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Case Report

Dicephalus, Arnold-Chiari Malformation, Spinal Dysraphism and Other Associated Anomalies in a Newborn Holstein Calf

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Abstract: A case of dicephalus, Arnold-Chiari malformation, spinal dysraphism and other associated defects was described in a newborn Holstein calf. The heads were medially and caudally joined with temporal bones and one common flattened occipital bone respectively. Cranial cavities became narrow dorsoventrally and included elongation of the compressed brains, and caudal displacement and hypoplasia of cerebelli. In the spinal column, variable degrees of dysraphism such as cervico-thoracal diplomyelia with hydromyelia and syringomyelia, lumbar spina bifida with spinal cord agenesis in the region were observed. The calf had also arthrogryposis of hind limbs and cleft palate of both heads.

Key Words: Anomaly, Arnold-Chiari malformation, calf, dicephalus, spinal dysraphism.

Yeni Doğan Holstein Buzağıda Disefalus, Arnold-Chiari Malformasyonu, Spinal Disrafizm ve İlgili Diğer Anomaliler

Özet: Bu çalışmada yeni doğan Holstein bir buzağıda disefalus, Arnold-Chiari malformasyonu, spinal disrafizm ve diğer malformasyonlar belirlendi. Buzağıdaki her iki kafa medial ve kaudal olarak temporal kemikler ile yassılaşmış bir adet oksipital kemik aracılığı ile birleşmişti. Kranial boşluklar dorsoventral olarak daralmıştı ve bu nedenle her iki beyinin basınca uğradığı ve serebellumların hipoplazik durumda kaudale doğru yer değiştirdiği gözlendi. Medulla spinaliste değişen derecelerde hidromiyeli ve siringomiyeli ile birlikte serviko-torakal diplomiyeli ve lumbar spinal agenezisli spina bifida gözlendi. Buzağıda arka bacaklarda artrogripozis ve her iki kafada damak yarığı mevcuttu.

Anahtar Sözcükler: Anomali, Arnold-Chiari malformasyonu, buzağı, disefalus, spinal disrafizm.

Introduction

Dicephalus is a malformation of the head resulting from incomplete twinning in humans and animals (1). Description of dicephalus is based on its external features, but duplication always involves internal structures with various concurrent defects (2). Dicephalus, Arnold-Chiari malformation (ACM), spina bifida (SB), spinal dysraphism (SD), and cleft palate (CP) are individually common malformations in cattle (1,3-10). Combination of these malformations in cattle has been reported rarely in veterinary literature (11-14) and has not been reported in Turkey. The present case describes dicephalus with ACM, spinal dysraphism, lumbar spina bifida, cleft palate, and associated other defects in a Holstein calf.

Case History

The malformed calf, a female Holstein, was stillborn at normal pregnancy period after natural breeding, and was presented for postmortem examination. The calving was assisted and the 6-year-old dam had given birth to three normal calves previously. There was no additional maternal history and pedigree information about the sire.

The tissue samples from the malformed calf were fixed in 10% neutral buffered formalin and processed routinely, sectioned and stained with hematoxylin-eosin (HE). The selected sections were also stained with phosphotungustic acid hematoxylin (PTAH) stains. The calf and dam were not examined for viruses known inducing congenital nervous system anomalies.

Results and Discussion

The calf had a single body with partially duplicated heads that were of almost the same size and shape. Externally, there were four eyes, three outer ears with a common auricle on the fusion line and a pair of mandibles curved from right to left (Figure 1). The paired and unpaired bones of the skull were formed. Both heads were joined each other with the temporal bones which had the fused petrous parts. The joined parts had one common middle ear cavity and two chains of three ossicle-the malleus, incus, and stapes. In the free sides of the heads, one normal petrous part in the temporal bones was present. In the cranium, one flat and wide occipital bone was found. Therefore the distance between the condylus of the occipital bones was larger than normal. A dilated single common foramen magnum with large condylus occipitalis was present. Each head had one tongue that joined each other with their roots and they also had CP. The widened common larynx and pharynx opened into a single trachea and esophagus respectively. The cranial cavities became narrow dorsoventrally, and especially the occipital lobes of the left-brain were compressed by tentorium cerebelli and partially herniated into common foramen magnum because of the inadequate development of the cranial cavities. Both cerebellums were hypoplastic in variable degrees, and the right one showed herniation with a tongue-like process through the dilated common foramen magnum to the vertebral foramen of the atlas indicating ACM (Figure 2). Especially, the herniated cerebellum had shallow grooves and vermis was reduced in size. Histologically, the left and right cerebellar cortex was disorganized in variable degree. In the severely affected folia of the vermis, with especially herniated cerebellum, the granular and

