

An anencephalic monocephalus diprosopus “headed twin”: postmortem and CT findings with emphasis on the cranial bones

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Monocephalus diprosopus is a form of conjoined twinning characterized by a single body, one unusual head and two faces or a spectrum of duplication of the craniofacial structures. Such cases have been mainly described according to postmortem pathologic examination.

This presented case is a 26-week-stillborn female fetus, with unusual facial appearance with four eyes, two mouths, two noses, two ears and a defective cranial vault. To our knowledge, a detailed computerized tomography (CT) examination of the aberrant facial and cranial bones of such a case has not been reported to date. In this reported case, we present an anencephalic monocephalus diprosopus “headed twin”, and describe the CT findings with emphasis on the cranial bones.

Key words: headed twin, monocephalus diprosopus, cranial computerized tomography CT, anencephaly, double tongue, dextrocardia, diaphragmal agenesis.

Monocephalus diprosopus is a form of conjoined twinning characterized by a single body, one unusual head and two faces or a spectrum of duplication of the craniofacial structures. This type of conjoined twinning is seen in less than 1% of cases. Such cases have been mainly described according to postmortem pathologic examination. The etiology of this anomaly is still obscure. In this presentation, we emphasize the computerized tomography (CT) findings (pre-autopsy) of the cranial and facial bones.

Case Report

We describe a 28-week stillborn fetus characterized by an unusual facial appearance with four eyes, two noses, two mouths, two ears, a short neck and a defective cranial vault. The shoulders were larger than normal (Fig. 1). The trunk and extremities were proportional and showed no other associated anomaly. This is the first child of a nonconsanguineous family. The mother was 18 and the father 20 years old. Prenatal sonography was not performed until 28 weeks of gestation. The family history was not contributory, but the mother underwent a

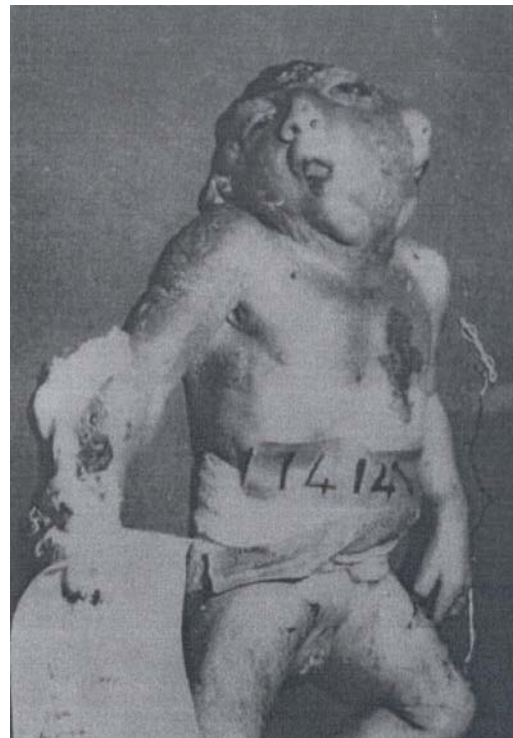


Fig. 1. Monocephalus diprosopus. Anencephalus with two faces and fused heads.

thyroid scan at two weeks' gestation. Postmortem chromosomal analysis from peripheral blood was normal (46 XX). The postmortem physical examination revealed an anencephalic fetus with facial appearance characterized by double eyes, noses and ears with short and wide neck. The weight was 130 g and length was 35 cm.

Computerized tomography was performed before the postmortem examination to study the cranial bones in detail. The CT scanogram of the whole body showed that the bones of the extremities and the trunk were normal and proportional (Fig. 2).



Fig. 2. CT scanogram of entire body. The bones of extremities and trunk are normal.

The facial bones were doubled and so badly malformed that identification of each bone was difficult. The two symmetrical mandibulae were separated by soft tissue. Each mandibula consisted of a double arch of bone with teeth arranged around the anterior margin and articulating laterally with the temporal bone (Fig. 3). The maxillary bones were doubled and symmetrical, but oriented obliquely with their long axis pointing laterally. The maxillary bone had lost its C shape and acquired an elongated narrow, inverse U shape, with alveolar processes arranged along the anterior border (Fig. 4).

