MRI of Hydrosyringomyelia Combined to Hydrocephalus and Occipital Dysplasia in a Dog

Chi-Bong Choi, Chun-Sik Bae[1] and Hwi-Yool Kim*

College of Veterinary Medicine, Konkuk University, Seoul 143-701, Korea, [1]College of Veterinary Medicine, Biotechnology Research Institute, Chonnam National University, Gwangju 500-757, Korea

Received July 19, 2005 / Accepted August 19, 2005

Hydrosyringomyelia is a dilation of the spinal cord central canal. In human it may be caused by congenital malformations such as Dandy-Walker syndrome and Chiari malformations or may be acquired as a result of infection, trauma or neoplasia. Hydrocephalus is an excessive accumulation of cerebrospinal fluid within the ventricles and occipital dysplasia is the dorsal extension of the foramen magnum. Hydrosyringomyelia and hydrocephalus can be confirmed by computed tomography or magnetic resonance imaging (MRI). A 3-year-old male maltese was presented with a history of long-term seizure. Blood examination was all unremarkable. On rostrodorsal-caudoventral oblique radiograph of the skull showed severe occipital dysplasia. On brain sonography through the persistent fontanelle, severe lateral ventriculomegaly was revealed. MRI examination revealed hydrocephalus and hydrosyringomyelia. Diuretic therapy didn't reduce clinical symptoms and surgical decompression was conducted. The dog responded well with ventriculo-peritoneal shunting. MRI is the most superior modality to diagnose hydrocephalus and hydrosyringomyelia, to plan therapy and to determine the prognosis.

Key words – MRI, hydrosyringomyelia, hydrocephalus, occipital dysplasia

Hydrosyringomyelia is characterized by the development of fluid-filled cavities within the spinal cord[15]. In veterinary medicine, there have been sporadic reports of hydrosyringomyelia often concurrent with developmental disorders of the craniocervical junction such as Chiari malformations and Dandy-Walker syndrome[1,2,6,8,12]. Hydrosyringomyelia with scoliosis secondary to suspected trauma has also been reported[7].

In this case, hydrosyringomyelia combined to hydrocephalus and deformity and overcrowing of the foramen magnum such as occipital dysplasia is described especially in the point of MRI.

Case Report

A 3-year-old neutered male maltese was referred for evaluation of long term seizure. On physical exam, open fontenelle with dome-shape head was found. Blood works and neurologic examination were unremarkable. Dorsal extension of foramen magnum was noted in the rostrodorsal-caudoventral oblique radiograph of the skull (Fig. 1).

Ultrasonography and brain MR images showed diffuse and symmetric dilatation of all ventricle (Fig. 2 and 3). These findings suggest severe occipital dysplasia and hydrocephalus, respectively.

MRI examination was performed using 3.0 T superconducting magnet MR equipment with a knee coil. On MR images, a long, tubular, sharply marginated, T1-low and T2-high, fluid signal intensity structure was noted in the central portion of cervical spinal cord, representing hydrosyringomyelia (Fig. 4).

Diuretic therapy did not reduce clinical symptoms and then ventriculo-peritoneal shunting was conducted. The dog has responded well with the surgical decompression.

Discussion

Pathologically, hydromyelia is simply ependymal lined distention of the central canal, whereas syringomyelia is defined as cerebrospinal fluid (CSF) dissection through the ependymal lining to form a paracentral cavity[14]. In practice, these two conditions often coexist and intercommunicate. Because even the histologic distinction between hydro- and syringomyelia is sometimes difficult and because their appearances on imaging studies are usually indistinguishable, these entities are often grouped under the term hydrosyringomyelia. This term describes any pathological cavity

*Corresponding author
Tel : +82-2-450-3710, Fax : +82-2-446-9876
E-mail : hykim@konkuk.ac.kr

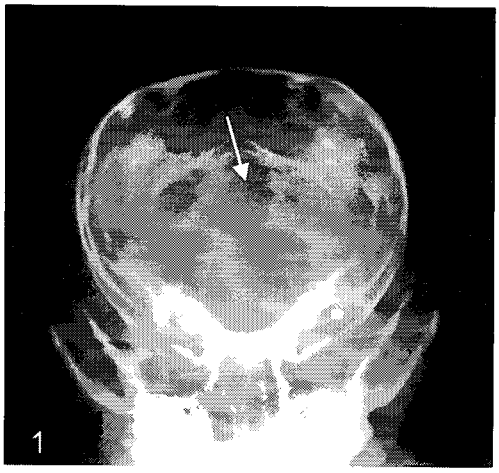

Fig. 1. Rostrodorsal-caudoventral oblique radiograph of the skull. Note the dorsal extent appearance of the foramen magnum (arrow). Severe occipital dysplasia.

