, with an acquisition time of 30 s per view. A zoom factor of 1.6 was used. Orthogonal transverse, coronal and sagittal planes were generated by filtered backprojection using a Wiener filter, followed by orbitomeatal reorientation of the reconstructed volume. The above procedure was repeated on twin L on day 14.

Results

Renal scintigraphy

Both kidneys were of normal (size), shape and position and presented with normal perfusion and concentration of the tracer in both twins. The excretion of tracer in the left kidney is delayed probably due to position during acquisition (patient lying on the right lateral side) (Figure 1).

Nuclear angiography and whole body quantification

The peak to peak time that the tracer bolus took to travel from the heart of L to the heart of J was 12 s after injecting twin L. The connection between twin L and J appeared to be of a venous nature through the posterior part of the central sinus in both twins. After injecting twin J, the bolus took 11 s to travel from the heart of J to the heart of L. Once again the connection appeared to be of a venous nature in the posterior part of their sinuses. The twin:twin whole body semiquantification of $^{99}Tc^{m}$ DTPA was 50%:50%. The distribution of $^{99}Tc^{m}$ HMPAO was 70%:30% in favour of J, when J was injected, and 70%:30% in favour of L, when L was injected (Figure 2). It was concluded that there was a fast and essentially equal shunting of blood through the sinus of L to J and of J to L with neither of the systemic circulations dominating.

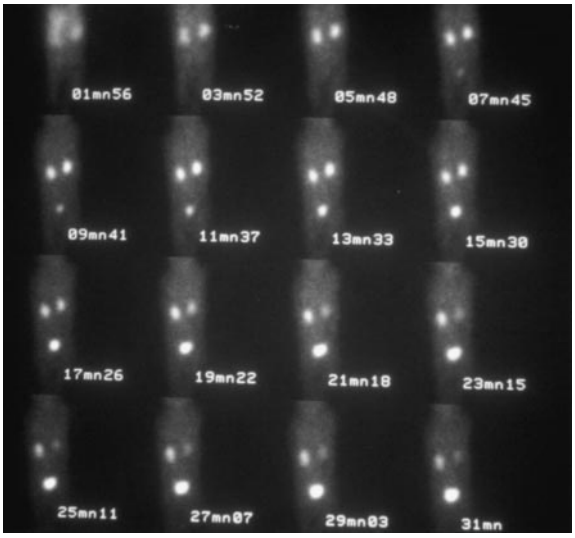

Figure 1. Renal scintigraphy of twin L over 30 min using $^{99}Tc^{m}$ DTPA.

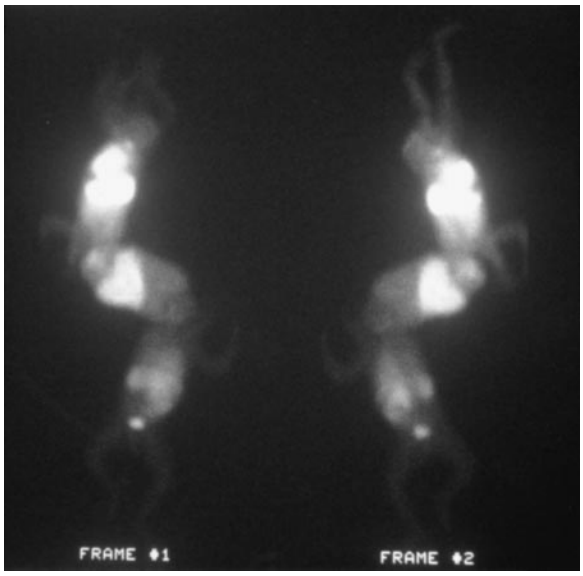

Figure 2. Whole body distribution of $^{99}Tc^{m}$ HMPAO in both twins after twin J was injected.

Liver scintigraphy

The liver was of normal size, shape and position in both twins; with normal tracer excretion into the gut (Figure 3).

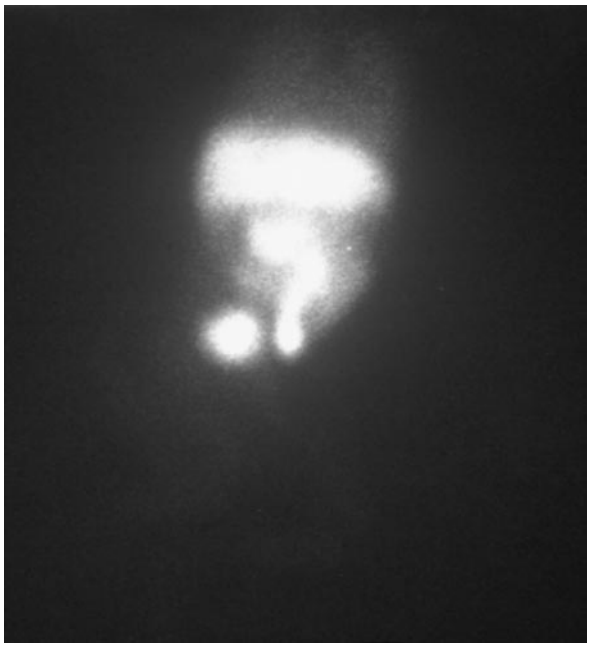

Figure 3. The liver of twin J 30 min post-injection of $^{99}Tc^{m}$ HMPAO.

Brain scintigraphy

There was an area of abnormally increased perfusion, superior to the right hemisphere of L and superior to left hemisphere of J. These findings were also noted on the 3D display (Figure 4) and interpreted as anatomical changes to accommodate the normal and separate brains in the shared cranium, as confirmed on CT and MRI (Figure 5).

