Arnold-Chiari Malformation and Associated Anomalies in a Dicephalic Newborn Calf

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Summary: A full-term female newborn calf of the Japanese black short-horn breed which was two-headed (dicephalus) and had two incomplete necks was carefully dissected. The calf was one of twins, the other being a normal male. There was no record of reproductive problems with the cow. The left and right heads were of almost the same size and shape. Each head had a nose, mouth, eyeballs, and ears. The brains were very congested. That is to say, in both the left and right brains, the longitudinal fissure was unclear, and these was narrower pattern in the cerebral gyri. The surfaces of the two brains displayed poly- or microgyria-like features. The cerebellar gyri ran longitudinally among the occipital lobes of the cerebral hemispheres. The caudal half of the cerebrum, the cerebellum and the brain stem were distorted ventrally at an angle of approximately 80 degrees. Twisting of the brain stem was observed in the right brain. The end of the medulla oblongata and a tongue-like process of the cerebellum extended through the foramen magnum of the skull into each enlarged vertebral canal (Arnold-Chiari malformation). They extended under the arch of the each 2nd cervical vertebra. The 4th ventricle was also displaced and irregularly dilated. The two necks were conjoined in the single 3rd cervical vertebra at an angle of approximately 45 degrees to each other. The one trunk was slightly distorted. The ribs revealed the presence of a left 14th extra rib. In the lumbar region, spina bifida was apparent. The pelvis was distorted asymmetrically. The tail was short. The viscera appeared as one set, but the rectum and anus were duplicated. Although the calf displayed female features, the vagina was undeveloped. The four limbs appeared normal and the joints moved freely.

Materials and Methods

A full-term female calf of the Japanese black short-horn breed which was two-headed and had two incomplete necks calf of had died just a few minutes after birth, was donated to our Department by a farmer from Shichinohe-Town, Aomori Prefecture. There was no record of reproductive problems with the cow, which had a normal pregnancy. The newborn calf was embalmed with 10% neutral buffered formalin via the common carotid artery. The body was carefully dissected and a photographic record was made of all recognizable anomalies. Radiographic studies made beforehand supplemented the anatomical evidence. Portions of the spinal cord from the “conjoined area” were preserved in 10% neutral buffered formalin, processed routinely for histological examination, and stained with hematoxylin-eosin (H-E).
Results

1. General features

Externally, the specimen had two well-developed heads (dicephalus). The left and right heads were of almost the same size and shape. Each head had orbits, eyeballs, and ears. The nose, mouth, and pharynx of each head appeared normal.

The calf had two incomplete necks. The necks presented a Y-shaped configuration at an angle of approximately 45 degrees to each other (Figs. 1 and 2). One trunk was slightly distorted (monosomus). The urachus appeared to be patent within an enlarged 3-cm-diameter umbilical ring. In the lumbar region, there was spina bifida. The tail was 22 cm in length and shorter than that of the normal. Although the calf displayed female features, the external pudendum and singular vagina were undeveloped.

The two necks were separated just cranial to the level of the single 3rd cervical vertebra. Although the atlas and axis in the left and right necks were normal in form, the foramen vertebrale of the left and right atlases was very enlarged. Individual vertebrae appeared somewhat asymmetric. Both axes articulated with the single 3rd cervical vertebra. The remaining skeleton of the specimen was mostly single caudal from the level of the 4th cervical vertebra. The main axis of the thoracic vertebral body was distorted so as to be more or less S-shaped. The 1st to 13th pairs of ribs were normal, but the presence of a left 14th extra rib was observed. The sternum curved abruptly. The pelvis was distorted asymmetrically. The four legs appeared normal. The joints moved freely (Figs. 1, 2 and 3).

The lateral muscles of the furcated necks, i.e., the mastoid and occipital parts of the sternocleidomastoid, the mandibular part of the sternocleidomastoid muscle, the sternohyoid muscle, and others, appeared almost normal. The medial muscles of the necks were either absent or small and fused mutually with the contralateral muscles.

Externally, the spina bifida appeared as a depression, 7.0 cm in length and 2.5 cm in width, over the whole lumbar vertebral region. A radiographo-osteological study revealed the spina bifida to be more complex. It was almost parallel to the vertebral canal. In this region, the vertebral arch was not formed (Figs. 3 and 4).

A partial furcation of the trachea and esophagus was observed. The esophagi and tracheae in the left and right necks joined in the ventro-midline part of the cervical vertebrae, respectively. The conjoined trachea led to a set of lungs. The lobation of the right lung revealed cranial, middle, and caudal lobes. The accessory lobe of the right lung was absent. In addition, there was a small extra lobe derived from the left wall of the trachea. The left lung was normal in shape.

The heart was situated at the level between the 2nd to 4th ribs. By cutting the pericardium, a double heart was exposed, with the two hearts fused at the atrial side. The conjoined esophagus passed through the esophageal meatus of the diaphragm and entered the rumen. The forestomach and true-stomach formed a single set and were normal in shape. The liver and intestinal tract appeared to be normal except for some duplicated structures in the caudal rectum, and anal canal and anus.

