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Case Report

Ethmoidal Meningoencephalocele in an Anatolian Shepherd Dog

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Abstract

Meningoencephalocele is an uncommon disorder in dogs, that has detrimental effects on both the duration and quality of life. Dogs afflicted by this condition may exhibit neurological deficits, epileptic seizures, and behavioral disorders. A 4-month-old female Anatolian Shepherd dog, weighing 13 kg presented for further investigation of a 3-week history of cluster, grand mal seizures. No abnormalities were evident on physical examination. A full neurological examination, including cranial nerve assessment was unremarkable, except a slight right proprioceptive deficit on the right front leg. MRI of the brain was performed. Protrusion of the right rostral rhinencephalon through the defect of the right cribriform plate and a right basal ethmoidal meningoencephalocele was detected. Due to the dog's age, aggressive temperament, and current clinical status, medical management was preferred, and the dog was managed on phenobarbital and levetiracetam. Seizures have remained well controlled 12 months post-diagnosis.

Keywords: Anatolian Shepherd Dog, Cluster seizure, Epilepsy, Meningoencephalocele, Magnetic resonance imaging, MRI

Introduction

In humans encephalocele refers to the abnormal protrusion of cranial contents beyond the normal confines of the skull. They may contain meninges (meningocele), brain matter and meninges (meningoencephalocele) or they may communicate with the ventricles (meningoencephalocystocele) [1]. A meningoencephalocele (MEC) is a protrusion of cerebral tissue and meninges through a cranial defect, whereas a meningocele (MC) is a herniation of the meninges only [2]. MECs protruding into the nasal cavity are named intranasal MECs [3].

This anomaly results from a focal failure of the neuroectoderm and surface ectoderm to separate during fetal development, occurring primarily during the 4th gestational week [4]. Genetic and environmental factors, including toxins, nutritional deficiencies, head traumas, and chronic intracranial hypertension, are identified as potential contributors to acquired cases [5-9]. Additionally, it has been reported that a meningoencephalocele formation occurred in a cat after the transfrontal craniotomy procedure. This situation indicates that this pathology can also develop iatrogenically [6]. In cases of congenital encephalocele, it has been observed that the meninges are often protruded [7].

Clinical presentations in both humans and dogs range from epileptic seizures to behavioral alterations. In the canine population, ethmoidal meningoencephaloceles are associated with diverse neurological signs, including aggressiveness, hyperactivity, and distinctive behaviors like stargazing or fly-catching [8]. Diagnosis necessitates a comprehensive approach involving clinical evaluation and advanced imaging techniques, with MRI being crucial for delineating intracranial compartments and herniated contents. Treatment options vary, with anti-epileptic drugs proving to be effective in some cases, while surgical intervention may be considered for drug-resistant cases or those presenting with cerebrospinal fluid rhinorrhea.

This report aims to comprehensively document and analyze a rare case of ethmoidal meningoencephalocele in a 4-month-old female Anatolian Shepherd dog presenting with cluster grand mal seizures. The investigation delves into the clinical presentation, diagnostic process utilizing MRI, and the subsequent management of the dog. The report emphasizes the challenges posed by the ethmoidal meningoencephalocele in a canine patient,

exploring the intricate relationship between anatomical anomalies, neurological manifestations, and treatment modalities. By providing detailed insights into this rare clinical scenario, the report contributes to the veterinary literature, fostering a deeper understanding of ethmoidal meningoencephaloceles in dogs and guiding clinicians in their diagnostic and therapeutic approaches to similar cases.

Case History

Informed consent was obtained from the animal owner to use the data obtained from the clinical examination.

Clinical Examination

A 4-month-old, intact female Anatolian Shepherd dog, weighing 13 kg, was admitted for evaluation of worsening cluster, grand mal seizures. The dog was adopted 3 months ago, medical history is unknown and was clinically healthy for the last 2 months. The first generalized seizure which lasted around 6 min was followed by 3 more seizures on the same day. No epileptic seizures occurred for the next 3 weeks, until the day before admission. The dog had 5 more grand mal seizures, each lasting around 4 min. During examination, the dog was bright, alert, and responsive. A full neurological examination, including cranial nerve assessment was unremarkable, except a slight right proprioceptive deficit on the right front leg. Complete blood count showed a mild increase in basophil (0.26 x 10^9/L; reference interval [RI] 0-0.1x10^9/L) and eosinophil (1.89 x 10^9/L; RI 0-1.6x10^9/L) counts. Serum biochemistry analysis also revealed a minor increase in ALP (139 U/L; RI 13.0-83.0 U/L) which is consistent with young age and cluster seizures. Serum TSH, total T4 and free T4 values were within the reference range.

