

CLINICAL PRESENTATION, DIAGNOSTIC AND THERAPEUTIC APPROACH OF VERMIAN DYSPLASIA IN A SIBERIAN HUSKY – A CASE STUDY

ASPECTELE CLINICE, DIAGNOSTICUL ȘI ABORDAREA TERAPEUTICĂ A DISPLAZIEI DE VERMIS LA UN HUSKY SIBERIAN – PREZENTARE DE CAZ

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ABSTRACT | REZUMAT

The pathology of the cerebellum includes the cerebellar malformations, which can be inherited or caused by lesions that affect the intrauterine development of the foetus. This paper aims to present the case of a four-month-old Siberian Husky female, which was referred for neurological evaluation due to ataxia, hypermetria, head tremor, and vocalization, which were observed immediately after the puppy was adopted. In order to establish the neurolocalisation of the disease, clinical and neurological examinations were performed. Considering that a cerebellar anomaly was suspected, an MRI scan was requested in order to confirm the diagnostic. Meanwhile, the previous treatment has been optimized and the diet has been improved with elements that support the brain function. The patient was reevaluated in four and twelve weeks after the first examination and then every three months after she started the medication and the diet has been changed. Therefore, the cerebellar syndrome in this case was consistent with diagnostic of cerebellar malformation - vermician dysplasia.

Keywords: neurological examination, cerebellum, vermis malformation, intracranial disease, MRI scan

Patologia cerebelului include malformațiile cerebelare, care pot fi congenitale sau cauzate de leziuni care afectează dezvoltarea intrauterină a fătului. Acest articol are ca scop prezentarea de caz a unei femele de Husky Siberian în vârstă de 4 luni, care a fost adusă la clinică pentru un consult neurologic de specialitate, prezentând ca semne clinice: ataxie, hipermetrie, tremor la nivelul capului și vocalizări, ce au fost observate imediat după ce pacientul a fost adoptat de către proprietari. Pentru a stabili neurolocalizarea leziunii, a fost efectuat un examen clinic și neurologic complet. Având în vedere faptul că a fost suspiciunată o anomalie cerebelară, s-a solicitat efectuarea unui RMN pentru confirmare. Între timp, tratamentul anterior a fost optimizat și dieta a fost îmbogățită cu elemente care să susțină funcțiile creierului. Pacientul a fost reevaluat la 4 și 12 săptămâni după primul consult și, ulterior, la fiecare 3 luni după începerea tratamentului și schimbarea dietei. Așadar, în acest caz sindromul cerebelar a putut fi diagnosticat ca malformație cerebelară, respectiv, displazie de vermis.

Cuvinte cheie: examinare neurologică, cerebel, malformație de vermis, afecțiune intracraniană, examen RMN

The cerebellum is the structure of the nervous system that coordinates muscular activity and controls the rate, range and force of a movement (4, 15). The cerebellar syndrome is one of the most easily recognizable pathology in the veterinary practice. A wide range of pathologies, including vascular, inflammatory, traumatic, anomalous, metabolic, idiopathic disorders and neoplasia, can cause this syndrome. Congenital malformations of the cerebellum are occasionally seen in dogs and they can be inherited or caused by lesions that affect the intrauterine development of the foetus (13). Anecdotal reports have described sporadic cases of vermis hypoplasia / agenesis with or without associated focal or generalized hypoplasia of the cerebellar hemispheres in dogs and the presence of a Dandy-Walker malformation was frequently discussed (1, 3, 5, 6, 7, 8, 11, 14).

The aim of this paper is to present the clinical, neurological and imaging features of a Siberian Husky with presumptive vermician dysplasia. To the authors' best knowledge, this is the first case published in our country that includes the imaging features of an abnormal development of the vermis in a dog.