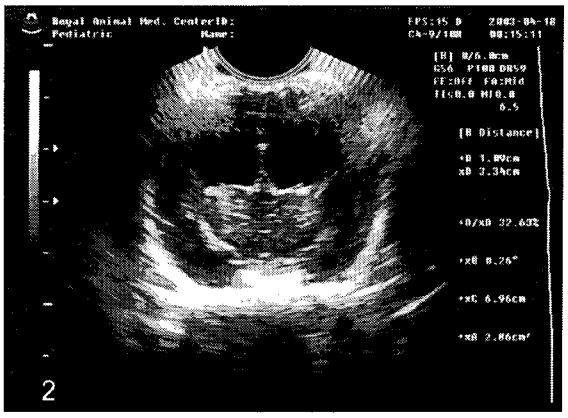

Fig. 2. Brain sonography through persistent open fontanelle.

that occupies the substance of the spinal cord, whether or not it is continuous with the central canal[4,14].

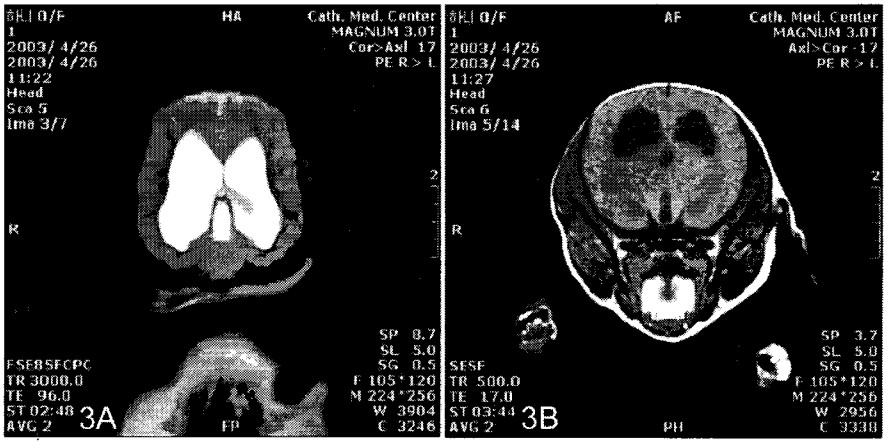

Fig. 3. On T2 weighted coronal image (A) and T1 weighted transverse image (B). Symmetric dilatation of lateral ventricles are noted and the 3rd ventricle is also dilated. T1WI-spin echo, TR-500 msec, TE-17 msec, NEX-2, T2WI-fast spin echo, TR-3000 msec, TE-96 msec, NEX-2

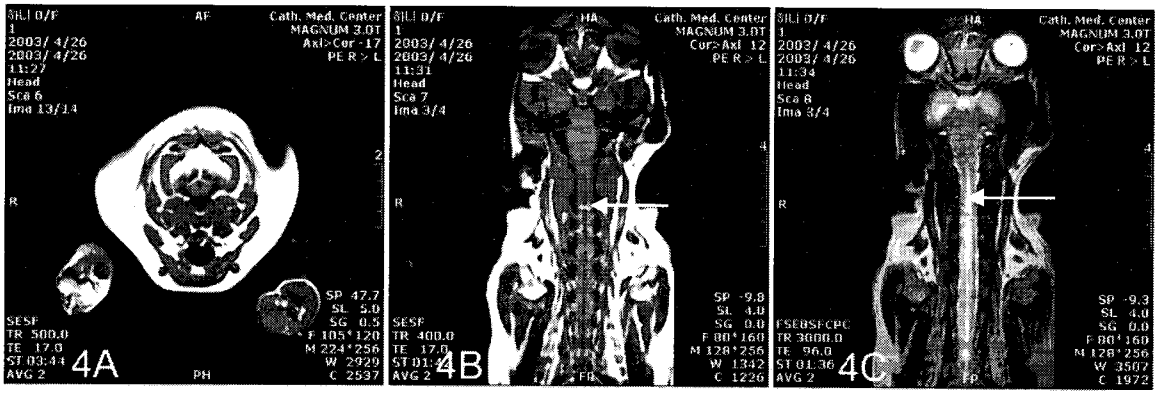

Fig. 4. MR images of Hydrosyringomyelia. On T1 weighted transverse image (A), this lesion is noted as a small round low signal intensity in central area. T1 weighted coronal image (B) of cervical region shows well defined, long tubular, low signal intensity lesion in the central part of the spinal cord. It is seen as homogenous bright signal intensity on T2 weighted coronal image (C) (arrow). T1WI-spin echo, TR-500 msec, TE-17 msec, NEX-2, T2WI-fast spin echo, TR-3000 msec, TE-96 msec, NEX-2.

