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IMAGING DIAGNOSIS

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MRI diagnosis of spontaneous intraventricular tension-pneumocephalus in a 10-month-old male Saarloos Wolfdog

Stefan Kohl | **Claudia Köhler** | **Ingmar Kiefer**

Small Animal Clinic, Faculty of Veterinary Medicine, University of Leipzig, Leipzig, Germany

Correspondence
Stefan Kohl, Small Animal Clinic, University of Leipzig, Faculty of Veterinary Medicine, 04103 Leipzig, Germany.
Email: stefan.kohl@kleintierklinik.uni-leipzig.de

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Abstract

A 10-month-old male Saarloos Wolfdog was presented with a history of multiple neurologic deficits that had acutely progressed. Neurologic examination findings localized signs to the cerebrum and brainstem. Magnetic resonance imaging revealed markedly enlarged and gas-filled lateral ventricles with a mass effect leading to cerebellar herniation. A right-sided defect of the cribriform plate with a dysplastic ethmoturbinate was identified as the inlet of air and origin of the intraventricular tension pneumocephalus. Surgical findings were consistent with a ruptured, congenital, nasal meningocele.

KEYWORDS

cranial nerve deficits, ethmoidal malformation, nasal cavity, ventriculoscopy

1 | SIGNALMENT, HISTORY, AND CLINICAL FINDINGS

A 10-month-old, 26 kg, male Saarloos Wolfdog was presented via emergency with a subacute history of lethargy and unspecific pain. The therapy of the referring veterinarian with NSAIDs showed no improvement. In the further course, the dog showed reduced consciousness, blepharospasm, intermittent head tilt, and propulsive circling motions. The dog presented proprioceptive deficits of the pelvic limbs since the age of 5 months that progressed to pelvic limb ataxia 3 months before presentation.

Clinical and neurological examination confirmed the reduced consciousness and revealed a sensitivity to touch of the head, deficits in the postural reactions of all four limbs with emphasis on the left side, and cranial nerve deficits, consistent with lesions within the right cerebrum as well as the brain stem. The remaining examination was unremarkable. The main differential diagnoses considering the young age were congenital malformations, in particular hydrocephalus or meningoencephalocele, followed by infectious or immune-mediated meningoencephalitis, traumatic brain damage, and metabolic encephalopathies such as lysosomal storage diseases. Magnetic resonance imaging was recommended for prognosis and treatment planning.

2 | IMAGING, DIAGNOSIS, AND OUTCOME

Magnetic resonance imaging of the brain was acquired with a 3.0T machine (Philips Ingenia 3.0T, Philips Healthcare, Hamburg, Germany) and a 16-channel knee-coil (dStream T/R Knee 16ch coil, Philips Healthcare, Hamburg, Germany). The imaging protocol consisted of 2D T2-weighted turbo spin-echo sequences in three planes (TR 4180–6415 ms, TE 100 ms, 3 mm, no gap) as well as 2D fluid-attenuated inversion recovery (FLAIR) (TR 11000 ms, TE 125 ms, TI 2800 ms, 3 mm, no gap), 2D T2-weighted fast field echo (T2*) (TR 700 ms, TE 16 ms, 3 mm, no gap) and pre- and postcontrast (0.1 mmol/kg administered intravenously, Dotarem®, Gadoteric acid 0,5 mmol/ml, Guerbet GmbH, Sulzbach, Germany) 2D T1-weighted fast field echo (TR 350 ms, TE 4.6 ms, 3 mm, no gap) sequences in the transverse plane.

