

Magnetic Resonance Imaging in 24 Dogs with Chiari Type 1-like Malformations

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Abstract : Chiari type 1 malformation is a developmental condition characterized by cerebellar herniation and syringohydromyelia in human beings. It has been reported as a common condition in the cavalier King Charles spaniel that is similar to human Chiari type 1 malformations. However, there are few documentations of diagnosed Chiari type 1 like malformation in other breed dogs. Abnormalities compatible with such a malformation were identified by magnetic resonance imaging in 24 dogs with neurologic signs in this study. The dogs were 15 females and 9 males. Their breed were variable, and 6 of the dogs were maltese, 5 were shih-tzu. The dogs had a variety of neurological signs and the severity of cerebellar herniation, syringohydromyelia, intracranial intra-arachnoid cyst or hydrocephalus.

Key words : Chiari malformation, dog, MRI, cerebellar herniation, syringohydromyelia.

Introduction

Chiari type 1 malformation is a condition in which the cerebellum, specifically the cerebellar tonsils 'slips' into the space where the spinal cord travels into the skull (2,4). Associated abnormalities include a small posterior fossa, inferior displacement of the medulla and/or the fourth ventricle, angulation of the cervicomedullary junction, hydrocephalus, syringohydromyelia and various osseous anomalies (4,11,13,22). A similar syndrome of cerebellar herniation, called Chiari type 1-like malformation, has been reported in dogs (3,5,18).

In human beings there is evidence that the pathogenesis of the condition is secondary to the underdevelopment of the occipital bone, which leads to 'overcrowding' of the posterior fossa and herniation of the cerebellar tonsils through the foramen magnum (13,14,21). This has the effect of altering the circulation of cerebrospinal fluid (CSF) at the foramen magnum and preventing the equilibration of the pressure between the intracranial and spinal subarachnoid spaces (4,15). The development of a pressure gradient across the foramen magnum, which is exacerbated whenever the patient performs a Valsalva manoeuvre (forced expiration against a closed glottis which may occur, for example, when defaecating), leads to progressive herniation of the cerebellar tonsils. The occlusion by the cerebellar tonsils of the subarachnoid space at the foramen magnum then leads to the development of syringohydromyelia. This occurs because the transiently increased intracranial pressure during each systole forces the cerebellar tonsils inferiorly, inducing a pressure wave that acts on the surface of the spinal

cord and promotes the leakage of CSF into the cord (15).

This syndrome has been reported in the cavalier King Charles spaniels (3,5,18). However, there are little documentations of diagnosed Chiari type 1-like malformation in other breed dogs. We report the characteristics of Chiari type 1-like malformation in 24 dogs.

Materials and Methods

Medical records of 24 small breed dogs were examined by MRI between March 2005 and January 2006 were reviewed. For dogs included in this study, information obtained from the database included age, breed, sex, clinical signs and concurrent diseases. Each of the dogs was examined physically and neurologically, and MRI examination of the brain and cervical spine using 0.2 tesla permanent magnet (VetMR[®], Esaote, Italy), including T1- and T2-weighted transverse and sagittal images.

Results

Twenty four dogs satisfied the criteria for inclusion in this study. In 24 dogs with Chiari type 1-like malformations, there were 15 females (2 neutered) and 9 males (4 neutered) and the median age was five years (range 5 months to 13 years). The breeds of the dogs were variable. Six of the dogs were maltese, 5 were shih-tzu and others are summarized in Table 1.

The dogs' neurological signs are also summarised in Table 1. Fourteen had seizures, six had ataxia, and other signs included tremor, tetraparesis, circling, and neck pain.

Based on MRI findings, all of the dogs had herniation of the cerebellum compatible with a Chiari type 1 malformation

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Table 1. Signalments, clinical signs and MRI findings of 24 small breed dogs