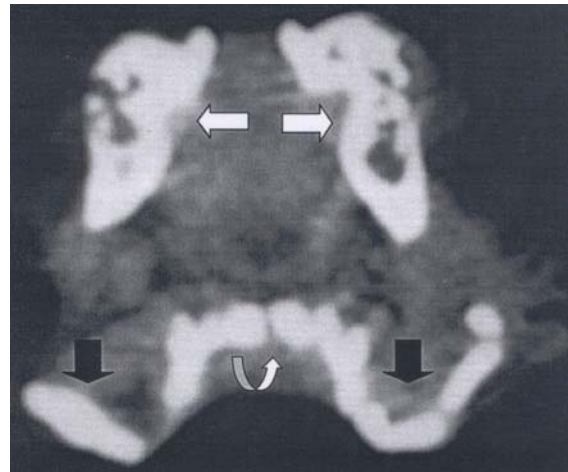


Fig. 3. Two mandibulae (white arrows) were symmetric and separated by soft tissue with the alveolar processes oriented anteriorly. Plate-like occipital squama (black arrows) with cervical spine bifida (curved arrow).



Fig. 4. Double maxillary bone (arrows) elongated in shape, pointing laterally and with anteriorly located alveolar processes.

The nasal cavities were like a thin canal containing some air. The double ethmoid bones were small and thin and the lateral ends articulated posteriorly with the presphenoid (Fig. 4).

The frontal bones were seen as double-bowed thin plates formatting the upper posterior border of the two eyes lying adjacent in the middle of the face. No parietal bone could be identified.

The sphenoid bone was single (Fig. 5). The postsphenoid was a big rectangular bone attached to the alisphenoid at the lateral border. The presphenoid was enlarged and wedge-shaped resembling a giant rostrum. The alisphenoid on both sides was thick, almost

oval-shaped. The orbitosphenoids could be identified as thin lamellae at both lateral ends of the alisphenoids. The pterygoid plates were round and thick. The foramen ovale and rotundum could not be distinguished.

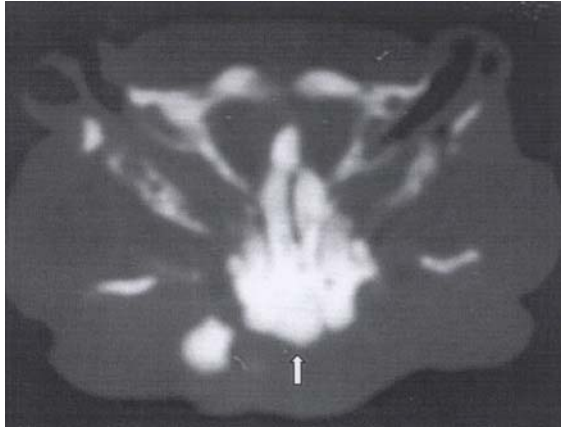


Fig. 5. The sphenoid bone (arrow): the presphenoid was enlarged and wedge-shaped. Both nasal cavities contained some air.

Two deformed and large petrous bones were situated at the usual skull-base location. The petrous bone had lost its pyramidal shape and become more rounded. The petrous apex was rotated almost 135° laterally. The internal auditory canal lay at the posterior-inferior border of the petrous bone. The external auditory canal was not discernable. The cochlea showed one and a half turns (Fig. 6). The

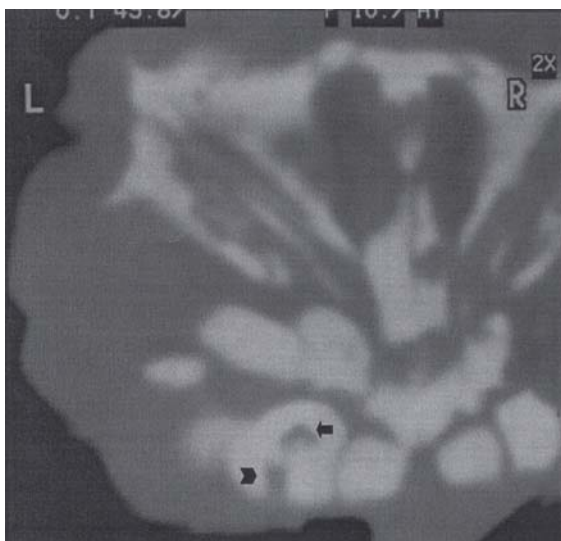


Fig. 6. Left petrous bone rotated with cochlea (arrow) and the internal acoustic canal (arrow-head) opening posteriorly.

vestibule was enlarged, and the semicircular canals were prominent (Fig. 7). The middle ear was large and deformed; there were a few calcifications which could represent the ossicle (Fig. 8). The temporal squama was seen as a linear small bone attached to the anterior-lateral border of the petrous bone. No mastoid bone could be identified.