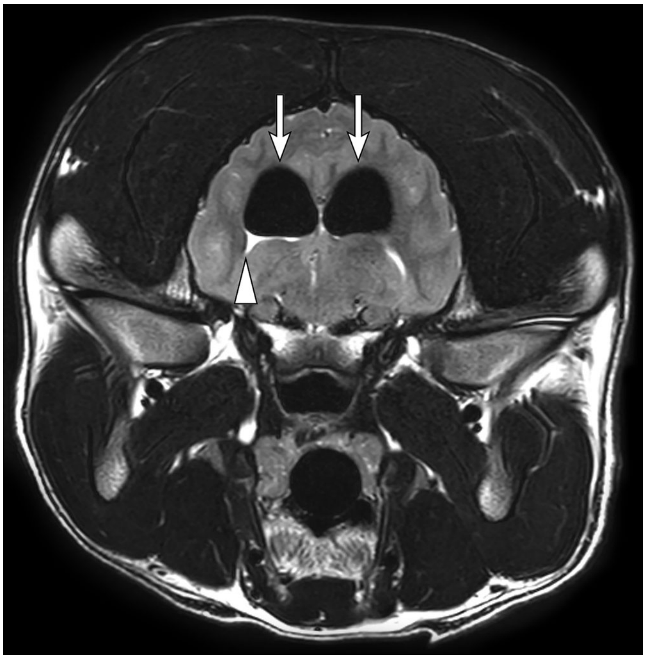

FIGURE 1 Transverse T2w-MRI image of the brain. Marked enlargement of the gas-filled lateral ventricles (arrows). The residual cerebrospinal fluid is apparent ventrally in the lateral ventricles (arrowhead)

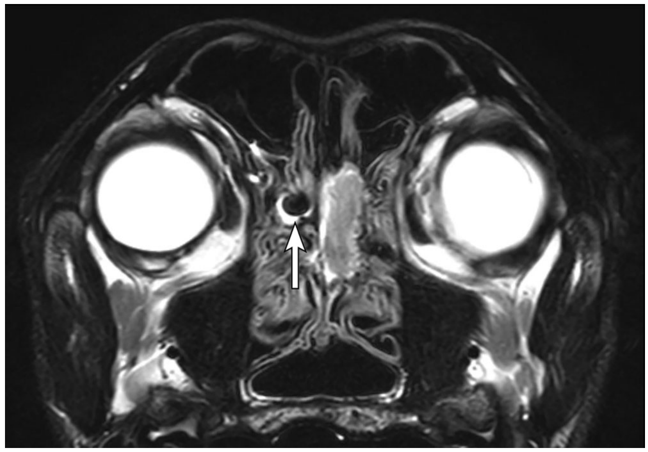

FIGURE 2 Transverse T2w-MRI image at the level of the cribriform plate. On the right side there is a well-demarcated defect of the cribriform plate terminating in a dysplastic ethmoturbinate (arrow). Within that turbinate, a gas bolus surrounded by cerebrospinal fluid is seen

The lateral ventricles were markedly enlarged and hypointense in all sequences, consistent with an intraventricular accumulation of gas. A small amount of T2-hyperintense, fluid-attenuated inversion recovery- and T1-hypointense cerebrospinal fluid remained ventrally in the lateral ventricles (Figure 1). The gas stretched dorsally to the right-lateral frontal bone, where no surrounding brain parenchyma or cerebrospinal fluid could be identified, and rostrally to the cribriform plate, that imposed to have a smoothly marginated defect of 5.7 mm through which the gas reached into the ethmoidal labyrinth where it ended within a dysplastic ethmoturbinate surrounded by fluid similar in signal intensity to that of cerebrospinal fluid (Figure 2). The white matter showed moderate periventricular T2- and FLAIR-hyperintensities, suggestive of edema. Sagittal images revealed marked caudal transtentorial and foraminal herniation of brain parenchyma secondary to increased intracranial pressure (Figure 3). Because brain parenchyma was not definitively observed rostral to the cribriform plate based on MRI findings, a primary consideration was given to a small meningocele that allowed air to communicate between the nasal cavity and the lateral ventricle through a postulated small defect. Therefore, MRI corroborated the main differential diagnosis of a congenital ethmoidal meningocele that led to an intraventricular tension pneumocephalus.

Following diagnostic imaging, the patient underwent an exploratory endoscopic rhino- and ventriculoscopy that confirmed the pathological connection between the nasal cavity and the right lateral ventricle. A partially intact thin membranous layer was observed at the level of the ethmoidal labyrinth without evidence of protruding brain parenchyma, which was therefore consistent with the postulated ruptured meningocele. An effort was made to evacuate the air from the ventricular system but did not show any signs of success. Because of the presence of multiple, severe lesions, limited treatment options, and a poor prognosis, the patient was euthanized without recovery from anesthesia after consultation with the owners.