2. Nervous system

The two separate, movable heads articulated with each atlas of the two separated necks. The brains were very congested. Each brain consisted of cerebral hemispheres, a brain stem and cerebellum (Figs. 5 and 6). The right brain was more or less smaller than the left one. The longitudinal fissure of each brain was unclear. In both the left and right brains, the cerebellum was extended caudally. Striking poly- or microgyri-like features were observed in the cerebrum and cerebellum. The gyri and grooves of the cerebellum, which was sandwiched between the occipital parts of the cerebral hemispheres, fitted and ran longitudinally. The caudal half of the cerebrum, the brain stem and the cerebellum were distorted ventrally at an angle of approximately 80 degrees. The cerebellar vermis appeared to be absent. All the cranial nerves ran several centimeters before leaving the cranial cavity.

The transverse fissure of the brain was of unclear V-shaped form. The so-called tongue-like process of cerebellum, the pons and the medulla oblongata extended through the large foramen magnum of the skull into the enlarged vertebral canal of the atlas and the axis. The two medullae oblongatae were flattened vertically and connected at the level of the 3rd cervical vertebra, and then formed a conjoined spinal cord (Fig. 4 and 7). The 4th ventricles were also displaced and irregularly dilated (Fig. 8). They were elongated toward the level of the axis, respectively. The 4th ventricles discharged into one central canal of the conjoined spinal cord. Microscopically, the cerebellum was composed of normal gray and white matter (Fig. 8). At the level of the 4th cervical vertebra, transverse sections of the spinal cord showed duplication (diplomyelia) of the gray and white matter with two well-developed median-ventral fissures (Fig. 9). That is, the white matter was partially fused, although the gray matter was not intermixed. Each medial gray column was smaller than its contra-lateral gray column. At the level of the 5th thoracic vertebra, the spinal cord had two central canals. At this level, the spinal cord was surrounded by a set of meninges.
Discussion

In the literature, cattle are reported to have the highest incidence of congenital duplicatory anomalies (Arthur, 1956; Leipold & Dennis, 1972a, 1972b; Leipold et al., 1972c). Greene et al. (1973) indicated that bovine double monsters had an incidence of 1-10 percent among all congenital anomalies. The types of duplication vary in degree and in the areas involved. The extent of bovine duplicatory malformation ranges from two completely separated heads to conjoined twins that constitute almost two complete individuals, as described by Dutton (1950), Frauchiger & Fankhauser (1952), Inoue & Nosaka (1955), Arthur (1956), Ikeda (1964), Dozsa (1966), Hofmann (1969), Schumacher et al. (1971), Leipold & Dennis (1972a, 1972b), Gruys (1973), Gordon & Lowe (1973), Nauriyal & Pandey (1979), Saperstein (1981), Dunn & Moreland (1983), Easton (1985), McGirr et al. (1987) and Misk & Hifny (1988).

Duplications of the head are found in about 75 percent of all bovine double monsters (Leipold et al., 1972c), but duplications of the entire head and neck are uncommon (McGirr et al., 1987). Duplications (dicephalus, diprosopus) of the head have been reported by Inoue & Nosaka (1955), Ikeda (1964), Hofmann (1969), Schumacher et al. (1971), Leipold & Dennis (1972a, 1972b), Gordon & Lowe (1973), Saperstein (1981), Easton (1985), McGirr et al. (1987), Misk & Hifny (1988) and others. Externally and radiographically, the anatomical features of the present case may be summarized as follows. The degree of doubling was complete from the 3rd cervical vertebra rostrally. That is, the separated heads were each attached to a spinal column through an atlas and axis which articulated onto a single 3rd cervical vertebra. There were four normal legs and one trunk. The external features resembled the cases of bovine duplicatory malformation reported by Dunn (1950), Arthur (1956), Ikeda (1964), Schumacher et al. (1971), Leipold & Dennis (1972b), Gruys (1973), Easton (1985), McGirr et al. (1987) and others. In particular, the present congenital anomalies appeared to agree with the cases of Leipold & Dennis (1972b) and Gruys (1973). Internally, there were some duplicatory malformations in the organs, as indicated above.

In the early literature, Chiari (1891) described anomalies of the hindbrain which showed variable displacement of the caudal vermis of the cerebellum into the cervical canal, accompanied by a similar caudal displacement of the caudal pons and medulla oblongata together with an elongated 4th ventricle. Arnold (1894) described a case of spina bifida in which there was caudal displacement of the cerebellum into the cervical canal. Malformation of the brains with displacement of the rhombencephalon into the cervical canal was first termed the Arnold-Chiari malformation (Arnold-Chiari'schen Missbildungen) by Schwalbe & Gredig (1907). It has also been called the Arnold-Chiari syndrome, Arnold-Chiari anomaly, or cerebello-medullary malformation syndrome. Cases of dicephalic calves showing abnormally large hemispheres have been described by Leipold & Dennis (1972a, 1972b), Easton (1985), McGirr et al. (1987) and others. However, they did not mention the Arnold-Chiari malformation.

