Case Report

Diprosopus, craniorachischisis, arthrogryposis, and other associated anomalies in a stillborn lamb

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Congenital malformations with multiple anomalies have been described infrequently in the veterinary literature. A stillborn male crossbred lamb with diprosopus, craniorachischisis, and arthrogryposis was examined macroscopically and histopathologically in this study. The left head was smaller than the right head. Micrencephaly, agnathia, and a rudimentary tongue, which was adherent to the palate, were present in the left head. Micrencephaly, brachygnathia superior, and cleft palate were present in the right head. Cerebellar agenesis and spinal cord hypoplasia were observed. The cerebrums and the spinal cord were covered with a tapering membranous structure. Neural and dermal tissues were noted to intervene upon microscopic examination of this structure. Disorganization of neurons was observed in both cerebrums, though it was more severe in the left one. This case demonstrates many congenital defects occurring together in a lamb.

Keywords: arthrogryposis, craniorachischisis, diprosopus, lamb

Congenital malformations are structural and functional abnormalities present at birth. They can affect a single structure or function, parts of various systems, or an entire system [14]. The incidence of these abnormalities is reported to vary between 0.2% and 2% in lambs [3]. Congenital malformations with multiple anomalies in a single animal have been reported to represent 32.9% of the total congenital malformations in lambs [2]. In this study, a lamb with diprosopus, craniorachischisis, arthrogryposis, and other associated malformations was examined grossly and histopathologically.

A stillborn male crossbred lamb with various malformations was presented for systemic necropsy to the Department of Pathology, Faculty of Veterinary Medicine, Kafkas University, Turkey. The mother, a 2-year-old in its first parity, was brought to the Department of Reproduction with a complaint of dystocia, and she gave birth to the malformed lamb after a full term pregnancy. On necropsy, tissue samples were collected, fixed in 10% neutral buffered formalin, processed routinely, sectioned at 5-μm thickness, and stained with hematoxylin, eosin, and Masson’s trichrome. In addition, to examine the details of the bones, the soft tissues were macerated in a 30% potassium hydroxide solution, and the lamb was radiographed.

There were two partially fused heads (Fig. 1A). The skin extending from right behind the heads to the lumbar level was missing, and the vertebral column was exposed. The brains were covered with a membranous tapering structure that continued through the hypoplastic spinal cord, which was also exposed (Fig. 1B). The right head was bigger than the left one. Each head had two eyes, two ears, a mouth, and a nose. The eyes of the right head were not aligned; the right eye was lower, and both were protruding from their orbits. Brachygnathia of the upper jaw was observed in the right head. The tongue was comparatively shorter than the normal. Cleft palate was present. The angulus mandibulae was larger in size than normal. The os hyoideum, pharynx, larynx, and esophagus were present. No lower jaw was noted in the left head. However, the presumptive location of the lower jaw was covered with skin (Fig. 1C). Both maxillae were deviated to the median. There was a rudimentary tongue that was adherent to the palate (Fig. 1D). The os hyoideum, pharynx, and larynx were absent, and there was no opening to the esophagus. The eyes of the left head were not aligned; the left eye was lower, and both eyes were protruding from the orbits, similar to the right head.

The concha nasalis dorsalis was absent in both heads, and the concha nasalis media and concha nasalis ventralis were approximately the same size. The two heads were joined to each other at the os occipitale, os temporale, and os
Fig. 1. (A) Frontal view of the diprosopic lamb. (B) Cranio-rachischisis. A tapering membranous structure covering the spinal cord (arrows). (C) Anognathia in the left head. Skin covering the presumptive location of the lower jaw (arrow). (D) A rudimentary tongue, which is adherent to the palate, in the left head (arrow). (E) Dorsal view of the lamb’s vertebral column. Severe deformation in the vertebral column. Scoliosis in the thoracal vertebrae (arrow). (F) Lateral radiograph of the lamb. Kyphosis in the cervical, thoracal, and lumbar vertebrae (arrows). (G) Microscopic view of disorganization in the left brain. Fibrous tissue in the brain (arrow). Masson’s trichrome. Scale bar = 56 μm. (H) Intervention of the neural (N) and dermal (D) tissues under the membranous structure. Masson’s trichrome. Scale bar = 140 μm.