3 | DISCUSSION

The term pneumocephalus refers to a pathological intracranial accumulation of gas that can be present in the epidural, subdural, or subarachnoid space as well as intraventricular and intraparenchymal.[1–7] First documentations of this condition date back to 1866[8] and 1884.[9] It is a rare diagnosis in human medicine and even rarer in veterinary medicine and in most cases stays asymptomatic or only leads to mild symptoms.[10,11]

In veterinary medicine, intracranial gas accumulation is predominantly described as a complication of craniotomy and rhinotomy as well as following severe trauma,[5,10,12–18] while in human medicine its main cause is trauma, followed by chronic otitis, neurosurgery, and ear nose and throat surgery.[19–22] The distribution pattern of the gas differs between human medicine, where it generally occurs in the subdural space, and veterinary medicine, wherein all cases of clinically relevant pneumocephalus the gas was within the ventricular system.[15]

If there is a steady inflow of gas while the escape is obstructed a tension pneumocephalus builds up with neurological symptoms relative to the degree of compression of the brain parenchyma, up to a compression of the brain stem due to a herniation of the cerebellum through the Foramen magnum.[23] Two mechanisms are described for the development of tension pneumocephalus: First, the “ball-valve effect,” where air enters through a defect when extracranial pressure exceeds the intracranial pressure, that is, while sneezing or coughing, while the discharge is obstructed by the resulting parenchymal pressure.[24] Sec-

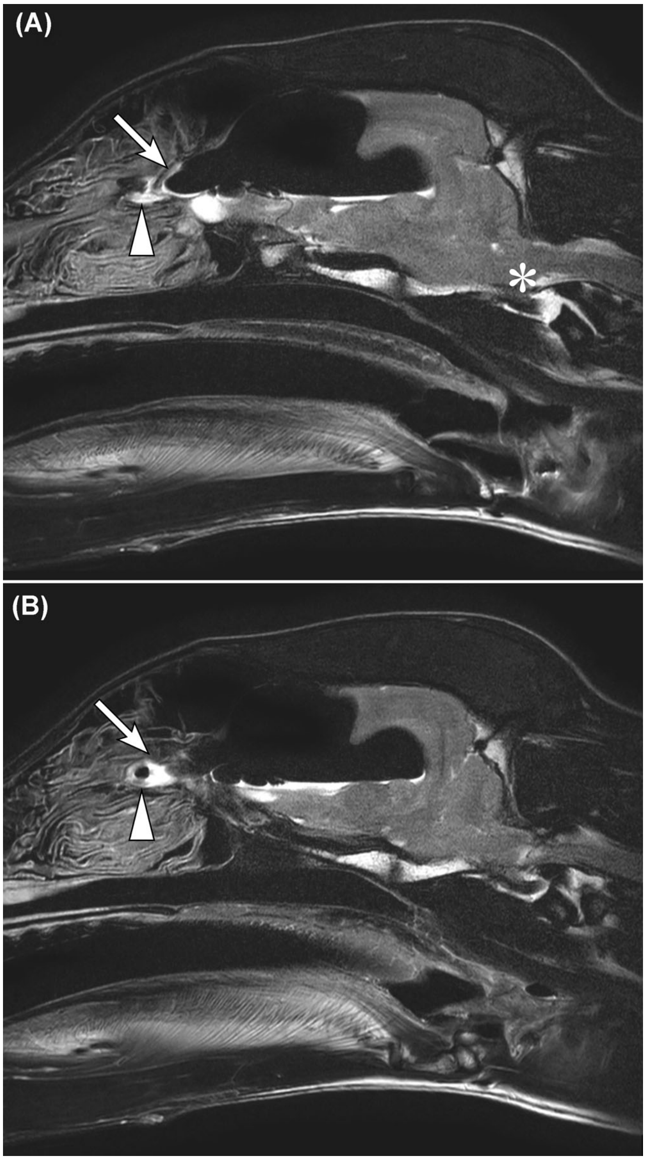

FIGURE 3 Parasagittal T2w-MRI images of the brain. There is a dorso-rostral extension of the gas-filled lateral ventricle. The connection of the intraventricular air with the nasal cavity through the cribrosal defect is clearly visible (arrows). The air ends surrounded by cerebrospinal fluid (arrowhead). There is also a marked herniation of the cerebellum into the Foramen magnum (asterisk, (A))

ond, the "inverted-soda-bottle effect," where a continuous loss of cerebrospinal fluid leads to a negative intracranial pressure, which results in the inflow of air.[25]