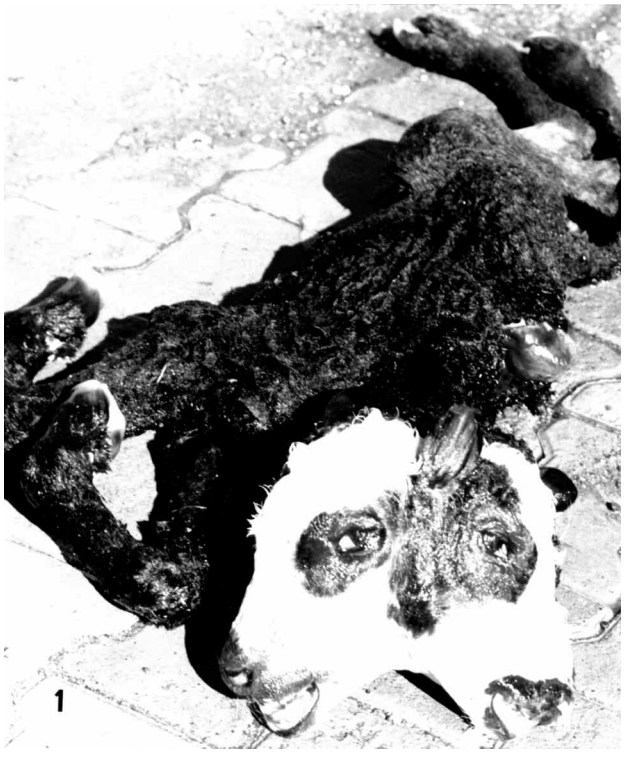

Figure 1. Dicephalic calf joined with temporal bones had four eyes and three ears. Note the lumbar spina bifida and arthrogryposis of hind limbs.

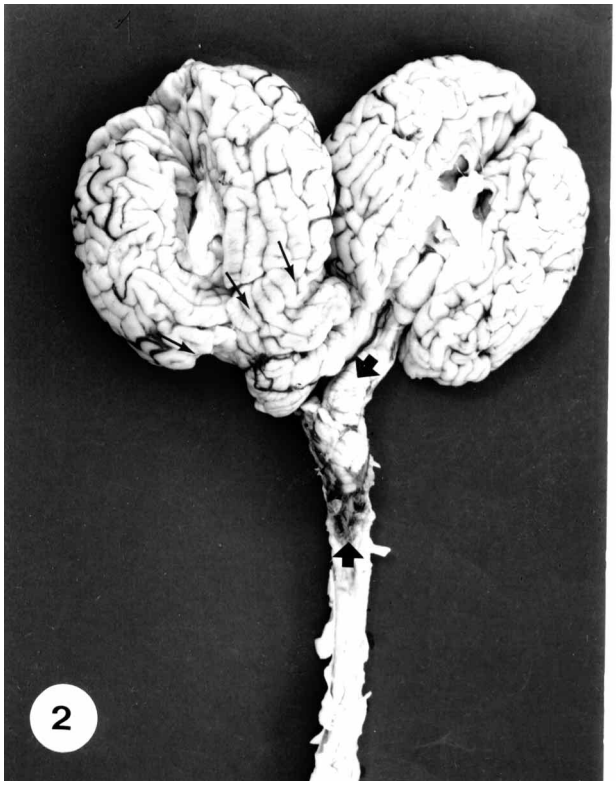

Figure 2. Arnold-Chiari malformation. The compression of the occipital lobe in the left-brain (thin arrows) and caudal displacement of both cerebelli, especially right one, with tongue-like process (thick arrow).