Discussion

The conjoining of twins is the result of the incomplete fission of a single embryo, rather than the fusion of two. The anomalous conjoining occurs in the second week of development, due to incomplete division of the embryonic disc of the blastocyst [6, 7]. In the case of craniopagus twins, the congenital defect results in an area of absent cranial cutaneous ectoderm (scalp) and ectomeninx (skull and dura) of varying severity and extent in the region where the developing telencephalic vesicles meet [1]. Craniopagus twinning can be classified by two methods.

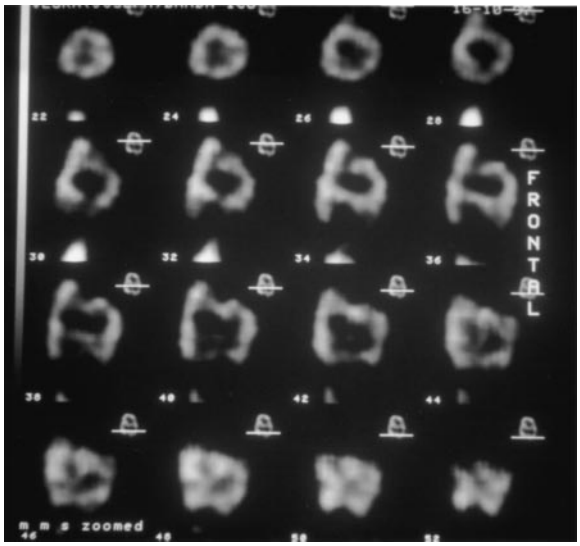

Figure 4. Coronal slices of $^{99}Tc^{m}$ HMPAO brain SPECT of twin L.

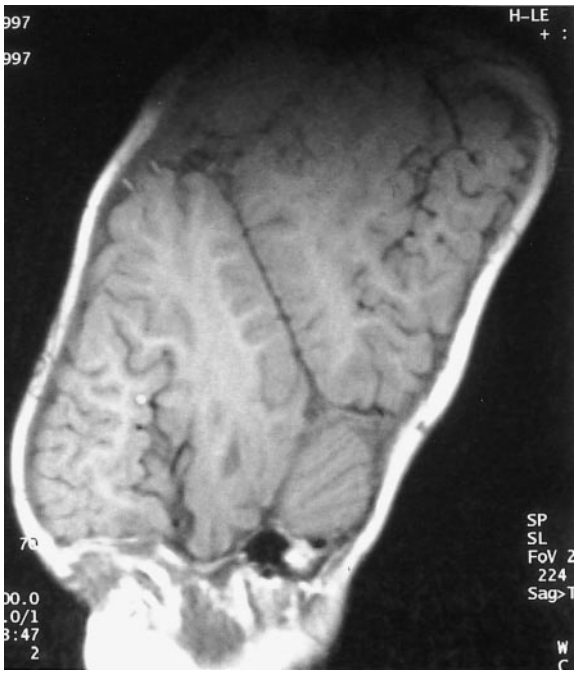

Figure 5. Coronal section of the MRI scan showing the abnormal anatomy of the brains.

The first is mainly topographical and considers only the site of conjunction. Thus, parietal, frontal, and occipital conjunctions have been described, the parietal being the most frequent [8]. The second classification stresses the extension and intimacy of union. Craniopagus is defined as partial when the surface common to the two heads is of limited extent, as in our case, and fusion is only superficial with partial involvement of the brain coverings. Bony and dural septa may partially or completely separate the two cranial cavities [9]. Cerebral deformities and venous cross-circulation are usually not prominent in the partial variety [7, 10]. Conjunction is more extensive and intimate in the total variety, in which the wide communication between the two cranial cavities may resemble a single skull containing two brains [11]. In these cases the development defect of the cranial ectomeninges is practically total. This defect may be responsible for the lack of development of the falx and of the superior sagittal sinus [6].

Although at present, CT and cerebral angiography are the fundamental diagnostic procedures for determining the possibility of separation, radionuclide studies can provide vital information which influences the final outcome.

CT shows the amplitude of the bone defect, morphology and mutual relationship of the two brains, as well as the presence of associated intracranial malformations. In this case, CT and MRI showed no intracranial defects. Cerebral angiography provides invaluable information on the cerebrovascular architecture of craniopagus twins [7, 12]. From a surgical viewpoint, the venographic findings are very important. In this case, the venous system was well developed and independent in each brain, and the two superior sagittal sinuses were separate, although more developed on twin J.

Nuclear scintigraphy provided indispensable functional information on the kidneys, liver and brain, apart from the semi-quantification of the circulation. The biochemical values of one twin can compensate for the other twin and thus appear as normal. Anatomical imaging modalities do not show the function of individual organs. The advantage of scintigraphy is that both anatomy and physiology can be investigated. Vital organ function can be determined without the interference of cross circulation and this fundamental information can be

utilized when planning the separation of the twins. $^{99}Tc^{m}$ HMPAO has excellent uptake primarily in the grey matter of the brain on first pass, prolonged retention of the activity in the brain, and slow regional redistribution [13]. In comparison, the extraction of $^{99}Tc^{m}$ DTPA by kidneys on first pass is not that efficient. It enters the extracellular fluid after intravenous administration and is removed from the circulation almost exclusively by glomerular filtration [14]. $^{99}Tc^{m}$ DTPA therefore remains in the bloodpool for longer than $^{99}Tc^{m}$ HMPAO. Both twins were shown to have good and independent function of the liver, kidneys, heart and brain with no dominant circulation of one twin to the other. The brain SPECT showed that the twins had separate brains with good cerebral perfusion and therefore good metabolism. The radionuclide studies correlated well with the intraoperative and postoperative findings.

Acknowledgments

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