The whole parietal bones and squama occipitalis of the occipital bones and foramen magnum were absent, and hence both brains were exposed caudally. All vertebrae from the cervical to the sacral spine were deformed. The discus intervertebrae were absent. The joining of the corpus vertebrae was chondroitic. Arcus vertebrae were absent at the tail, and hence the vertebral canal was not observed, except in the caudal region. The processus transversales were absent. Since the cervical vertebrae were severely deformed, the atlas and axis could not be distinguished. Therefore, the typical atlanto-occipital and atlanto-axial joints were undetermined. There were five rib-like bones in each side located on the dorso-lateralis of the thoracal vertebrae. The vertebral canal was present at the caudal level. Scoliosis was present in the thoracal vertebrae (Fig. 1E). There was kyphosis at the cervical, thoracal, and lumbar levels (Fig. 1F). The ossa coxae were close to each other, and the pelvic cavity was narrowed.

The right brain weighed 28.1 g. A pair of olfactory, optic, and oculomotor nerves, a sella turcica, and a hypophysis were present. The right brain bowed slightly to the median in accordance with the shape of the cranium. The left brain weighed 2.4 g. Olfactory, optic, and oculomotor nerves were present in the left brain. However, the sella turcica and hypophysis were absent. A hypoplastic medulla oblongata was present, and the cerebella were absent in both heads.

Arthrogryposis flexio was observed in both articulations metacarpophalangeae and the right articulations metatarsophalangeae. There was a hemorrhagic substance in the thoracal cavity. Petechiae were also present on the surfaces of the lungs and heart.

Microscopically, disorganized cerebral structures, degeneration and necrosis of neurons, hyperemia, and extravasal erythrocytes were observed in both brains. Disorganization was more severe in the smaller brain (Fig. 1G). On microscopic sections taken from the membranous structure, a tapering narrow band of epidermis was noted; right under it was a dermis composed of fibroblasts, fibrocytes, collagen, and lymphocytes. No hair follicles or sweat or sebaceous glands were noted at this location. Where the epidermis ended in this membranous structure, only dermis was visible. Dermal and neural tissues were intervening, and in some places, neural tissue islands were observed in this membranous structure (Fig. 1H).

Congenital duplications have been a matter of interest for centuries. An incomplete division of the zygote at a considerably late stage of embryonic development is considered to be the reason for congenital duplication [14]. These malformations can appear as a graded series from a slight duplication to almost complete separation of the twins, and hence can be classified as attached, free symmetrical and attached, or free asymmetrical twins [7,14]. Cranial duplications can be either diprosopus or dicephalus [7,16]. While diprosopus is characterized by fusion of the craniofacial structures of the two heads [5,11,13,15], partial duplication of the spine with the presence of two heads is described as dicephalus [8,10,12]. We regarded this case as diprosopus because of the fusion of the two heads at the occipital level. In most reported diprosopic animals, the two heads and the features of them have more or less resembled each other [11,13,15,17,18]. However, the two heads in this lamb were very different, not only in size, but also in anatomic structure.

Cleft palate is one of the most common anomalies, and it is commonly associated with diprosopus [4,9,19]. We also observed cleft palate in the right head of this diprosopic sphenoides.
lamb. Among the other defects, agnathia in the left head and brachygna-thia superior in the right head are noteworthy in estimating the developmental difference of the two heads.

In the present study, the weights of the right and left brains were recorded as 28.1 g and 2.4 g, respectively, which were below the normal range of 58-70 g [6]. Therefore, both brains were described as micrencephalic. It has been reported that there is generally a fused cerebellum [4,15,17-19] or two cerebella [5,11] present in diprosopus. However, cerebellar agenesis was observed in the present diprosopic lamb.

Spina bifida is a broad term used to describe neural tube defects characterized by a failure in closure of the vertebral arches. The most severe form of this malformation is called rachischisis, which is characterized by an open spine [14]. If rachischisis combines with a closure defect of the cranium, it is called craniorachischisis [1]. We have also described a case of diprosopus with craniorachischisis in a lamb.

The etiology of most congenital malformations is unknown, simply because of the complexity of the mechanisms leading to the formation of an abnormality. Genetic and environmental factors, or their interaction, have been proposed as the most common causes of congenital abnormalities [3]. Whatever the cause of the congenital defects, the countless varieties of a certain anomaly from animal to animal might be explained by the degree and time course of the effects of several etiologic factors of presumed lesser importance. In the current investigation, since we were unable to obtain the pedigree and history of the mother, no etiologic cause or causes could be ascertained.

In this study, we described the gross and histopathologic findings in a diprosopic conjoined twin lamb with craniorachischisis and arthrogryposis. The importance of these malformations, in general, lies in the embryologic development of the fetus and the particular mechanisms that relate to such significant changes during organogenesis. The precise etiology of these malformations is still largely unknown and requires further investigation.

References

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