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Ethmoidal Meningoencephalocele Associated with Seizure in a Juvenile Alaskan Malamute

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Abstract : Signalment: An 8-month-old female Alaskan malamute was presented for progressive cluster seizure disorder. Results: There were no abnormalities on neurological examination, survey radiographs, or blood analysis. Magnetic resonance (MR) imaging and computed tomography revealed extension of the olfactory bulb and frontal lobe into the nasal cavity. They also confirmed abnormal anatomy of the nasal turbinates within the rostral part of the nasal cavity and the absence of a cribriform plate. On T2-weighted and fluid-attenuated inversion recovery images, the herniated brain showed heterogeneous and hyperintense signals consistent with intraparenchymal edema. Transverse MR images showed brain herniation into the right frontal cavity and an asymmetrical lateral ventricle because of a left midline shift. On contrast-enhanced MR images, the protruding brain parenchyma was mildly enhanced. Ethmoidal encephalocele was suspected as the final diagnosis. Despite symptomatic treatment, the dog continued to exhibit seizures and was euthanized. Clinical relevance: Ethmoidal encephalocele is a rare disease in dogs. However, it could be considered as a cause of seizure in young dogs.

Key words **:** dog, ethmoidal meningoencephalocele, seizure.

Introduction

Encephalocele is defined as a congenital malformation of the central nervous system (CNS) with protrusion of cranial content through defects in the cranial bones. Development of encephalocele is usually caused by a disturbance in the separation of the surface ectoderm and neuroectoderm during neural tube formation (2,6,17). In humans, encephalocele is classified according to the location of the skull defects and the content of the protrusion (4). On the basis of human classification and terminology, ethmoidal meningoencephalocele is a subtype of encephalocele caused by the protrusion of cerebral tissue and meninges through a congenital deficit in the ethmoid (17). Herniated brain lesions that include part of the olfactory bulb or frontal lobe are associated with seizures in dogs. Due to the intricate architecture of the CNS, abnormalities occurring during brain development are particularly liable to cause deficits and derangements, such as seizure activity. In dogs, lesions in olfactory bulbs and cerebral frontal lobes can be accompanied by seizure activity (5,9). Advanced imaging methods, such as magnetic resonance imaging (MRI) and computed tomography (CT), may enable precise diagnosis and treatment of encephalocele.

This case report presents findings of ethmoidal meningoencephalocele with signs of cluster seizure activities on CT and MR.

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Case

An 8-month-old female Alaskan malamute weighing 12 kg was referred for progressive cluster seizure disorder. Clinical signs were observed to begin when the dog was 2 months old. Seizures were accompanied by salivation, unconsciousness, and limb stiffness and continued to worsen. Additionally, the dog showed mild dyspnea with nasal congestion.

On blood analysis, alkaline phosphatase (ALP) was 666 IU/L beyond the normal range (29-97 IU/L), but no other abnormalities were evident. Additionally, no abnormalities were found on survey radiography or physical and neurological examination. Magnetic resonance examination was performed under general anesthesia using a 0.3-Tesla MRI scanner (Airis II, Hitachi, Japan) and human joint coil at Chungbuk University Veterinary Medical Center. The brain MRI protocol included pre- and post-contrast T1-weighted imaging, T2-weighted imaging, and fluid-attenuation inversion recovery (FLAIR) imaging sequences. The imaging sequences were obtained in the transverse, dorsal, and sagittal planes. The MRI studies revealed a protrusion of the right cerebral olfactory bulb into the nasal cavity through a bony defect that was seen in the cribriform plate of the ethmoid bone. On T2-weighted and FLAIR images, herniated cerebral tissues showed heterogeneous and hyperintense signals, consistent with cerebral edema (Fig 1). Additionally, MRI showed an asymmetrical lateral ventricle on transverse images (Fig 2). On contrast-enhanced T1-weighted images, the lesions showed no or mild contrast enhancement. There was neither evidence of hydrocephalus nor other abnormality that