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CASE PRESENTATION

SIGNALMENT AND NEUROLOGICAL EXAMINATION

The patient, a four-month-old Siberian Husky intact female, was brought to the Clinic of the Faculty of Vete-

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rietary Medicine in Bucharest for a complete neurological consult. The symptomatology consisted of an acute episode of vocalization with an abnormal stance: wide-base stance on both pelvic and thoracic limbs, head tremor, the tendency to bump into walls and furniture, which happened a week before the moment of the examination. During that week, the patient received treatment with diuretics (Mannitol), antibiotics (Clindamycin), barbiturates (Phenobarbital) and vitamins (B1, B6, B12) prescribed by his attending veterinarian, but the progression of the disease could not be stopped. Previously, the puppy was vaccinated and dewormed according to the standard protocol.

In our clinic, we started with a complete physical examination (16) which revealed a normal color of the mucous membranes with a capillary refill time of 2 seconds, a respiratory rate of 20 respirations per minute, a cardiac frequency of 155 beats per minute, synchronic with the pulse. The temperature was 38.2°C and all the palpable lymph nodes were mobile, painless and of normal size. The patient did not expressed pain when the abdomen was deeply palpated.

The physical examination was followed by the neurological examination, which included evaluation of the mental status, posture, cranial nerves, proprioception, gait, spinal reflexes and sensory testing in order to establish the localization of the lesion within the nervous system. The mental status was depressed, with minimum response to the environment and stimuli. The posture was characterized by a permanent lateral decubitus and inability to stand or walk (so the gait could not be evaluated in that moment). Postural reactions assessed were: proprioceptive positioning in which the animal was unable to return his paw to the normal position after it was turned over in all four limbs, visual placing in which the animal reached the table, but expressing hypermetria in all four limbs and extensor postural thrust that reveal a wide-base stance on hind limbs. Abnormal movements were observed –permanent tremor of the head, which intensified when the dog was trying to reach a fixed target. For cranial nerves, we tested the pupillary light reflex, which was normal, the menace response which was absent on both sides, in the cotton ball test the patient eyes followed the objects, the palpebral response and the physiological nystagmus were both present. In addition, a pathological horizontal nystagmus was observed and a mild anisocoria on the right eye (Fig. 1). The pupils show movements of myosis and mydriasis, which follow one another alternatively. The spinal reflexes were normal in all four limbs and the panniculus and anal reflexes were present.

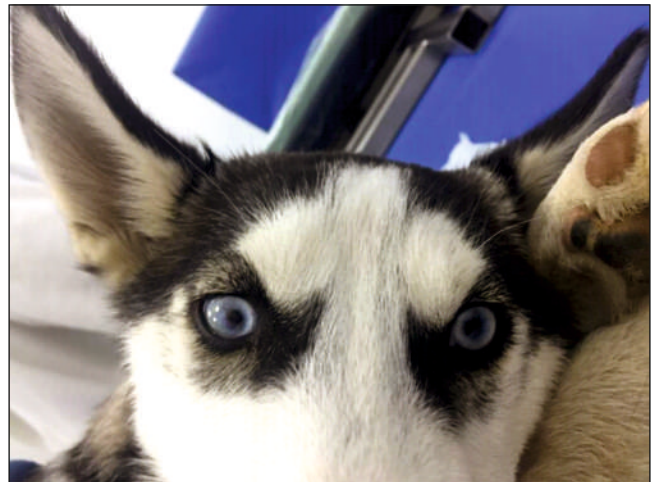


Fig. 1. Head of the patient at the moment of presentation. Permanent decubitus, horizontal nystagmus and a mild anisocoria in the right eye.

After the neurological examination was finished, all the findings have been correlated and the lesion was described as multifocal and localized in the cerebellum and the central vestibular apparatus. A list of differential diagnoses has been taken into consideration using the acronym VITAMIND (Vascular, Inflammatory, Trauma, Anomaly, Metabolic, Idiopathic, Neoplasia, and Degenerative). For the symptomatology of this case, we suspected a vascular, inflammatory, traumatic, anomalous, neoplastic or degenerative pathology, according to the acronym. In order to confirm and to obtain an accurate diagnostic, the owners were informed and agreed to the CNS magnetic resonance imaging (MRI) examination. Before proceeding with the MRI, a cardiological examination was performed and blood was collected for a set of biochemistry and hematology. The results came back normal, so the patient was scheduled for the imaging investigation.

