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SHORT COMMUNICATION

Internal hydrocephalus combined with pachygyria in a wild-born brown bear cub

Anna Kübber-Heiss · Andreas Zedrosser · Georg Rauer · Wolfgang Zenker · Peter Schmidt · Jon M. Arnemo

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Abstract An abandoned wild-born male brown bear (*Ursus arctos*) cub of the year was found and subsequently placed in a zoo. At 7 months of age, the cub showed first signs of ataxia, and at 13 months of age, it was unable to move the hind legs and exhibited outbursts of aggressive behavior and self-mutilation. The animal was euthanized, and necropsy revealed alterations of the brain with obviously flattened gyri, profound enlargement of both lateral ventricles and considerable accumulation of cerebrospinal fluid, disruption of the septum pellucidum, and atrophy of the hippocampus. The animal was diagnosed with an internal hydrocephalus and pachygyria. Genetic evidence showed that the father of the described cub was also the father of the cub's mother, suggesting the possibility of congenital hydrocephalus.

Keywords Brain · Brown bear · Hydrocephalus · Pachygyria · *Ursus arctos*

Introduction

Hydrocephalus refers to an accumulation of cerebrospinal fluid (CSF) within the head as a result of a disturbance of its secretion, circulation, or absorption (Ironside and Pickard 2002). Two basic mechanisms cause CSF to increase in volume: compensatory and obstructive. In compensatory hydrocephalus, CSF increases in volume to take up the space where parenchyma has been destroyed, failed to develop, or both. In obstructive hydrocephalus, CSF accumulates in front of an obstruction to its normal

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A. Kübber-Heiss (✉) · P. Schmidt
Department of Pathobiology,
Institute of Pathology and Forensic Veterinary Medicine,
University of Veterinary Medicine Vienna,
Veterinärplatz 1,
1120 Vienna, Austria
e-mail: anna.kuebber@vu-wien.ac.at

A. Zedrosser
Department of Integrative Biology,
Institute for Wildlife Biology and Game Management,
University of Natural Resources and Applied Life Sciences,
Vienna,
Peter Jordan Str. 76,
1190 Vienna, Austria

A. Zedrosser
Department of Ecology and Natural Resource Management,
Norwegian University of Life Sciences,
Post Box 5003, 1432 Ås, Norway

G. Rauer
Research Institute of Wildlife Ecology,
University of Veterinary Medicine Vienna,
Savoyenstrasse 1,
1160 Vienna, Austria

W. Zenker
Zoo Vienna,
Maxingstrasse 13A,
1140 Vienna, Austria

J. M. Arnemo
Faculty of Forestry and Wildlife Management,
Hedmark University College,
Campus Evenstad,
2418 Elverum, Norway

circulatory pattern or at its site of resorption into the venous system (Summers et al. 1995). Ventricles exposed to this elevation of fluid pressure will be enlarged, compressing and destroying cortical neurons (Summers et al. 1995). Agyria and pachygyria denote macroscopic abnormalities of the cortical surface associated microscopically with a thickened cortical ribbon. Agyria implies absence of gyri and pachygyria and reduced numbers of broadened gyri; the difference is one of degree (Harding and Copp 2002). Hydrocephalus internus is a frequently encountered congenital defect in domestic animals (Axthelm et al. 1981; Grappendorf 1995), and sometimes causes death or obvious clinical signs in calves (Greene et al. 1978) and dogs (Pumarola and van Niel 1992); however, it has rarely been documented in wild animals (e.g., Mandara et al. 2007).

Case report

A lone male brown bear (*Ursus arctos*) cub of the year at an approximate age of 4 months entered a road construction site from the surrounding forest on 25 May 1999 near the city of Mariazell (47.1518 N, 15.1518 E) in Austria. The cub appeared physically weak and constantly approached people; its mother did not appear to be nearby. A game warden tried to release the cub back into the forest; however, the cub returned to the construction site. Due to its weak condition, the cub was fed diluted milk and then placed in a wire-mesh cage in the forest overnight, with the expectation that the mother would return and destroy the cage to retrieve the cub. Because the mother did not return, the cub was considered abandoned and transferred to the Zoo in Vienna the next day. Upon its arrival, the animal weighed 5 kg and was in poor physical condition possibly from malnutrition. After a few days of intensive care, the cub seemed to recover well and quickly increased in both size and mass. At the age of 7 months, minor ataxia in the hind legs became obvious; at the age of 13 months, the animal suddenly was unable to move its hind legs and exhibited outbursts of aggression with vocalization and self-mutilation by biting his own hind legs. Initial treatment consisted of 250 mg prednisolone (Solu-Darcotin®, Fa. Merck KGaA, Darmstadt, Germany) and 100 mg flunixin (Finadyne®, Schering-Plough Animal Health, Boxmeer, Netherlands) administered with a dart from a blowpipe. This caused additional aggressive outbursts with more self-mutilation and tremors in the head and neck area. These aggressive outbursts occurred several times over several hours and continued despite a subsequent treatment of 200 mg prednisolone. Because the symptoms continued until the next morning, the animal was sedated and then euthanized on 19 March 2000 (240 mg tiletamine-zolazepam, Zoletil®, Virbac International, Carros Cedex, France; 0.6 mg medetomidine, Domitor®, Orion Corporation Animal Health, Turku, Finland; and 4,000 mg embutramide, T-61®, Intervet GesmBH, Vienna, Austria). At the time of its death, the animal appeared in otherwise good physical condition, weighing 66 kg at the age of 13 months.