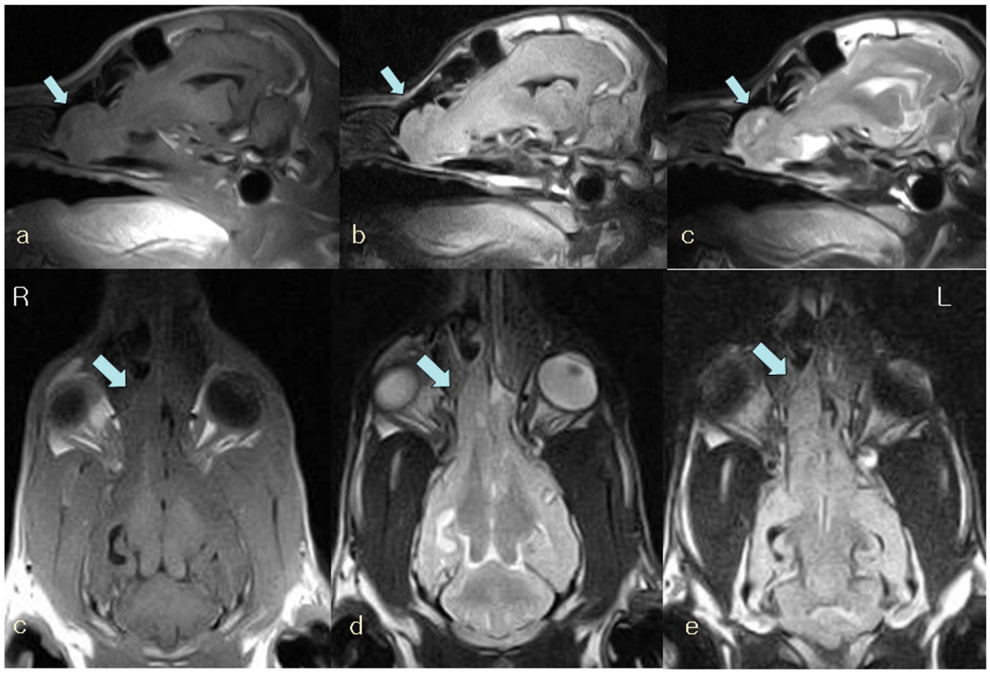

Fig 1. Magnetic resonance images of the head: top row - sagittal plane, and bottom row - dorsal plane. (a, c) T1-weighted images. (b, d) T2-weighted images. (c, e) Fluid-attenuated inversion recovery (FLAIR) images. All images show extrusion of the olfactory bulb into the nasal cavity. T2-weighted and FLAIR images also reveal heterogeneous and hyperintense signals in herniated cerebral tissue.

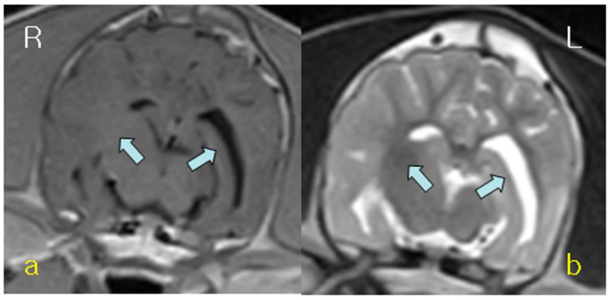

Fig 2. Transverse plane magnetic resonance images at the lateral ventricle level: (a) T1-weighted image. (b) T2-weighted image. The images show the asymmetric lateral ventricle.

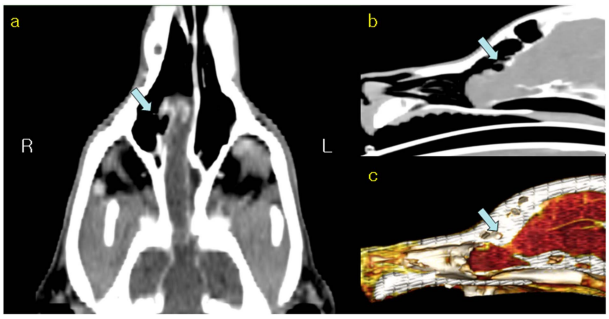

Fig 3. Computed tomography (CT) images. (a,b) Two-dimensional dorsal and sagittal images show the absence of the cribriform plate of the ethmoid bone and extension of cerebral tissue through the bony defect. (c) Three-dimensional sagittal CT image.

could have caused the seizure.

CT of the head, using a helical CT scanner (Hi speed, GE Medical Co, USA), was performed immediately after magnetic resonance examination and used the following acquisition parameters: 120 kVp, 150 mA, 2-mm slice thickness, and a 512 × 512 voxel matrix. CT images of the head showed a defect in the cribriform plate of the ethmoid bone and the extension of the olfactory bulb into the nasal cavity (Fig 3). In addition, shifting of the nasal septum to the left and malformation of endoturbinates in the ethmoidal labyrinth and nasal cavity were observed (Fig 4).

Cerebrospinal fluid (CSF) was collected through the cerebellomedullary cistern to investigate possible intracranial causes of seizure. There were no remarkable findings on CSF analysis.