In the described case the intraventricular air had a marked mass effect with severe compression of almost all parts of the brain. The underlying mechanism of development cannot be resolved with certainty. The detection of cerebrospinal fluid within the ethmoturbinate would be in line with the description of the "inverted-soda-bottle effect" but rhinorrhea was never observed with the patient. The "ball-valve effect" seems possible with the cribrosal defect only open in phases of increased extracranial pressure and apart from that sealed by brain parenchyma.

A similar case was described by Shea et al., where a 1.5-year-old Border collie with a known nasal meningoencephalocele – the leakage of brain parenchyma through a pathological opening of the cranium[26] – presented with acute left forebrain signs 14 months after initial diagnosis. The control MRI revealed an intraventricular tension-pneumocephalus with a gas-filled tract leading into the nasally herniated brain parenchyma.[27] Considering the similarities between this case and the case presented here, a previously undiagnosed meningocele seems likely as the underlying cause and entry point of the air, given the cerebrospinal fluid surrounding the air at the level of the pathologic opening.

Most cases of pneumocephalus only lead to mild symptoms and thus have no need for therapy or only symptomatic therapy since small volumes of air are reabsorbed within 2–3 weeks.[28,29] The latter includes strict rest, elevated head posture, and avoiding situations with increased extracranial pressure, such as coughing.[11] The presence of a tension pneumocephalus however requires urgent therapy of which the main goal is to decrease the intracranial pressure. Emergency measures include aspiration of air through a detected bony defect or through a drill hole.[11,20,30,31] Eventually, a craniotomy is required to close the dural leak.[14,28,31,32] Therefore, the diagnostic workup of patients with intracranial gas accumulation needs to be focused on the detection of the – inevitably existing – bony defect that allows the inflow of air.

Most of the described veterinary cases of pneumocephalus occurred as complications following craniotomy. Therefore, the main therapy was the surgical exploration of the craniotomy site together with the repair of the initial dural defect.[10,14] Case reports of animals with defects of the cribrosal plate leading to pneumocephalus exclusively describe dogs with chronic rhinosinusitis and following rhinoscopy.[16–18] In only one of those cases a surgical repair of the defect was attempted with a fat-graft and following a progressive postoperative deterioration with a bone- and fascia-graft. Three days later, the patient had gone into status epilepticus and underwent a CT scan revealing a rotation of the bone-graft into the brain, which finally lead to the decision of euthanasia.[17]

The young age at presentation with only 10 months as well as the appearance of the defect within the cribriform plate and the dysplastic ethmoturbinate – most likely resembling a meningocele – strengthens the assumption of it being a congenital malformation with an acute decompensation in the form of a tension pneumocephalus.

This case presents a patient with a tension pneumocephalus, most likely arising from a previously undiagnosed nasal meningocele. Being a rare disease, pneumocephalus should still be included in the list of differential diagnoses of young patients with acute deterioration of neurological state – with and even without known congenital malformations of the skull.

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LIST OF AUTHOR CONTRIBUTIONS

Category 1

(a) Conception and Design: Kohl, Köhler, Kiefer
(b) Acquisition of Data: Kohl, Köhler
(c) Analysis and Interpretation of Data: Kohl, Kiefer

Category 2

(a) Drafting the Article: Kohl
(b) Revising the Article for Intellectual Content: Köhler, Kiefer

Category 3

(a) Final Approval of the Article: Kohl, Köhler, Kiefer

Category 4

(a) Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved: Kohl, Köhler, Kiefer

CONFLICT OF INTEREST

The authors declare that there are no conflict of interest.

ORCID

Stefan Kohl https://orcid.org/0000-0002-1438-3159

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