Two Calves of Arnold-Chiari Malformation and Their Craniums

Takeo HIRAGA, and Mitsuo ABE

Department of Veterinary Anatomy, Rakuno Gakuen University, 582 Bunkyo-dai-Midorimachi, Ebetsu 069, Japan

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ABSTRACT. Two calves with Arnold-Chiari malformation were examined macroscopically. The brain and cranium appeared to be compressed dorso-ventrally and parts of the cerebellum, medulla oblongata and fourth ventricle herniated through the enlarged foramen magnum to the vertebral foramen of the atlas. The internal cranial base composed of the sphenoid and occipital bones was abnormally flat, so the hypophyseal fossa and dorsum sellae were obscure. It was considered that the malformation probably resulted primarily from an abnormal development of these bones, the osteogenesis of which was induced by the notochord, and the brain secondly showed its abnormal shape and position. —**KEY WORDS:** Arnold-Chiari malformation, calf, congenital anomaly, cranium.

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Arnold-Chiari malformation (ACM), named after Arnold [1] and Chiari [4] by Schwalbe and Gredig [16], is defined as a herniation of the medulla oblongata and cerebellum into the foramen magnum [15]. In cattle this anomaly has been thought to occur very rarely [12] and some case reports have been published [5, 8, 9, 11, 17, 19]. The pathogenesis has not been clarified.

Recently we found two cases of ACM while investigating many other anomalous calves. This paper describes the anatomical findings on ACM in detail, especially on abnormalities of the ACM cranium in comparison with the normal, and discusses a possible pathogenesis of this anomaly from an embryological point of view.

MATERIALS AND METHODS

Two full term-born Holstein-Friesian calves with ACM were examined macroscopically. Each dam had previously given birth to 5 normal calves. Case 1 was a female and weighed 47 kg. She showed spina bifida, arthrogryposis of both pelvic limbs externally, and was sacrificed because of an inability to stand 4 days after birth. Case 2 was a

male and weighed 48 kg. He showed slight spina bifida and taillessness, and died 3 days after birth. As the control, the brain and cranium of 5 normal calves under 1 week of age were used. Bony tissues were prepared by maceration for detailed observation including measurement.

RESULTS

The brain, weighing 240 g in case 1 and 290 g in case 2, was about normal in size and appeared to be compressed vertically. Each side of the cerebral hemisphere was separated into two parts by a shallow groove (Fig. 1), in which the underdeveloped tentorium cerebelli were found. All the gyri and sulci of their caudal parts ran parallel longitudinally. The dorsal surface of the cerebellum appeared to be flat, and the boundaries between the vermis and hemispheres were obscure. In the sagittal section of the brain, the cerebral hemisphere extended more caudally than normal, so that the cerebellum in company with the medulla oblongata and fourth ventricle seemed to be pushed out through the foramen magnum to the vertebral foramen of the atlas (Fig. 2).

Caudal displacement of the brain stem was also indicated by a stretched condition of the intracranial parts of the third and following cranial nerves. No case was associated with internal hydrocephalus.

Fig. 1. Dorsal view of the brain. Arrow indicates the groove separating each side of the cerebral hemisphere into two parts. Dorsal surface of the cerebellum appears to be flat.

The cranium appeared more slender than normal (Fig. 3). The internal cranial base, composed of the presphenoid bone, basisphenoid bone and basilar part of the occipital bone, was abnormally flat in spite of their normal width and length. Due to the obscurity of the hypophyseal fossa and dorsum sellae, the hypophysis appeared to be in a more raised position than normal (Figs. 4 and 5). The cranial cavity appeared to be compressed dorso-ventrally corresponding to the shape of the brain. The foramen magnum was very large and its vertical diameter was longer than its horizontal diameter (Fig. 6 and Table 1).

In case 1, spina bifida was observed in the total sacral region. Fusion of both adrenal glands in the midline, hypoplastic spiral loop of the colon and a wry tail were also seen in this calf. In case 2, only the first and second sacral vertebrae were affected with spina bifida. This calf had only four sacral and no caudal vertebrae.

DISCUSSION

The findings on the brain of the present two cases were almost identical to those reported previously on ACM calves [5, 8, 9, 11, 17, 19]. ACM in calves seems to have

Fig. 2. Sagittal section of the brain showing an abnormal shape of the cerebral hemisphere and the herniation of the rhombencephalon through the foramen magnum (black line).

Fig. 3. Left lateral view of the cranium. Since the vertex is abnormally flat, the cranium appears slender and compressed dorso-ventrally as a whole.