No.	Breed	Sex	Age	Clinical Signs	Concurrent Disease	Severity of Cerebellar Herniation
1	ST	F	3y11m	Seizure, tetraparesis	ME, HC	Mild
2	ST	MC	3y4m	Seizure	ME, IVDD	Mild
3	MT	F	3y	Tremor	SH, ICIAC, HC	Mild
4	PK	F	2y6m	Seizure	ICIAC, HC	Mild
5	ST	F	5y	Seizure		Mild
6	CS	F	2y	Seizure, circling		Mild
7	Papillon	F	13y	Tetraparesis	SH, HC	Mild
8	PD	M	5y6m	Ataxia	SH, HC	Mild
9	YT	F	12y	Seizure, circling	SH, HC, ICIAC	Mild
10	Mongrel	FS	11y1m	Seizure	SH, HC	Mild
11	ST	F	1y	Seizure		Mild
12	MT	MC	1y4m	Seizure	ME, IVDD	Mild
13	ST	MC	6y6m	Ataxia, neck pain	IVDD, ICIAC	Mild
14	MT	F	3y	Seizure	ME, HC, SH	Mild
15	CS	F	5y4m	Seizure		Mild
16	MT	F	6y	Tetraparesis	GME, SH, HC, ICIAC	Mild
17	YT	FS	7y4m	Tetraparesis	IVDD, HC	Mild
18	PD	M	6y	Neck pain	IVDD	Mild
19	MT	MC	5y	Tremor		Mild
20	PD	M	7y6m	Seizure, ataxia	SH	Mild
21	CH	F	3y1m	Tremor, ataxia	SH, HC, otitis media	Marked
22	CKCS	F	5m	Seizure, circling	ME, otitis media	Marked
23	MT	M	9m	Seizure, ataxia	ME, SH	Marked
24	CH	M	1y7m	Ataxia, circling	SH, Cerebellar infarction	Marked

ST: Shih-tzu, MT: Maltese, PK: Pekingese, CS: Cocker spaniel, PD: Poodle, YT: Yorkshire terrier, CH: Chihuahua, CKCS: Cavalier King Charles spaniel
ME: meningoencephalitis, HC: hydrocephalus, IVDD: intervertebral disk disease, SH: syringohydromyelia, ICIAC: intracranial intra-arachnoid cyst, GME: granulomatous meningoencephalitis

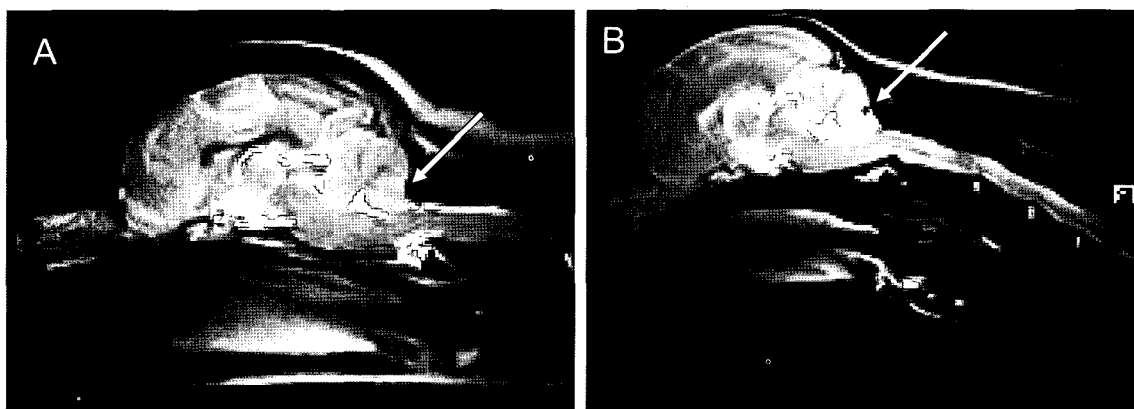


Fig 1. Subjective evaluation of the cerebellar herniation observed in magnetic resonance images in the median plane. Mild (A in dog 6) and marked (B in dog 23) caudal displacement of cerebellar tonsils (arrows).

(Fig 1). The severity of cerebellar herniation was evaluated subjectively in MR median plane images. Twenty dogs had mild herniated cerebellar tonsil, while only 4 dogs had marked cerebellar herniation. Eleven of 24 dogs had hydrocephalus,

and 10 had syringohydromyelia with Chiari type 1-like malformation. Other concurrent diseases presented these dogs contained intervertebral disk disease, intracranial intra-arachnoid cyst, meningoencephalitis, cerebellar infarction and otitis

