Kafkas Univ Vet Fak Derg
17 (2): 333-336, 2011
DOI:10.9775/kvfd.2010.3078

CASE REPORT

A Rare Case of Diprosopus, Tetraophthalmus and Meningoencephalocele in A Lamb

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Makale Kodu (Article Code): KVFD-2010-3078

Summary

A female, dystocic, two-headed dead lamb from a 3-year old Sakiz sheep is described. The two faces of the foetus fused at occipital, partly parietal and temporal bones on a single trunk. On each face there were two eyes, one ear and one normally developed oral cavity with only one laryngopharynx and oesophagus. A flaccid, hyperemic, membrane-like tissue protruded from the defective frontal and parietal bones. There were no other skeletal abnormalities but incomplete costal articulation. Microscopic examination of the membranous tissue revealed mostly autolytic neurons. Since diprosopus cases with defective skull development and meningoencephalocele are rare events, this case was evaluated for presentation.

Keywords: *Diprosopus, Lamb, Meningoencephalocele, Tetraophthalmus*

Bir Kuzuda Nadir Rastlanan Diprosopus, Tetraophthalmus ve Meningoensefalosel Olgusu

Özet

Üç yaşlı Sakız ırkı bir koyundan güç doğumla doğan iki yüzlü, ölü, dişi bir kuzu tanımlandı. İki yüz tek bir gövde üzerine oturmuş, oksipital bölge ile kısmen parietal ve temporal bölgelerden kaynaşmıştı. Her iki yüzde iki göz, birer kulak, normal gelişmiş ağız boşluğu ancak tek bir laringofarinks ve özofagus vardı. Hiperemik ve gevşek zar benzeri bir doku hatalı şekillenmiş frontal ve parietal kemiklerden dışarı çıkmıştı. Kosta kondral eklemler gelişmemişti; başka bir iskelet anomalisi görülmedi. Zarsal dokunun mikroskobik muayenesinde çoğunlukla otolitik nöronlara rastlandı. Yetersiz kafatası gelişimi, meningoensefalosel ile beraber olan ve nadir karşılaşılan diprosopus olgusundan dolayı, bu olgunun sunumu yapıldı.

Anahtar sözcükler: *Diprosopus, Kuzu, Meningoensefalosel, Tetraophthalmus*

INTRODUCTION

Congenital defects are structural or functional abnormalities and can effect an isolated portion of a body system, entire system or parts of several systems and may cause obstetrical problems [1]. The majority of human and animal malformations are because of a multifactorial aetiology [2]. Congenital defects are caused by genetic or environmental factors (infectious diseases, viruses, drugs, poisonings, plants, mineral salts and vitamin (A, D, E) deficiency, hormonal factors and physical reasons) or by their interactions [3-5]. These factors cause oocyte structure, maturations and organogenesis alterations probably as a result of some metabolic or circulatory disturbances, on the basis of a not well -known mechanism, whereas traumas or compressions could damage the molecular disposition of cells, producing static alterations. Finally, genetic defects are pathological or pathophysiological results caused by mutant genes or chromosomal aberrations [4,5], the most known of which result from recessive genes [6], and are recognized only when they occur in characteristic intra-generational familial frequencies and intergenerational patterns [4]. Except for later-differentiating structures such as the cerebellum, palate and urogenital system, the fetus becomes increasingly resistant to teratogenic agents as it ages [1,3]. Conjoined twins, considered to be monozygotic twins imperfectly separated, from a graded

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series of slight duplication to almost separated individuals and occur extremely rarely in horses, occasionally in dogs and cats, and uncommonly in cattle, pigs and sheep [7,8]. The diprosopus, craniofacial duplication, abnormality, characterized by the presence of 2 faces with a single head on a single trunk, is one of the rarest craniofacial malformations and a rare form of conjoined twinning [9,10]. Diprosopus, have often been reported in calves and lambs [11-13], goats [14], cats [15] and rarely in foals [16]. Although several cases [2,3,12,17] of diprosopus have been reported in ewes, no literature about a diprosopus case with defective frontal and parietal bone development and meningo-encephalocele has been reported to the knowledge of the authors. Therefore, this case of diprosopic lamb was described with clinical and pathological findings.