molecular layers were diminished in size and there was no folial pattern or division into lobes (Figure 3). Both brains were joined at the same level of the foramen magnum just caudal to pons with Y-shaped appearance (Figure 4). The remaining brain lobes were normal in appearance. Each head had one pituitary gland and the development, origin and number of the cranial nerves in the brains and spinal cord were normal, but they were not examined microscopically. The trunk had cervical kyphosis and lumbar SB characterized by agenesis of the dorsal arches of the vertebrae. In the region not covered with skin, spinal cord and associated tissues were absent. Both hind limbs were arthrogrypotic.

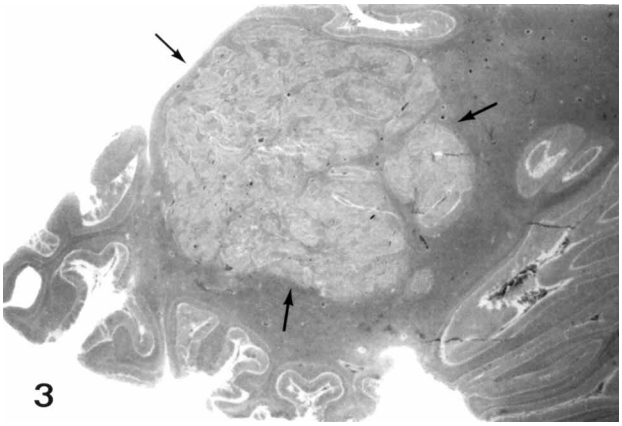

Figure 3. Herniated right cerebellum. Note the dysplastic area (thin arrows) in the herniated lobe of the vermis. HE, X 2,5.

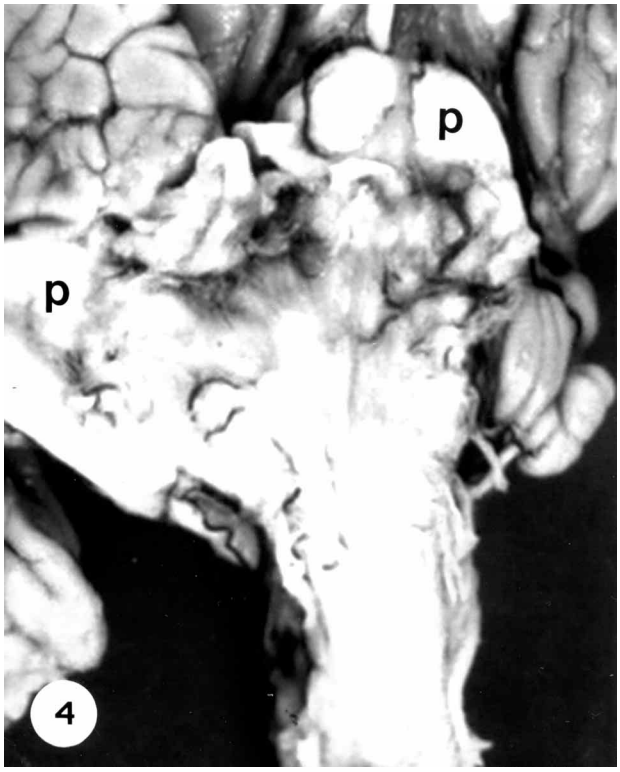

Figure 4. Ventral aspect of the brains joined at just caudal to pons (P) with Y-shaped appearance.