In the present case, the brain stem, especially the medulla oblongata, and the tongue-like process of the cerebellum extended to the 2nd cervical vertebrae. This represented a case of protrusive displacement of the medulla oblongata and herniation of the cerebellum. Such a malformation must be due to congenital failure according to the smallness of the encasement of the brain. It must also reflect an abnormal overgrowth of the central nervous system in relation to the skull. Polysor microgyria of the cerebral and cerebellar cortex are often said to accompany the Arnold-Chiari malformation. In the present case, the surface features of the anomalous brain, which deviated from the normal bovine brain (Louw, 1989), were similar to those of previously reported calves (Frauchiger & Fankhauser, 1952; Cameron, 1957; van den Akker, 1962; Cho & Leipold, 1977).

The Arnold-Chiari malformation is also frequently accompanied by spina bifida, as described by Schwalbe & Gredig (1907), Frauchiger & Fankhauser (1952), Cameron (1957), Peach (1965), Gruys (1973) and Cho & Leipold (1977). Noden & de Lahunta (1985) and Saluja (1988) indicated that spina bifida presents a failure of the laminae to form dorsally over the vertebral foramen. It is not an anomaly of the spinal cord. In the case of the present study, the spina bifida was found clearly in the lumbar region. The first description of the malformation in a calf with spina bifida was given by Frauchiger & Fankhauser (1952). The present case was accompanied with diplomyelia in the cervical and thoracic spinal cord. Diplomyelia, a congenital malformation characterized by more or less perfect duplication of the spinal cord, has been described in detail by Herren & Edwards (1940), Cameron (1957), Gruys (1973) and Noden & de Lahunta (1985). The present case was a dicephalic and atlodidymic monosomic monster with the Arnold-Chiari malformation, diplomyelia, and spina bifida. Such a malformation may have been due to congenital failure of the developmental balance between the processes of differentiation of the central nervous system and the skull-vertebral column.

According to Inoue & Nosaka (1955), van den Akker (1962), Cho & Leipold (1977), Easton (1985), McGirr et al. (1987) and others, there are several hypotheses for the etiology of conjoined twins: the fission concept of imperfect twinning, the fusion concept of two embryonic masses combining, etc. The observations made on our atlodidymic specimen appear to be con-
sistent with the hypothesis that a singular early embryonic axis first split. In any case, various modified hypotheses have been put forward for the development of duplicatory anomalies (Leipold et al., 1972c; Noden & deLahunta, 1985).

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References

Explanation of Figures

Plate I

Fig. 1. Lateral view of a Japanese short-horn newborn calf with two heads and two incomplete necks. Each head had a nose, mouth, eyeballs, and ears. The four limbs appeared almost normal.
Fig. 2. X-ray photograph of the skeleton showing anomalous fusion and duplications. The left and right heads were almost the same in shape and size. The two necks conjoined in the single 3rd cervical vertebra at an angle of approximately 45 degrees to each other.
Fig. 3. A and B. X-ray photographs showing the anomalous skeleton of the caudal trunk. Extensive spina bifida (A) can be discerned in the lumbar region. The caudal trunk and pelvis were distorted asymmetrically (A and B).
Fig. 4. A and B. Dorsal view of the spina bifida which appeared in the lumbar region of the double monster calf (A). Note the absence of laminae of the lumbar vertebrae (B).
Fig. 5.  A, Dorsal view of the left brain of the double monster calf. The brains were very congested. The longitudinal fissure was unclear. The cerebral and cerebellar cortex showed poly- or microgyria-like features. The cerebellar gyri and sulci ran longitudinally. B, Lateral view of the left brain of the double monster calf. Arrows show the V-shaped transverse fissure corresponding to the tentorium. The tongue-like process of the cerebellum extended caudally.
Plate VI

Fig. 6. A, Dorsal view of the right brain of the double monster calf. The right brain also showed poly- or microgyria-like features. B, Lateral view of the right brain of the double monster calf. Arrows show the V-shaped transverse fissure corresponding to the tentorium. From this deep impressions, the caudal parts of the brain were distorted ventrad. Twisting of the brain stem was observed in the right brain. That is, the caudal half of the cerebrum, the cerebellum and the brain stem were distorted ventrally at an angle of approximately 80 degrees.

Plate VII

Fig. 7. Dorsal view of the two prolonged brain stems (medullae oblongatae) and two tongue-like processes of the cerebellum which extended caudally to the necks (arrows). At the cranial level of the 3rd cervical vertebra, the medullae oblongatae were conjoined at an angle of approximately 45 degrees to each other.

Fig. 8. Micrographs of transverse sections of the left and right prolonged brain stems and cerebellum of the double monster calf. A and B show the level at the middle part of the 1st cervical vertebra. Note the absence of the cerebellar vermis. C and D show the level of the caudal part of the 2nd cervical vertebra. E and F show the level of the middle part of the 3rd cervical vertebra. The asterisk indicates the elongated and irregularly dilated 4th ventricle.
Plate VIII

Fig. 9. A and B. Micrograph of a transverse section of the spinal cord at the level of the cranial part of the 4th cervical vertebra. A shows the duplication of the gray and white matter. Note the smaller medial gray columns. Arrows indicate the two median ventral fissures. B show a histological preparation of Fig. 9-A.