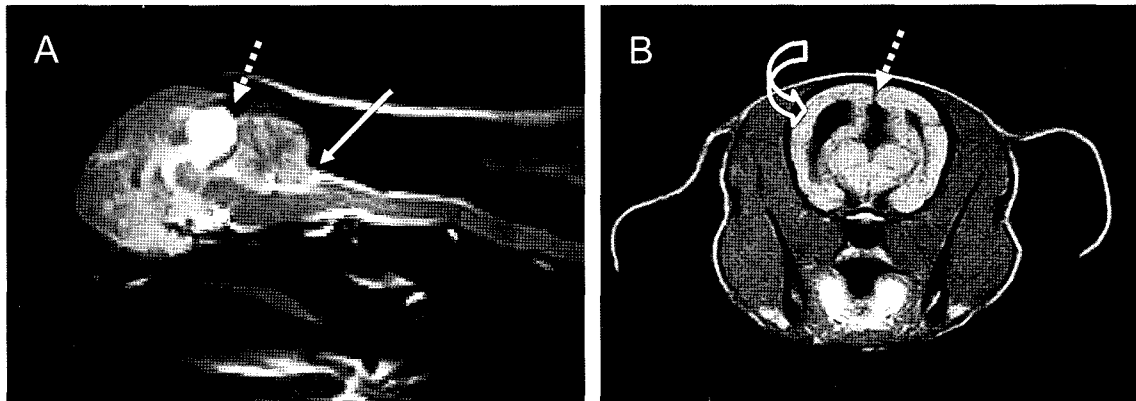


Fig 2. Sagittal T2 weighted image(A) and transverse T1 weighted image show coincident diseases, intracranial intra-arachnoid cyst (dotted arrows) and hydrocephalus (curved arrow) in dog 4 with mild Chiari type 1-like malformation.

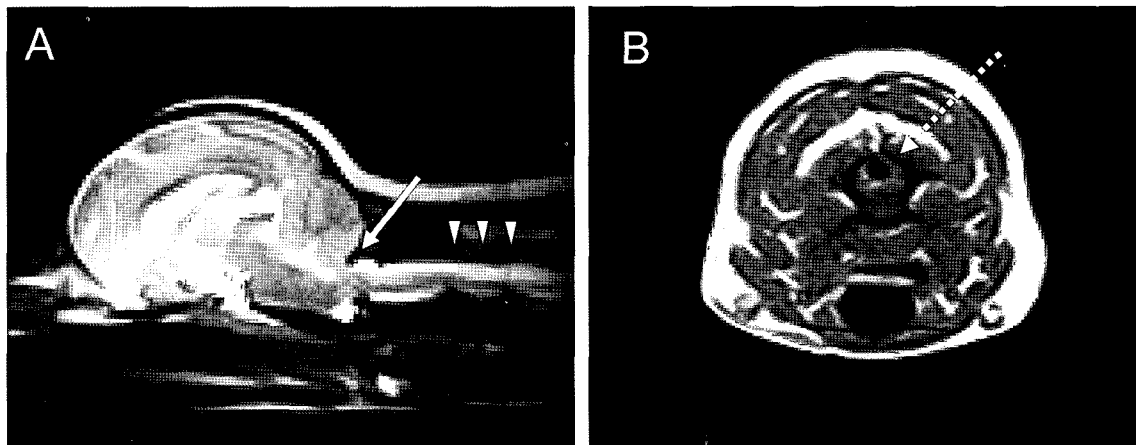


Fig 3. Mid-sagittal T2 weighted MR image (A) of cranium in dog 9 represents hydrocephalus, mild intra-arachnoid cyst, Chiari type 1-like malformation (long arrow) and hyperintense syringohydromyelia (arrow heads). Syringohydromyelia are evident as hypointense signal on T1 weighted transverse image (B) of cervical spinal cord (dotted arrow).

media (Table 1). Typical MR images of mild and marked Chiari type 1-like malformations and concurrent diseases represented at Figs 1-3.

Discussion

All 24 of these small breed dogs had an abnormally shaped cerebellum. On mid-sagittal MR images, cerebellum had a concave caudal aspect and a herniated vermis towards or through the foramen magnum. Among these dogs, females (54%) were most likely to be affected with Chiari type 1-like malformations, followed by males (21%), castrated males (17%), and spayed females (8%). The number of females in our study was higher than previously reported numbers (10). They reported there was no difference between male and female prevalence in 40 cavalier King Charles spaniels with Chiari type 1-like malformation. Although the discrepancy between these findings could reflect the breed difference, more patient number should be required for the accurate tendency understanding.

Studies reveal that Chiari type 1-like malformation can occur in dogs ranging 6 months to 10 years of age (17). Mean age of dogs in the present study was five years (range, 5 months to 13 years), which was similar to the values reported in recent studies (10,17).