Fig. 4. Caudal view of the internal cranial base showing its flattened appearance. The hypophyseal fossa and dorsum sellae are obscure.

wide variations, for example, Herzog [11] described 4 types and Wehner [19] 5 types based on the brain anomaly. Both of the present cases showed similarities to Herzog's type 3 and Wehner's type A and were characterized by the partial herniation of the cerebellum through the foramen magnum and the presence of the parallel gyri at the caudal parts of the cerebral hemispheres.

The cranium of ACM calves has not received much attention before now. In the present study, we noted the following anatomical findings on the ACM cranium in

comparison with the normal: (1) the cranium and cranial cavity have a compressed appearance dorso-ventrally; (2) the internal cranial base is so flat that the hypophyseal fossa and dorsum sellae are obscure; (3) the foramen magnum is wide and the vertical diameter is longer than its horizontal diameter. Of these observations, the second (2) abnormality is very significant, as endochondral osteogenesis of the internal cranial base is induced by the notochord [15]. These findings on the internal cranial base had not been detected, even in cases with hydrocephalus showing an excessive en-

largement of the cranial cavity [10].

There are several theories on the pathogenesis of ACM. Cho and Leipold [6] reviewed the literature on ACM and the following five theories are listed: (1) ACM may result from mechanical forces such as

Fig. 5. The floor of the cranial cavity after removing the brain. The hypophysis (arrow) is exposed without being covered by the meninges.

increased intracranial pressure caused by internal hydrocephalus [4]; (2) traction resulting from the lower end of the spinal cord being fixed to the walls of a meningo-myelocoele in spina bifida [13]; (3) a disturbed pressure balance arising from leakage of cerebrospinal fluid from the spina bifida into the amniotic cavity [3]; (4) generalized overgrowth of the central nervous system [2]; (5) abnormal embryonic development of unknown morphogenesis on the neural tissue and bony structures [7]. However, a theory which can precisely explain all cases of ACM has not been presented. There is some doubt on the first three theories, because ACM without spina bifida as well as ACM without hydrocephalus has been reported in calves [5, 11, 17, 19]. The fourth theory is also questionable, because the brains observed in the present study were almost normal in size. The last theory has been supported by some investigators [5, 14].

From the findings observed in this study, it seems to be an adequate hypothesis that ACM results primarily from the abnormally flat development of the internal cranial base. Because of this abnormality, it is suggested that the brain and hypophysis together are raised from the normal position

Fig. 6. Ventrocaudal view of the cranium showing an enlargement of the foramen magnum. Note the incisura (arrow) based on a disconnection between the lateral part of the occipital bone.

Table 1. Measurements of cranial cavity and foramen magnum on two ACM and 5 normal calves

	ACM calf		Normal calves
	Case 1	Case 2	Mean±SD
Cranial cavity			
Maximum width/length	0.76	0.75	0.79±0.03
Maximum height/length	0.52	0.46	0.62±0.03
Foramen magnum			
Vertical diameter (cm) (V)	4.28	3.64	2.72±0.26
Horizontal diameter (cm) (H)	3.61	3.16	3.04±0.26
V/H	1.19	1.15	0.89±0.06

and, consequently, the rhombencephalon is pushed out through the foramen magnum. The absence of the hypophyseal fossa and dorsum sellae certainly supports this hypothesis, which may be in favour of the fifth and final theory mentioned above regarding the pathogenesis of ACM.

ACM has been thought to be associated with spina bifida and hydrocephalus [15], and the former was also observed in the present two cases. Warkany et al. [18] reported ACM with spina bifida as early as ten weeks' gestation and that with hydrocephalus at 18 weeks in human fetuses. In keeping with their findings, it may be thought that ACM, spina bifida and hydrocephalus are affected by the teratological factor in embryogenesis at nearly the same critical points.

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要 約

Arnold-Chiari 奇形の子ウシ 2 例：平賀武夫・阿部光雄（酪農学園大学酪農学部家畜解剖学教室）—— Arnold-Chiari 奇形の子ウシ 2 例を肉眼的に観察した。脳と頭蓋は背腹に扁平で、小脳、延髄および第四脳室の一部は拡張した大孔を通り、環椎椎孔内に逸脱していた。蝶形骨と後頭骨の底部により構成される内頭蓋底は非常に平坦であり、下垂体窩と鞍背は不明瞭であった。この奇形は、脊索に誘導されて発生するこれらの骨の異常がまず起こり、二次的に脳の異常を引き起こしたものと考えられた。