CASE HISTORY

A 3-year-old pregnant Sakiz breed ewe was referred to the Department of Veterinary Obstetrics and Gynecology of the University of Uludag for dystocia with a history of extended term. A diprosopic lamb was detected as jammed in vaginal canal with head region; fetal fluid was discharged after vaginal examination. The dead female lamb was delivered by extraction force. The owner declared that the ewe had delivered an alive lamb in previous breeding seasons and that congenital anomalies had not been encountered for a long time in the herd. The lamb was sent to the Department of Veterinary Pathology of the University of Uludag for necropsy.

Gross examination of the lamb revealed a single trunk with two partially fused heads joined at the occipital, parietal and temporal regions *(Fig. 1)*. There were two eyes and one ear on two heads. Both faces were completely separated from each other but fused at caudal region. The bones forming the face elongated from behind of proccesus zygomaticus to temporal bones at cranio-dorsal, and was coalescing temporo-mandibular joint and part of corpus mandibulae at the cranio-dorsal. Symphisis mandibulae was completely shaped and joined with the arm of corpus mandibula. Although development of oral cavity and tongue was normal, only a single laryngo-pharynx and oesophagus was observed. Absence of calvarium, no skin development and protrusion of a hyperemic, thick and membrane covered brain like tissue were also noticed. Neither anatomical nor any structural organization was observed within the membranous structure *(Fig. 2)*. Microscopically mostly autolytic neuron like cells were seen *(Fig. 3)*. While all four legs developed

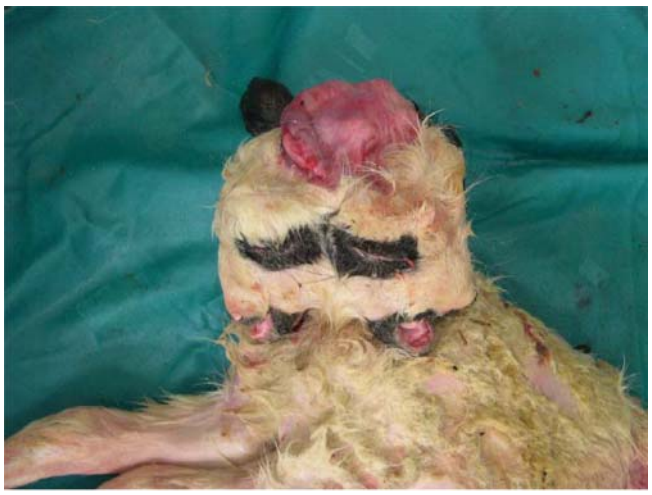

Fig 1. Cranial appearance of the partial duplication with two eyes and one ear on the each face

Şekil 1. Her bir yüz üzerinde bir kulak ve iki göz ile birlikte kısmi duplikasyonun cranial görünümü

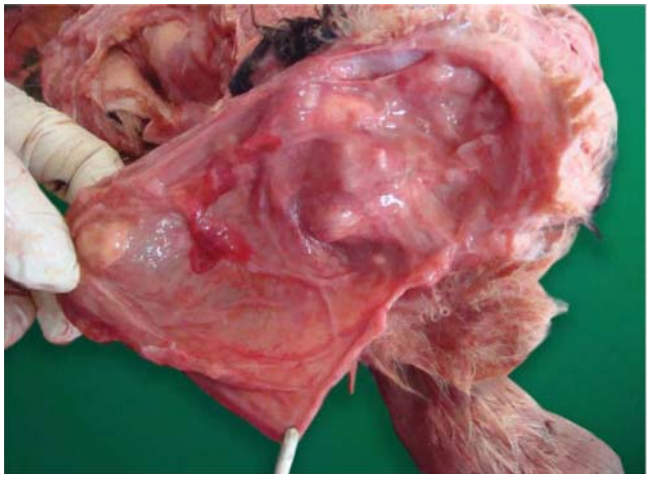

Fig 2. Following removal of the membrane like tissue, exposure of the cavum cranium without any normal anatomical development

Şekil 2. Membran benzeri dokunun uzaklaştırılmasını takiben normal anatomik gelişimin olmadığı cavum craiumun açılmış görünüşü