IMAGING

The magnetic resonance examination was performed on the neurocranium with a VET MR GRADE device from ESAOTE with a power of 0.3 Tesla.

Protocols used to obtain images consisting of T1 Spin Eco (SE) and T2 Fast Spin Eco (FSE) sequences in three planes (sagittal, transverse, and dorsal) and post contrast images were obtained in T1 sequences, after intravenous contrast administration. The animal underwent inhalation anesthesia to obtain high quality images and free of motion artifacts.

By interpreting the MRI results, the diagnosis was compatible with a congenital anomaly in the vermis (Fig. 2, Fig. 3, Fig. 4, Fig. 5, Fig. 6 and Fig. 7).

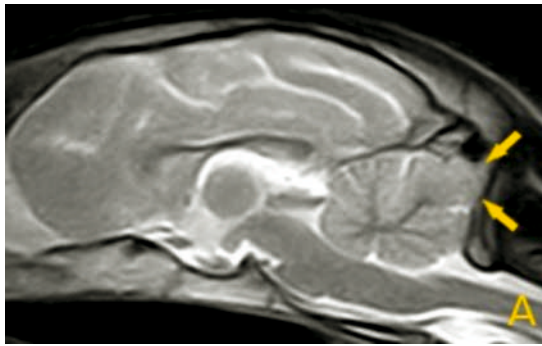


Fig. 2. Mid sagittal T2 plane of a 4-month-old Husky dog brain with a presumptive abnormal development of the vermis (yellow arrows)

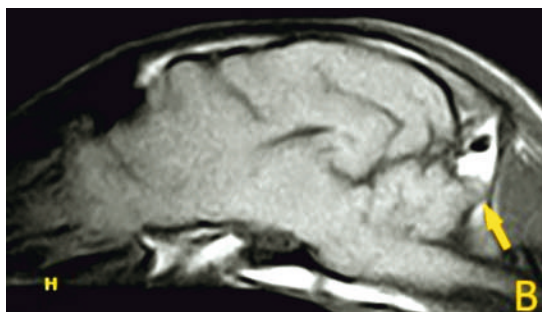


Fig. 3. Sagittal T1 plane (left side) of a 4-month-old Husky dog brain with a presumptive abnormal development of the vermis (yellow arrow)

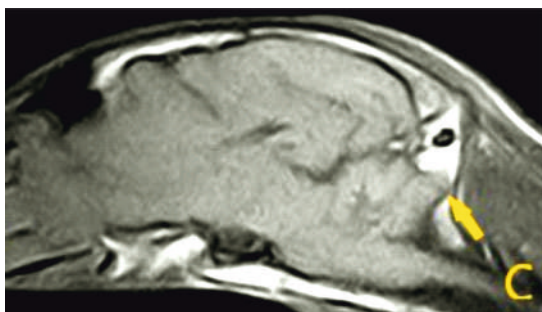


Fig. 4. Sagittal T1 plane (left side) of a 4-month-old Husky dog brain with a presumptive abnormal development of the vermis (yellow arrow), no contrast enhancement.