Hydromyelia and syringomyelia can be congenital or acquired. The pathogenesis of congenital hydromyelia is debatable. The dysraphic theory suggests that a neural tube closure defect is responsible, whereas the hydrodynamic theory maintains that disturbed. CSF outflow from the fourth ventricle is responsible. Syringomyelia then supervenes as the dilated ependymal canal ruptures into the spinal cord parenchyma[14,16].

In the dog, CSF circulates from the ventricular to the subarachnoid space by way of the lateral apertures of the fourth ventricle (foramenae of Luschka). The dog does not have a foramen of Magendie. Flow of CSF is thought to be due to the pulsation of blood in the choroid plexuses. With each pulsation the CSF pressure rises and surges towards the lateral apertures[3].

In an experimental study, ventriculography in normal cats resulted in contrast leaving the ventricular system via the lateral apertures to opacify the subarachnoid space, but filling of the central canal by contrast medium was never observed[5]. Whether, in the dog, CSF flow occurs from the ventricular system to the central canal, flows within the latter, or whether or not there is a communication between the central canal and sacral subarachnoid space could not be ascertained from the literature. In experimental canine hydrocephalus/hydromyelia, a communication was occasionally found between the central canal and sacral subarachnoid space[17].

Magnetic resonance imaging is the ideal non-invasive imaging technique to screen for suspected hydrosyringomyelia and a possible etiology[10]. Cerebrospinal fluid behaves like water and the ventricular system appears black (hypointensive signal) under T1 weighting compared with brain parenchyma and white (hyperintensive signal) under T2 weighting[14]. Fluid T1 and T2 relaxation times are similar to that of CSF[13]. Substantial flow within the hydrosyringomyelic cavity is possible. Since cortical bone has dark signal on MRI and appears to subarachnoid space on short Spin-echo techniques the posterior margin of the foramen magnum is seldom identified, making identification of small cerebellar prolapses difficult[9,13]. Although it is possible to increase the signal of CSF compared to cortical bone by increasing the TE and TR in Spin-echo techniques, the resulting loss of resolution usually makes it impossible to image the edge of the foramen[9,11]. The degree of subarachnoid space narrowing caused by cerebellar ectopia is difficult to assess[11]. Cavitation due to cord neoplasia can be distinguished from hydromyelia by means of MRI. The cystic and solid parts of the neoplasm have different signal intensities and can be distinguished from hydrosyringomyelia on that basis[10]. In humans with hydromyelia, CSF analysis was normal and fluid aspirated percutaneously from cavitatory lesions was within normal limits and similar to each other in 74 patients[7,9].

Diagnostic ultrasonography of the occipital region may prove to be of benefit to visualize cerebellar or ventricular abnormalities, particularly in the presence of occipital dysplasia[9]. The malformations described, and their various subtypes, may be difficult to accurately classify by means of survey radiographs, contrast medium studies, ultrasonography and even MRI, particularly considering the wide range of canine skull types. Greater accessibility for animals to MRI may result in increasing numbers of patients with hydrosyringomyelia being diagnosed[9].

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초록 : 개에서 뇌수두증과 후두골 이형성증을 동반한 척수공동증의 자기공명영상학적 평가 1례

최치봉 · 배춘식[1] · 김휘율*
(건국대학교 수의과대학, [1]전남대학교 수의과대학 및 생물공학연구소)

척수공동증(hydrosyringomyelia)은 척수내의 중심관(central canal)이 확장되는 질환으로 사람에 있어서는 Dandy-Walker 증후군이나 Chiari 기형과 같은 선천적인 기형에 의해 발생되거나 감염, 외상 및 종양과 관련되어서도 발생된다. 뇌수두증(hydrocephalus)은 뇌척수액이 뇌실내에 과도하게 축적되는 것을 말하며 후두골 이형성증(occipital dysplasia)은 후두공이 과도하게 위쪽으로 연장되어 열린 질병이다. 척수공동증과 뇌수두증은 컴퓨터단층촬영이나 자기공명영상 검사에 의해 확진되어질 수 있다. 3세령의 수컷 말티즈가 장기간의 발작 증세를 주증으로 내원하였다. 혈액검사상 특이소견은 없었으나 rostrodorsal-caudoventral oblique의 두개골 방사선 검사에서 심한 두개골 이형성증 소견이 확인되었고 열려진 천문을 통한 초음파 검사상 양측 외측 뇌실이 모두 심하게 확장된 것이 확인되었다. 자기공명영상 검사에서 뇌수두증과 척수공동증이 진단되었다. 이뇨제를 중심으로 한 내과적 처치에 반응을 하지 않아 외과적 감압술(ventriculo-peritoneal shunting)을 시행하였고 현재까지 잘 유지되고 있다. 자기공명영상검사법은 뇌수두증과 척수공동증을 진단하고 그에 따른 치료계획을 수립하며 예후를 평가하는데 있어 가장 우수한 진단 기법이다.