In the spinal cord, histologically, diplomyelia was observed with various degrees of dysraphism. Especially at the fusion level of both brains, two central canals and two ventral fissures with a common covering of leptomeninges and dura were confirmed duplication of the spinal cord (Figures 5, 6). Spinal dysraphism was characterized by variable degree of hydromyelia, with the partial pseudostratified ependymal cells lining the central canal and two syringomyelic canals. Direct communication

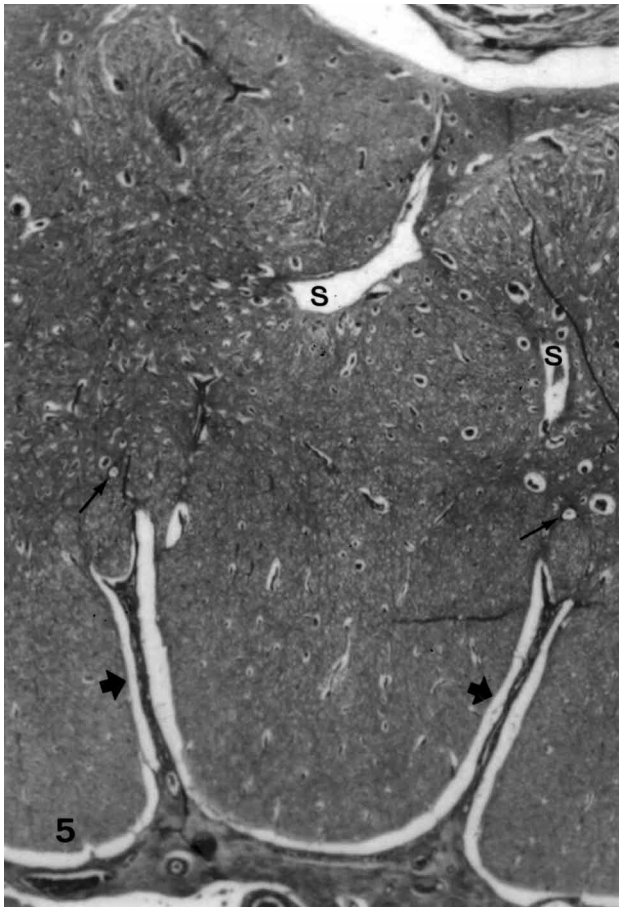

Figure 5. Two syringomyelic canals (S) in cervical segments of spinal column covered by single dura and piamater, double ventral fissures (thick arrows) and double central canals (thin arrows). HE stain, X 20.

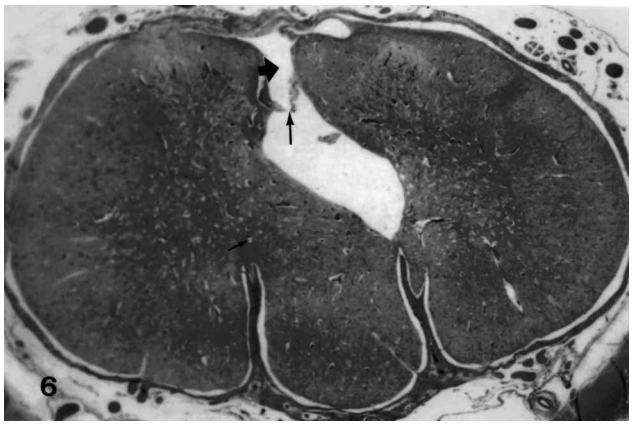

Figure 6. Syringomyelic canals and hydromyelia in the mid-thoracal segments of the spinal cord. Note the membrane (thin arrow) between a syringomyelic (thick arrow) canal and hydromyelia beginning from the right central canal. Small thin arrow indicates left central canal. HE, X 5.

between syringomyelic canal and hydromyelic central canal was detected in the some sections of the spinal cord (Figure 6). The caudal thoracic parts of the spinal column, which were bilaterally compressed due to probably abnormal conformation of the caudal thoracic vertebrae, were both dysplasic and had abnormal morphological features. Moreover, it was observed cleft-like central canal lined by the partial pseudostratified ependymal cells (myeloschisis), including hemorrhage with rarefaction of the neuropil and some microrosette formations in the two sides of it (Figure 7). There were a variety of alterations to the anatomy of the fissures, including a short penetration or absence of ventral median and dorsal median fissures, and ventral median fissure duplication with deep penetration including some small vessels were also observed. In these areas, therefore, the demarcation of the cuneate and gracilis fascicles in the funiculus dorsalis was absent (Figure 6). Proliferation of the astrocytes or minimal inflammatory reaction was found in neuronal tissues examined. No abnormal change in the other organs examined was observed.