Imaging, Diagnosis and Outcome

The dog was sedated with 10 µg/kg-IM medetomidine (Domitor®, Zoetis, Estonia) and 0.1 mg/kg-IM butorphanol (Butomidor®, RichterPharma, Austria) combination. Anesthesia was induced with 0.2 mg/kg-IV propofol (Propofol® 200 mg/20 mL, Abbott, Türkiye) and maintained by an inhalation anesthetic, isoflurane at a concentration of 1% (Forane®, liquid, Abbott, England) in 100% oxygen. MRI of the brain was performed using a 1.5 Tesla MRI system (GE Signa Excite 1.5T). T2-weighted fast recovery fast spin echo (frFSE), fluid-attenuation inversion recovery (FLAIR) in sagittal, dorsal, and transverse planes. T1-weighted fast spin echo (fSE) was performed with 4 mm slice thickness in transverse plane. Post-intravenous gadolinium contrast (0.1 mmol/kg - Gadovist®, Bayer, Türkiye) T1-weighted fat sat fast spin echo sequence (FS-T1-fSE) was obtained with 4 mm slice thickness, in transverse and dorsal planes.

MRI revealed protrusion of the right rostral rhinencephalon through the defect of the right cribriform plate, which was measured approximately 1 cm in diameter. Herniation of the neural parenchyma to the nasal cavity was measured at 22x19x12 mm. Bilateral deep ventricular white matter and the signal intensity of gray and white matter in bilateral cerebral hemispheres were evaluated as normal. The herniation of the neural parenchyma to the right rostral rhinencephalon was consistent with a right

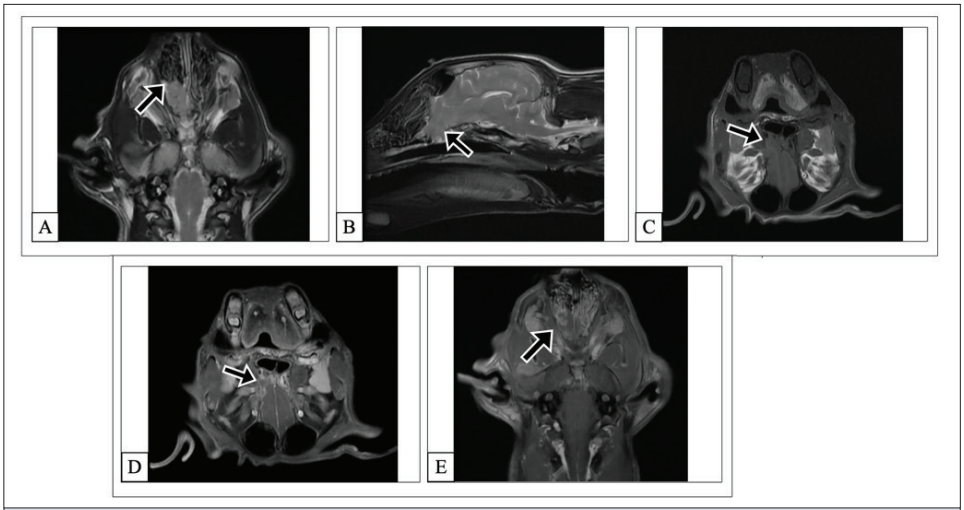

Fig 1. Magnetic resonance images of the dog's brain. **A**- Dorsal fast recovery Fast Spin Echo T2-weighted image. Brain tissue (rhinencephalon) and meninges *(black arrow)* and the absence of the normal structures in the caudal part of the right nose, **B**- Sagittal fast recovery Fast Spin Echo T2-weighted image through the skull. Rhinencephalon is seen in the right ethmoidal conchae *(black arrow)*, **C**- Transverse fast Spin Echo T-1 weighted image pre-contrast at the level of olfactory bulb and rhinencephalon *(black arrow)*, **D**- Post-contrast transverse fast Spin Echo T-1 weighted image with mild contrast enhancement of the meninges (black arrow), **E**- Post-contrast dorsal fast Spin Echo T-1 weighted image, mild contrast enhancement of the protruded meninges *(black arrow)*

ethmoidal MEC. In the post-contrast examination, mild meningeal contrast enhancement was observed around the protruding rhinencephalon tissue in both transverse and dorsal planes in T1- weighted sequences *(Fig. 1)*.

Cerebrospinal fluid analysis was performed after the magnetic resonance imaging process, which did not reveal any abnormalities. The dog recovered uneventfully from anesthesia. Conservative therapy with antiepileptic medication was preferred. Surgical correction of the meningoencephalocele and defect was not performed due to the dog's age, aggressive temperament, and current clinical status. The dog was discharged on the same day of diagnostic imaging.

Levetiracetam (Keppra, UCB Pharma, USA) was prescribed at 20 mg/kg PO q 8 h in combination with phenobarbital (Luminal, Bayer, Türkiye) 2 mg/kg PO q 12 h. 2 days after the patient was discharged, a single generalized epileptic seizure recurred, which was lasted less than 1 min. Phenobarbital and levetiracetam doses were adapted to the dog's changing weight each month and serum phenobarbital concentration was measured at 22.4 µg/mL in the fourth month and 19.7 µg/mL in the 9th month of therapy (range, 15-40 µg/mL). Seizures have been well controlled since last seizure and no seizure activity had been observed for 12 months post-diagnosis of the meningoencephalocele.