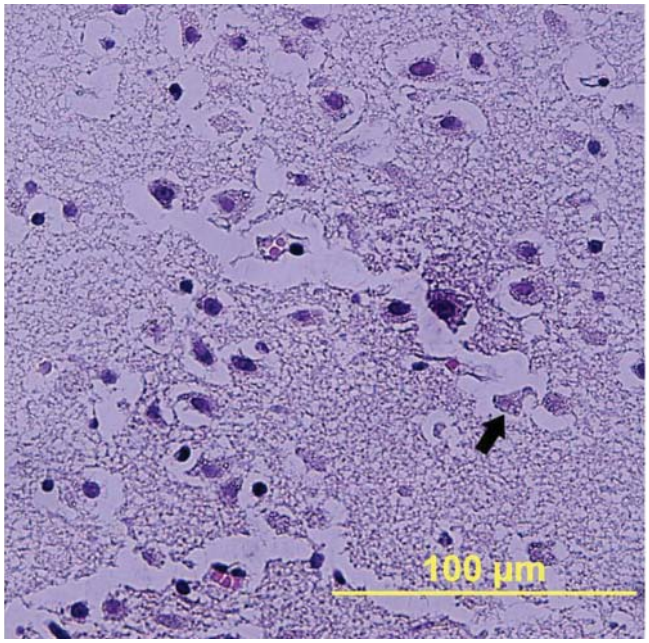

Fig 3. Microscopic appearance of severe autolysis in meningo-encephalocelic tissue, x400, H&E

Şekil 3. Meningoensefalosel şekillenmiş dokuda şiddetli otolizin mikroskobik görünümü, x400, H&E

normally, development of costochondral articulations were not complete. Thus, 7th to 10th ribs did not joint with the sternum and their ends were free and protruding outwards. Internal organs were in normal appearance, yet were severely autolytic.

DISCUSSION

Ewes have the highest incidence of craniofacial defects in mammals, including man [3]. Duplication of the caudal parts of the body is most frequent among the ewe conjoined twin anomalies. In a survey study, two of the 27 embryonic duplications were determined as diprosopus and Merino breed was the most common breed in which such defects occur [18]. Diprosopus has been reported most frequently in domestic animals such as calves and lambs, However, any race and gender predisposition have not reported so far [11,19]. In this case, although the stillborn animal as a result of dystocia, in previous reports that have been reported from time to time to offspring born alive [2,14]. Diprosopus and dicephalus should not be confused with, but in dicephalus has two heads and two different shaped neck [8,20,21]. It has been reported that face was shaped full or partialy to the pharynx and larynx and that next part the neck and body shape have been reported as a single columna vertebralis [13,18,22] in previous reports. Besides diprosopus is seen together with such as other anomalies; cleft palate, face and jaw anomalies, atresia ani, cryptorchism, arthrogryposis, craniorachischisis, heart and central nervous system disorders [11,13,18,22]. The os hyoideum, pharynx, larynx, and esophagus were present. It has been reported that there is generally a fused cerebellum [13,22] or two cerebella [2,7] present in diprosopus. However, cerebellar agenesis was observed in the present diprosopic lamb.

With its macroscopical findings, this is a case of diprosopus, tetraophthalmus. Since there is no cerebral tissue development within the sac like structure that fills the cranial cavity, it is also an encephalomeningocele.

While the observation of two cerebri and partially developed cerebellum formation was made in previous reports of diprosopus of sheep [7,12] and calve [23], in our case only a single sac that can be accepted as a representative of one cerebrum, which comprised of only a thin layer of cells, was observed; and no cerebellar development could be detected. This case with the presence of developmental abnormalities at the cranium, but not at the rest of the body can be considered as an example of support to the proposition that teratogens show their effect much more on the cranial part compared to the caudal part of the body [17].

Our case has some similarities with the case of a diprosopus of a kid in which there were two partially developed faces with the development of two eyes [24]. Yet the kid was born alive and development of the cerebrum, despite the hypoplastic cerebellum formation, was complete. In the case presented here, there is no development of cerebrum and cerebellum and developmental abnormality of the brain in such severity has not been reported in sheep or goats. However, in humans, there are reports of anencephaly cooccurring with diprosopus [9].

There is not sufficient research on the morphopathogenesis of diprosopus in both humans and animals. However, viruses and protozoan agents might have etiological role and circulatory disturbances resulting in hypoxia might be a cause. Due to the insufficient data, effects of such factors could not be evaluated in this presented report.

This case presented here with the absence of calvarium and protruding sac like structure instead of a skin cover and absence of anatomic development of cerebrum and cerebellum differs from the diprosopus cases that have been reported before.