The high frequency of congenital duplications in calves has been reported in some countries. Hiraga and Abe (6) reported that congenital duplications in calves accounted for 17.5% of all the anomalies and, among the congenital duplications observed, cranial duplication has 64% in ratio. They have classified cranial duplications as five types. According to their classification, the present case was nearly consistent with type 4 of cranial duplication with two heads at a common neck face in the opposite

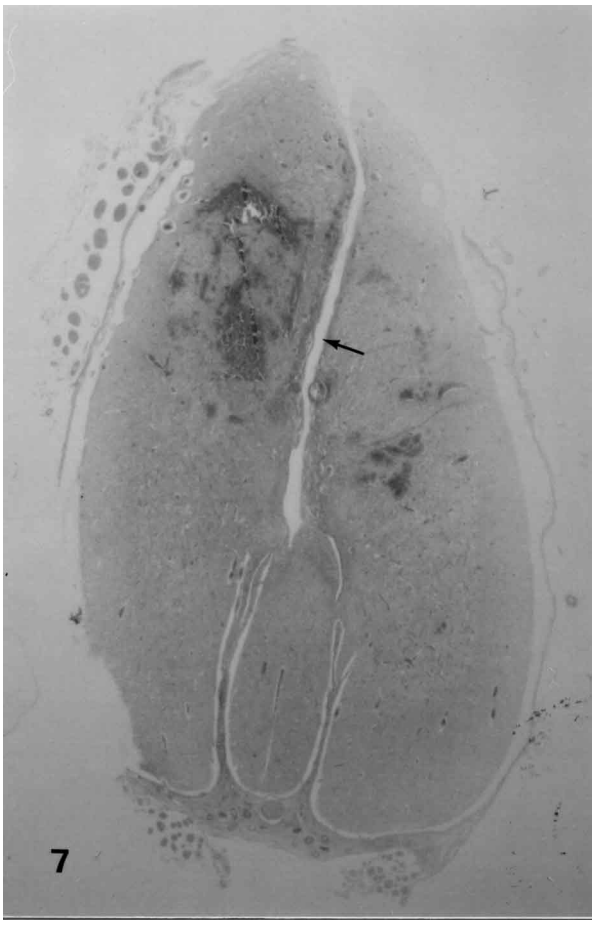

Figure 7. The caudal segment of the thoracal spinal cord. Myeloschisis (arrow) in the central canal and hemorrhage foci in neuropil. HE, X 2,5.

direction, four eyes, three ears, two mandibles, four cerebral hemispheres, two cerebelli, and two pituitary glands.

Pathogenesis of the ACM is obscure. In calves, the common cause of the many malformations is an in utero or perinatal viral infection. In the present case, a study for diagnosis of causative agents was not performed. In fact, ACM appears to be a specific syndrome of malformations that occurs sporadically without a proven cause (7,15). It may result from mechanical forces such as increased intracranial pressure caused by internal hydrocephalus or traction resulting from the lower end of the spinal cord being fixed to the walls of a meningomyelocele in SB, a disturbed pressure balance arising from leakage of cerebrospinal fluid from the SB into amniotic cavity and generalized overgrowth of CNS (5). These theories may be questionable because cases of ACM without