Based on these findings, the final diagnosis was determined to be ethmoidal encepahalocele.

Phenobarbital (3 mg/kg, every 12 hours) was administered orally for seizure prevention. Oral administration of prednisolone (1 mg/kg, every 12 hours) for 10 days and furosemide (1 mg/kg, every 12 hours) was given for edema. Although there was no seizure activity during 4 weeks after the first treatment, seizures recurred and was continued to worsen.

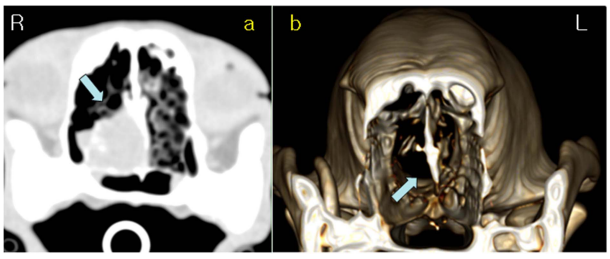

Fig 4. Transverse computed tomography images. (a) Two-dimensional image reveals malformation of the endoturbinates in the nasal cavity. (b) Three-dimensional image shows the nasal septum shifting to the left.

The dog was therefore euthanized at the request of the owner.

Discussion

Encephaloceles are rare and have been reported in only a few cases in the veterinary literature (15,3,11,10,14). In contrast, humans have a relatively frequent incidence of encephaloceles, as high as 1 in 3000-5000 live births, with abundant case reports (16). In addition, the classification of encephaloceles is highly specific, based on whether the protruded contents are limited only to the meninges and cerebral tissue or whether the ventricle is also included, and whether the skull defect is located in the cranial vault, frontoethmoid, occipital, or basal areas (2,7,13,17).

Ethmoidal meningoencephalocele is a type of encephalocele that is characterized as a protrusion of cerebral tissue and meninges through a defect of the cribriform plate of the ethmoid bone caused by a congenital neural tube deformation. In dogs, the herniation of cerebral tissue includes the olfactory bulb and extends rostrally to the nasal cavitiy. It is rarely reported to be connected with facial deformities because the rostral structures are different from those in humans with fronto- and naso-ethmoidal encephaloceles (15,8). Therefore, diagnosis can be delayed despite congenital deformations, and so advanced imaging, like CT or MRI, is important for diagnosis and understanding, especially of ethmoidal meningoencephalocele. In previous reports in the veterinary literature, MR images of encephalocele were characterized by hyperintense signals in the frontal sinuses, consistent with accumulated fluid on T2-weighted scans. Contrast-enhanced images provided no evidence of a breakdown of the blood/brain barrier within the herniated contents (10). However, another report interpreted slight and heterogeneous contrast enhancement of the protruding brain tissue (11). In the human literature, MR images of encephalocele demonstrated hyperintense legions with edema on T2-weighted and FLAIR images and relatively iso- to hypo-intense lesions on T1-weighted images (12). In the dog in this case, the herniated cerebral tissues showed heterogeneous and hyperintense signals with edema on T2-weighted and FLAIR images, and the lesions showed no to mild contrast enhancement. Thus, it was not clear whether the blood/brain barrier within the protruded part of the brain had broken down. There are no cases reported previously in which frontoethmoidal or basal encephalocele was associated with seizure activity, because the most common intractable epilepsy in humans is mesial temporal lobe epilepsy. In contrast, the regions of the brain associated with seizure activity in dogs are the olfactory bulb and frontal lobe (5,9).

Abnormalities of the brain causing seizures are pressure changes in the ventricles or abnormal 'wiring' patterns in juvenile dogs, and subtle changes in brain structures make seizure activity possible. According to a previous report, pathologic changes associated with herniated brain tissue in a juvenile dog were gross hemorrhage, malacia, degeneration of white matter, and inflammatory cell infiltration that could have all contributed to abnormal excitability of cortical neurons (10). Ethmoidal meningoencephaloceles in which the lesion is not only located in the olfactory bulb and frontal lobe but that also causes a change in brain structures has obvious potential for recurring seizure activity in dogs. It can also cause incompetence of the nasal apparatus, as suggested by the history of nasal congestion in the dog in this case. A limitation in this case is that postmortem and histopathologic examinations were not performed. However, ethmoidal meningoencephalocele was diagnosed and thought to cause the seizure activity on the basis of findings from advanced imaging, such CT and MRI, and other examinations.

Conclusion

Ethmoidal meningoencephalocele could be considered as a cause of seizure activity in juvenile dogs.

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