Discussion

A focal failure of the neuroectoderm and surface ectoderm to separate during fetal development results in congenital meningoencephalocele formation. In humans, it is considered a late neurulation defect taking place during the 4th gestational week may result in MECs [4]. In cases of congenital encephalocele, it has been observed that the meninges are often protruded [7].

The most common clinical signs were epileptic seizures, behavioral alterations including aggressiveness, hyperactivity, intermittent yelping, stargazing or fly-catching behavior in dogs [8]. In most cases, as outlined by Lazzerini et al.[11], seizures serve as the primary neurological manifestation in dogs with ethmoidal MEC, although it's important to note that seizure features don't consistently seem to align with the location of the defect. Hence, although the underlying causes of seizures in individuals with encephalocele are thought to stem from cortical traction or herniation, additional factors such as hemorrhage, white matter degeneration, or the presence of infectious or non-infectious inflammatory infiltrates may also contribute to abnormal neuronal excitability [12]. Additionally, cases of cerebrospinal fluid rhinorrhea have been documented in dogs with frontoethmoidal encephaloceles [11-13]. In Lazzerini et al.[11]'s research, when examining dogs with frontoethmoidal encephaloceles, neurological abnormalities such as circling, proprioceptive deficits, and reduced menace response were observed. Interestingly, this study revealed that 6 out of 17 dogs with frontoethmoidal encephaloceles displayed normal neurological examination findings.

The diagnosis of encephaloceles necessitates a dual approach, combining clinical assessment with advanced imaging methods. Following the clinical and neurological evaluation, an MRI becomes essential, while a CT scan can provide valuable insights into the craniofacial skeleton's visualization. Additionally, MRI serves the purpose of delineating the communication between intracranial compartments and the herniated contents [12].

Dogs affected by MECs may respond to medical treatment with anti-epileptic drugs [11]. However, the development of drug resistance can serve as an indication for considering surgical intervention [11-14]. In cases where surgery is not performed, the response to anti-epileptic drugs may vary. Notably, some dogs afflicted with frontoethmoidal encephaloceles may eventually be euthanized due to unmanageable seizures [11-12]. Within the Lazzerini et al.[11] study involving 11 dogs treated with anti-epileptic drugs, 2 out of the 11 dogs were seizure-free, while 3 out of 11 experienced a seizure reduction of at least ≥50%.

The only abnormal finding obtained during the neurological examination of the patient presenting with complaints of generalized epileptic seizures is a slight proprioceptive deficit on the right front leg. In a study conducted by Lazzerini et al.[11] in 2017, which included 22 dogs diagnosed with meningoencephalocele or meningocele, lateralized proprioceptive deficits were observed in 9 dogs, while 7 dogs had no abnormal neurological findings reported. Although the pathological mechanism causing unilateral proprioceptive deficit is not fully understood, it is believed to be the result of the mechanical traction effect of the protruding tissue on the remaining brain tissue. In this case, it is observed that the rhinencephalon, which not only transmits the sense of smell but also conveys instinctive and emotional behaviors, is protruding. Despite the patient's young age, the aggressive behavioral pattern exhibited is also thought to be related to the protrusion of the rhinencephalon.

It has been observed that clinical and neurological examination findings are not sufficient for the diagnosis of this pathology, and advanced imaging methods must be included in the diagnostic process. Due to the presence of a calvarial defect in MEC cases, diagnosis can be made not only with MRI but also with CT.

The patient's T1-weighted post-contrast MR images show mild contrast enhancement in the meninges associated with the protruded rhinencephalon tissue. Such contrast enhancement is thought to be potentially due to focal meningitis or low cerebrospinal fluid pressure

and resulting intracranial hypotension. Although no pathology was detected in the CSF analysis of the case, it should be noted that CSF analysis findings may not always be consistent in cases of focal meningitis. It is thought that intracranial hypotension may have developed due to the disruption of intracranial volume balance caused by the protruding brain tissue.

Although the possibility of traumatic MEC formation cannot be ruled out since the medical history of the dog is unknown, the fact that the dog started showing clinical symptoms at a very young age increases the likelihood of this case having a primary neural tube defect etiology.

In human medicine, surgery is considered as first-line treatment for encephaloceles [13]. Surgery was recommended in this case; however, the dog was elected to be managed on anti-epileptic drugs. If refractory epilepsy or CSF rhinorrhea ever develop, surgical intervention is planned right after a follow-up MRI and CT scan.

In conclusion, neither medical nor surgical treatment for MECs in dogs is warranted. Although MECs are rare, it is important to include them as a potential diagnosis when assessing young dogs displaying neurological deficits, seizures, or changes in behavior. Even though surgical treatment is considered as the first-line treatment, no seizure activity was observed in 12 months following the diagnosis and the medical treatment was considered favorable for this patient.

Declarations

Availability of Data and Materials: The data that support the findings of this case report are available from the corresponding author (D. P. A.) upon reasonable request.

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Conflict of Interest: The author declared that there is no conflict of interest.

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