hydrocephalus or meningomyelocele and with normal brain size have been reported in calves (4,6). However, in human, there is consistent association between SB manifesta (aperta) and Chiari II malformation and all neonates with SB also have Chiari II malformation. This association should be regarded as part of the malformation sequence (16). Actually, bone anomalies seem to be important in the pathogenesis of ACM and three characteristic features of ACM in monocephalic and dicephalic calves include: 1) dorsoventral compression, 2) platybasia and 3) enlargement of the foramen magnum (6,12). In children, 71% of patients had the large foramen magnum and 41% had the flat posterior fossa (17). The experimental model of ACM in hamsters, using a single high dose of vitamin A in the 8th day of gestation, has made possible the analyses of the developmental interrelationships among the various skeletal and neurological disorders (18). The present case revealed cranial bone abnormalities such as flattening of occipital bones and becoming narrow dorsoventrally of cranial cavities, especially in the left cranium severely affected with ACM, enlargement of common foramen magnum, and SB. Cases of ACM in both monocephalic and dicephalic calves are usually associated with SB, meningomyelocele and/or internal hydrocephalus, arthrogryposis. Moreover, SB with meningomyelocele occurs in the lumbosacral area or less frequently in the cervical region (5,11,15,17). Other defects associated with ACM and SB are neuromyodysplasia, lordosis, cleft palate, taillessness, perosomus elumbus, and exencephaly (5). The present case was a dicephalic with ACM. SB was found clearly in the lumbar region with a total absence of spinal cord in this segment, and the calf exhibited diplomyelia of cervical/thoracic spinal cord with spinal dysraphism, arthrogryposis of hind limbs, cleft palate of both heads, kyphosis, but hydrocephalus, meningomyelocele and other defects mentioned above were not detected. Similar coexisting findings, excepting for spinal cord aplasia, have been reported in dicephalic calves with ACM (12-14). However, spinal cord aplasia is extremely rare in cattle (4,5,15). In the present case, the combination of ACM, cervico-thoracal diplomyelia, SB with lumbar spinal column aplasia, and other defects may have been due to congenital developmental imbalance between the processes of differentiation of the central nervous system and the skull-vertebral column.

SB is usually classified according to the severity of the lesion. SB occulta is used to describe simple failure of the dorsal bony arches of the vertebrae to fuse without involvement of the other tissues whereas SB manifesta (aperta) refers to amore severe lesion with the formation of a cleft extending thorough all the tissues dorsal to the spinal cord (3). In recent human terminology, the term "dysraphism" refers to a defect of closure of the neural tube, and should be therefore apply to abnormalities of the primary neurulation only. SD is classically categorized as open SD (OSD) characterized by exposure of the nervous tissue and/or meninges to the environment through a congenital bony defect. Conversely, closed SD (CSD) is covered by skin without exposure of neural tissue. The term SB merely refers to defective fusion of posterior spinal bony elements. SB aperta or cystica and SB occulta were used to refer to OSD and CSD, respectively, but are no longer widely used (16). The present case could be classified as a case of OSD, or according to former classification, SB manifesta.

In previous cases of monocephalic calves, spinal lesions are well documented, (5,9,15) but descriptions in dicephalic calves are scarce (12-14). In the present case, diplomyelia of cervical and thoracal segments of spinal cord with myelodysplasia was observed. At the various levels of spinal cord, the presence of double both central canal and ventral fissure warranted diplomyelia. Diplomyelia occurs within the common covering of leptomeninges and dura, and the degree of duplication is often with asymmetrical gray matter development. Conversely, diastomatomyelia consists of two separate spinal cords, each contained in its own meningeal covering and in separate vertebral canals (15). Moreover, variable degrees of hydromyelia and syringomyelia in cervical and thoracic segments of the spinal column were detected. Spinal dysraphism in monocephalic calves usually involves the caudal thoracic and lumbar spinal cord segments with or without spina bifida, arthrogryposis, or both (15). The dysraphic lesions described here occurred throughout all the segments of spinal cord with SB and arthrogryposis. Moreover, concomitant occurrence of the segmental diplomyelia and amyelia was made an interesting contribution to veterinary literature. In conclusion, the simultaneous defects in our case could be considered another variation on developmental defects of the neural tissue and bony structures in a dicephalic calf with ACM. Moreover, to clarify pathogenesis and etiology of dicephalus and associated malformations, further cases and etiologic investigations should be made.

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