Fig. 7. Right petrous bone with enlarged deformed vestibule (arrow) and prominent semicircular (arrowheads) canals.

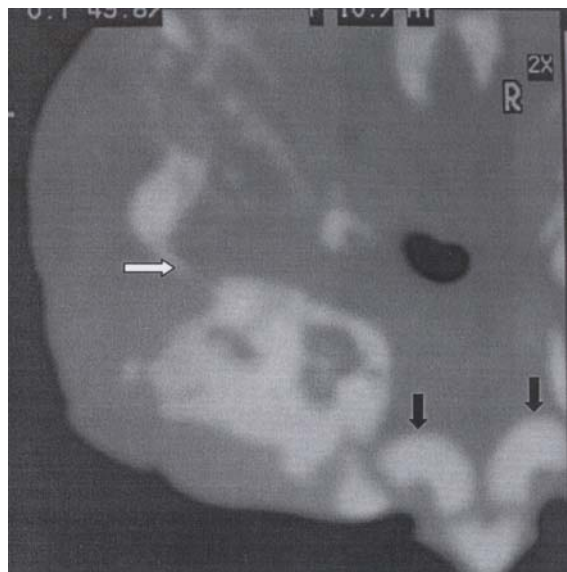


Fig. 8. The left petrous bone with enlarged vestibule and rudimentary temporal squama (white arrow). The inverted occipital condyles (black arrows) and inverted basis occipitalis.

The squamos part of the occipital bones was split. The squamos occipital bone on both sides was seen as a curved thin bony plate, lying free at the lateral end and medially lying close to the posterior arc of the 2nd cervical vertebra (Fig. 3). The occipital condyles were inverted and comma-shaped. Atriangular bone was found posterior to the condyles that might represent the ossification of the basis occipitalis, which was absent at the expected location.

A proper foramen magnum did not exist due to the disorganized occipital bone. The first four cervical vertebrae had wide posterior spina bifida of 2 cm (Fig. 3). The body and posterior elements consisted of multiple ossification centers, orderly arranged around the split spinal canal. The fifth and lower vertebrae up to the 2nd lumbar vertebra level were of a more normal shape with a narrower spina bifida. No cerebral, cerebellar or brainstem soft tissue could be identified. The two nasopharynges combined to form one pharynx and a single trachea could be followed. The thymus was extremely enlarged. Situs inversus totalis with right-sided heart and left-sided liver was noticed (Fig. 9). CT of the thorax and abdomen showed no other major pathology.

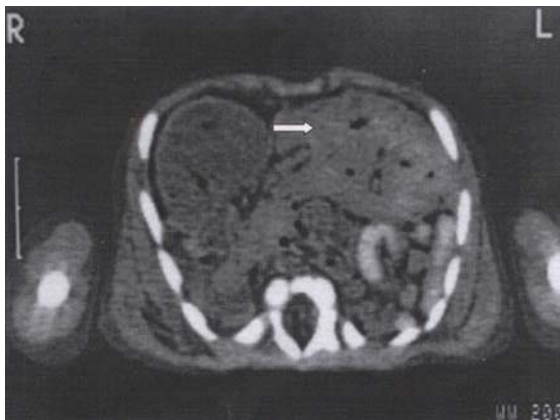


Fig. 9. Left-sided liver (arrow), right-sided spleen.

Postmortem examination showed single trachea, double tongue and dextrocardia. Left diaphragmal agenesis was noted.

Discussion

Monocephalus diprosopus is a rare form of conjoined twinning, and the incidence of this anomaly in association with anencephaly is very rare. One such case was described by Moerman in 1983¹; however, to our knowledge, a detailed CT examination of the aberrant facial and cranial bones has not yet been published. In our case, the shape of the doubled mandibular and maxillary bones was deformed and a correlation with normal bones was difficult. The sphenoid bone was atypical, yet similar to the sphenoid bone in anencephaly described by Virapongse in 1985^{2,3}. The occipital bone was dysplastic, showed atypical inverted occipital condyles and ossification of the basis occipitalis.

In conclusion, we believe that the detailed CT description and documentation, done prior to autopsy, may help to enlighten the anomaly of the cranial bones. As far as we know, this is the first detailed published report of detailed CT findings of an anencephalic headed twin.

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