REFERENCES

1. Long S: Abnormal development of the conceptus and its consequences. **In,** Noakes DE, Parkinson TJ, England GCW (Eds): Arthur's Veterinary Reproduction and Obstetrics. 8th ed., pp. 119-143, W.B. Saunders, London, 2001.

2. Mazzulo G, Germana A, De Vico G, Germana G: Diprosopiasis in a lamb. A case report. *Anat Histol Embryol*, 32, 60-62, 2003.

3. Dennis SM, Lelpold HW: Congenital and inherited defects in sheep. **In,** Morrow DA (Ed): Current Therapy in Theriogenology - 2. pp. 864-866, W.B. Saunders Company, Philadelpia, 1986.

4. Leipold HW, Dennis SM, Huston K: Embryonic duplications in cattle. *Cornell Vet*, 62, 572-580, 1972.

5. Kaçar C, Özcan K, Takçı İ, Gürbulak K, Özen H, Karaman M: Diprosopus, craniorachischisis, arthrogryposis, and other associated anomalies in a stillborn lamb. *J Vet Sci,* 9 (4): 429-431), 2008.

6. Dennis, SM, Leipold HW: Ovine congenital defects. *Vet Bull,* 49, 233-239, 1979.

7. Fischer KRS, Partlow GD, Walker AF: Clinical and anatomical observations of a two-headed lamb. *Anat Rec*, 24, 432-440, 1986.

8. Leipold HW, Dennis SM: Dicephalus in two calves. *Am J Vet Res*, 33, 421-423, 1972.

9. Al Muti Zaitoun A, Chang J, Booker M: Diprosopus (partially duplicated head) associated with anencephaly: A case report. *Path Res Pract*, 195 (1): 45-52, 1999.

10. Chervenak FA, Pinto MM, Heller CI, Norooz H: Obstetric significance of fetal craniofacial duplication. A case report. *J Reprod Med*, 30, 74-76, 1985.

11. Sarperstein G: Diprosopus in a hereford calf. *Vet Rec*, 108 (11): 234-235, 1981.

12. Kerr NJ: Diprosopus with multiple craniofacial, musculoskeletal, and cardiac defects in a purebred Suffolk lamb. *Can Vet J*, 48, 1074-1076, 2007.

13. Özcan K, Öztürkler Y, Takçı İ: Diprosopus in a cross bred calf. *Indian Vet J*, 82, 650-651, 2005.

14. Sönmez G, Özbilgin S, Serbest A, Mısırlıoğlu D: Bir oğlakta diprosopus olgusu. *Uludağ Univ Vet Fak Derg,* 3, 93-98, 1992.

15. Camon J, Ruberte J, Ordoneez G: Diprosopia in a cat. *J Vet Med*

(Series A), 37, 278-284, 1990.

16. Götz HJ: A case of diprosopus in a foal. *Tierarztl Prax*, 19, 82-83, 1991.

17. Shojaeı B, Derakhshanfar M, Oloumi M, Hashemnia S: Diprosopus, sipina bifida and kyphoscoliosis in a lamb - A case report. *Veterinarski Arhiv*, 76 (5): 461-469, 2006.

18. Dennis SM: Embriyonic duplications in sheeps. *Aust Vet J,* 51, 83-87, 1975.

19. Leipold HW, Dennis M: A diprosopus in newborn calves. *Cornell Vet*, 62, 282-288, 1972.

20. Güngör Ö, Yıldız S, Çolak A: Dicephalus in a calf. *Vet Hek Der Derg*, 75, 34-35, 2004.

21. Majeed MA, Hussain SS, Hur G: The structure of a doubleheaded buffalo calf (Dicephalus dipus dibrachius). *Vet Rec*, 88, 393-395, 1971.

22. Türkütanıt SS, Sağlam YS, Bozoğlu H: Diprosopus in a calf. *Istanbul Univ Vet Fak Derg*, 22, 253-256. 1996.

23. Özyıldız Z, Oral H, Kurt B, Yıldız S, Güngör Ö: İki melez buzağıda diprosopus. *Kafkas Univ Vet Fak Derg,* 15 (2): 305-308, 2009.

24. Mukaratirwa S, Sayi ST: Partial facial duplication (diprosopus) in a goat kid. *J S Afr Vet Assoc,* 77 (1): 42-44, 2006.