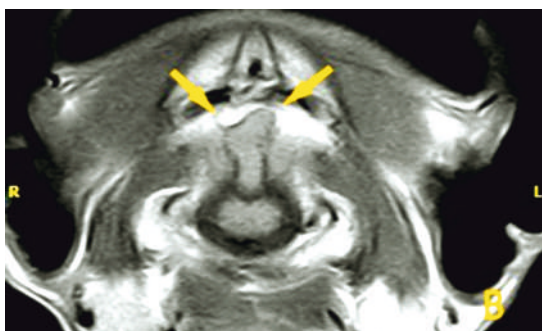


Fig. 5. Transversal T1 plane of the caudal part of the brain, abnormal development and shape of the vermis (yellow arrows)

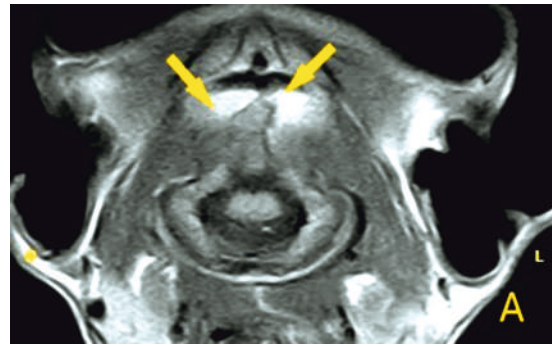


Fig. 6. Transversal T1 plane of the caudal part of the brain, abnormal development and shape of the vermis (yellow arrows), no contrast enhancement.



Fig. 7. Dorsal T2 plane in the ventral side of the brain, abnormal shape of the vermis (yellow arrows).

TREATMENT AND CLINICAL COURSE

The established treatment was palliative and consisted of betahistine dihydrochloride, pills with filtered extract obtained from calf blood, syrup with omega 3 acid and Agaricus. In addition, dietary recommendations included optimized nutrition to support the brain function. The owner was informed that there is no known treatment for this condition, but supportive care may ameliorate symptoms slightly in some cases. He agreed to follow our recommendations.

After four weeks of treatment, the dog came back for reevaluation, when an important progression was noticed. The patient was able to stand and walk, the mental status was normal, alert, the tremor on the head was reduced and the food was no longer dropped from the mouth while eating. Although during gait, hypermetria and a wide-base stance was observed on both thoracic and pelvic limbs, the dog was able to run and to go up and down stairs (Fig. 8). At the following controls, an increasingly good evolution was observed.

Currently, the dog comes every 3 months for control, and it is under an optimized treatment that contains supplements plus a special diet for the brain.

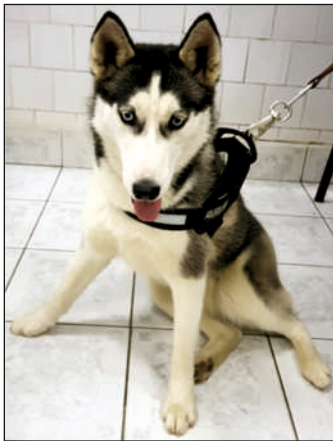


Fig. 8. The patient after 2 months of treatment. The posture improved, the mental status is normal and the pathological nystagmus dissapeared. A wide-base stance can be observed on thoracic limbs.

DISCUSSIONS

In humans, cerebellar vermian dysplasia is one of the many features included in the hereditary cerebellar ataxias, which are a heterogeneous group of neurological disorders (9, 10, 12). Evidence for inherited cerebellar hypoplasia is rare in the veterinary literature. Animals with cerebellar syndrome are easy to recognize because of the specific clinical signs: dysmetric gait, hypermetric wheelbarrowing, head tremor, rolling, cranial nerve deficits and sometimes nystagmus due to paradoxical vestibular disease that may occur with involvement of the flocculus, the nodulus or the caudal cerebellar peduncle (2). Although there is no effective treatment cited in literature for this condition, mild cases can have a fair prognosis, especially if the signs do not progress. In this case, by establishing the correct diagnosis and offering treatment to support brain function, good quality of life for the patient was obtained and the owners were satisfied with the results.

CONCLUSIONS

In the present case report, a cerebellar malformation was suspected in a four-month-old Siberian Husky based on the clinical signs and the neurological examination. MRI imaging confirmed vermian dysplasia. Treatment was instituted throughout life, with the owner's agreement, knowing that it might not be effective. The evolution was favorable, the clinical signs improved and finally a good quality of life was achieved for the patient.

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