Fig. 1 The flaccid brain with extremely flattened convolutions due to an internal hydrocephalus of a 13-month-old wild-born brown bear. The cerebellum (*white arrow*) appears to be normal. It is visible between the very soft hemispheres

Necropsy revealed a pronounced subcutaneous edema of the ventral body of the bear, potentially caused by prolonged recumbency. In the left thigh, multiple perforations were found in the skin and subcutis ranging from a few millimeters to several centimeters. Beneath the perforations, an extensive subcutaneous phlegmon was observed extending from the hip to the stifle. In the femoral muscles, a necrotic area of approximately 20 cm in diameter and 2 cm thick was noted, and multiple skin-erosions in the pads of feet on both hind legs were found. These lesions were a result of self-mutilation prior to euthanasia. Parenchymatous organs appeared severely hyperaemic,

Fig. 2 Sections of the brain (formalin-fixed) showing severe loss of subcortical tissue and thick cortical ribbon (*white arrow*) in a 13-month-old wild-born brown bear

and the gastrointestinal tract was normal. The skull was not deformed, and no injuries or abnormalities of the skeletal system were apparent. The surface of the brain appeared very smooth, almost all anatomical gyri and sulci were missing, and Rolandic and Sylvian fissures were poorly defined (Fig. 1). On coronal sectioning, the cortical ribbon appeared thicker than normal, and the underlying white matter was markedly reduced or almost inexistent. There was a profound enlargement of both lateral ventricles and the third ventricle, with a striking accumulation of CSF, disruption of the septum pellucidum, and atrophy of the hippocampus (Fig. 2). The fourth ventricle, cerebellum, aquaeductus mesencephali, and whole spinal cord appeared normal. No obstruction of normal CSF drainage was detected. Tissue samples of the brain were fixed in 7% neutral buffered formalin, embedded in paraffin wax, cut at 3 µm, and stained with hematoxylin and eosin. Histologic examination of brain sections revealed a focal loss of flattened ventricular ependyma, thin compressed remnants of cerebral hemispheres, and sponginess of subependymal areas. Occasional neuronophagia was seen in the gray matter of the cerebral cortex. Inflammation was not observed in the brain or leptomeninges. Based on the above findings and on the flaccid brain with a lack of cortical convolutions, the animal was diagnosed with an internal hydrocephalus in combination with pachygyria.

Discussion

Descriptions of hydrocephalus in exotic, zoo, or wild animals are rare (e.g., Halstead and Kiel 1962; Sorjonen et al. 1982; Abramo and Poli 1991). Champoux et al. (1997) reported hydrocephalus as an occasional clinical finding in nonhuman primates with clinical signs such as disturbed consciousness, lethargy/depression, tendency to sleep, ataxia, and blindness. Mandara et al. (2007) described an internal hydrocephalus and associated periventricular encephalitis in a wild red fox (*Vulpes vulpes*) cub; the clinical signs included ataxia, disorientation, and blindness. Sorjonen et al. (1982) found a hydrocephalus in combination with ependymitis and encephalitis in a 7-week-old Asiatic black bear (*Ursus thibetanus*) cub with gross enlargement of the cranium characterized by necrosis and gliosis of the neuropil circumscribing the dilated lateral and third ventricles. Raymond et al. (1998) found an internal hydrocephalus in a 5-month-old American black bear (*Ursus americanus*). The animal experienced seizures, opisthotonus, and nystagmus to the left prior to death, and the hydrocephalus was associated with a paraventriculitis. It is interesting to note that hydrocephalus usually is mechanically caused by obstruction of CSF outflow (Koestner and Jones 1996); however, no obstruction was found in the described case. The clinical symptoms associated with hydrocephalus were also observed in the described case, though not obvious for untrained observers until the animal was 7 months old.

The potential causes of hydrocephalus are many and can include hereditary structural defects, bacterial or viral infections, neoplasia, trauma, parasitic infestation, and nutritional disorders (Bruni et al. 1988; Champoux et al. 1997; Koestner and Jones 1996; Mandara et al. 2007). In the present case, no inflammatory process or other specific cause could be identified.

In addition to an internal hydrocephalus, with uncertain etiopathogenesis, a pronounced lack of cerebral convolutions was observed in the described bear cub. This malformation is associated with breed disposition in dogs such as Lhasa Apso (Greene et al. 1976), Wire-haired Fox Terriers, Irish Setters, and in domestic cat breeds such as Korat (Summers et al. 1995). The developmental mechanism responsible for gyral malformation is unknown; however, a genetic basis is assumed (Summers et al. 1995).

DNA analysis showed that the cub was the offspring of a 10-year-old male, released in an augmentation program in 1993 and a 3-year-old female born in the area (Zedrosser et al. 2004). The male was also the father of the mother of the described cub, and 2 years later, this female successfully raised two cubs fathered by the same male. In brown bears, inbreeding generally is uncommon (Bellemain et al. 2006); however, in very small populations, as in Austria, it may occur more often. It remains unknown if inbreeding in brown bears is associated with abnormalities such as the described case. The evidence suggests a congenital hydrocephalus.

When the bear cub appeared in the road construction site, it already appeared to have been abandoned by its mother. Abandonment of dependent offspring has been described in brown bears (Tait 1980); however, the reasons are often unclear. Possibly the mental and physical handicaps of the cub were already obvious to the mother and caused abandonment.

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