In our study, 25% of the dogs with Chiari type 1-like malformations were maltese dogs, 20% were shih-tzu dogs, and 13% were poodles. Previous studies reported cavalier King Charles spaniels are apparently over-represented with Chiari type 1-like malformations (20). A familial or genetic basis is suspected (19) in this breed. However, this study contained only one cavalier King Charles spaniel dog. The discrepancy between these breed distributions likely represents the difference in pet populations at the respective countries; on the other hands, shih-tzu, maltese and poodle are very preferential breeds in Korea but cavalier King Charles spaniels are not. Other author has encountered a number of small-breed dogs not cavalier King Charles spaniels with syringohydromyelia and Chiari type 1-like malformations of the caudal fossa; the

majority of these dogs have been toy and miniature poodles (6).

The symptoms reported in other articles are very variable and include neck pain, torticollis or scoliosis, spinal hyperesthesia, and neurologic deficits relating to cervical spinal cord dysfunction. Intracranial signs, such as facial paresis and vestibular dysfunction have also been reported (17). Paroxysmal involuntary scratching of the neck and flank has been associated with this condition (5,18). Clinical signs in 24 dogs were variable and contained seizure, ataxia, neck pain, and circling in the present study. Human patients with Chiari type I malformation present in adolescence or early adulthood with a variety of symptoms occurring in various combinations. There are foramen magnum compression with ataxia, sensory deficits, cerebellar signs, lower cranial nerve palsies and headaches, central cord syndrome with dissociated sensory loss, segmental weakness and long tract signs possibly combined with lower cranial nerve palsies and cerebellar syndrome (16). Seizures may occur in people with the condition (7), and Lu *et al.* (10) reported seizures occurred in seven of 40 dogs, suggesting that the condition should be considered as a possible underlying cause in cavalier King Charles spaniel with epilepsy. In our study, seizures were the most common clinical signs (54%). This finding might be due to concurrent intracranial diseases, such as hydrocephalus and meningoencephalitis not Chiari type 1-like malformation.

Syringohydromyelia is commonly associated with Chiari type 1 malformation in human and has been suggested to play an important part in the pathogenesis of intramedullary cavities (9). It has reported hydromyelia occur in 20-85% of patients with Chiari type 1 malformation, and hydrocephalus in up to 55% of patients (1,12). Other authors state that hydrocephalus does not occur (8). In dogs, it is believed that hindbrain anomalies such as Chiari type 1-like malformation and intrarachnoid cysts are not merely coincidental malformations that occur concomitantly with syringohydromyelia by disrupting normal CSF flow mechanisms (6). In the present study, the percentage of occurrence were 46% (hydrocephalus) and 42% (syringohydromyelia). The twenty nine percentage dogs were simultaneously affected by both diseases. Dog breeds predisposed to hydrocephalus include the chihuahua, Yorkshire terrier, toy poodle, Pekingese and maltese (17). It was largely overlapped with the breeds of this study.

There was no correlation between the neurological sign and the severity of the herniation of the cerebellar tonsil, or the presence of syringohydromyelia or hydrocephalus as previous reports (8,10).

The recognition that the malformation is very prevalent in small breed dogs is due to the increased availability of MRI. Especially, exact midsagittal images most effectively display abnormalities of cerebellar shape. Computed tomography could not show cerebellar abnormalities and small syringohydromyelia but might useful for evaluating occipital bone dysplasia associated with Chiari type 1-like malformation.

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자기공명영상을 이용한 24마리의 개에서의 Chiari 1형 유사 기형 진단

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요 약: 사람에서, Chiari 1형 기형은 소뇌 탈출과 척수공동증이 특징인 발달성 장애이다. 이러한 사람의 Chiari 1형 기형과 유사한 질환이 cavalier King Charles spaniels에서 흔히 나타났다. 그러나, 이러한 Chiari 1형 유사 기형이 다른 종의 개에서 진단 보고된 증례가 거의 없는 실정이다. 이에 본 연구에서는 24마리의 개에서 자기공명영상 장치를 이용하여 Chiari 1형 유사 기형으로 진단된 증례를 보고하고자 한다. 이 개들은 다양한 신경 증상과 다양한 정도의 소뇌 탈출, 척수공동증, 두개관내 거미막 낭종 또는 뇌수종의 병발 질환들을 나타내었다.

주요어: Chiari 기형, 개, MRI, 소뇌 탈출, 척수공동증