The full details of the published version of the article are as follows:

TITLE: What Is Your Neurologic Diagnosis?

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JOURNAL TITLE: Journal of the American Veterinary Medical Association

VOLUME/EDITION: 249/1

PUBLISHER: American Veterinary Medical Association

PUBLICATION DATE: July 1, 2016

DOI: 10.2460/javma.249.1.55

In cooperation with

What Is Your Neurologic Diagnosis?

A 4-year-old 11.4-kg (25.1-lb) sexually intact female Shetland Sheepdog was referred to the University of Wisconsin Veterinary Care neurology service because of sudden-onset ataxia of 12 hours' duration. At the initial evaluation, the dog's abnormalities were limited to the neurologic system.

Neurologic examination

Observation

Mental	Alert		Depressed	X	Disoriented		Stupor		Coma		
Posture	Normal		Head tilt	X	Tremor		Falling				
Gait	Normal		Ataxia	X	Pelvic limbs		All 4	X	Circling		
Paresis	Pelvic limbs		Tetra		Hemi		Mono				
Other	Head tilt to the right.										

Postural reactions

Key: 4 = exaggerated, clonus; 3 = exaggerated; 2 = normal; 1 = diminished; 0 = none; NE = not evaluated

	LF	RF	LR	RR
Wheelbarrow	NE	NE		
Hopping	NE	NE	NE	NE
Ext postural thrust			NE	NE
Proprioceptive pos	2	0	0	0
Hemistand/walk	NE	NE	NE	NE
Placing–tactile	NE	NE		
Placing–visual	NE	NE		

Spinal reflexes

	LF	RF	LR	RR
Quadriceps			2	2
Extensor carpi	2	2		
Flexion	2	2	2	2
Crossed extensor	2	2	2	2
Perineal			2	2

Cranial nerves

	L	R		L	R	Comments CN
II, VII–Vision menace	2	2	VIII–Nystagmus, resting	2	2	Nystagmus (horizontal) became apparent with a change in head posture.
II, III–Pupils resting	2	2	VIII–Nystagmus, change	3	3	
Stim L	2	2	V–Sensation	2	2	
Stim R	2	2	VII–Facial mm	2	2	
II–Fundus	2	2	V, VII–Palpebral flex	2	2	
III, IV, VI–Strabismus, resting	2	2	IX, X–Gag	2	2	
III, IV, VI, VIII–Strabismus, position	2	2	XII–Tongue	2	2	

Sensation (Locate and describe abnormal):

Hyperesthesia	2	
Superficial pain	2	
Cutaneous reflex	2	
Deep pain	NE	

What is the problem? Where is the lesion? What are the most probable causes of this problem? What is your plan to establish a diagnosis? Please turn the page.

Assessment

Anatomic diagnosis

Problem	Rule out location
Depressed mentation	Most consistent with brainstem lesion localization; however, bilateral lesions of the prosencephalon could not be excluded fully.
Right head tilt	Right vestibular system (central or peripheral) or right or left portions of the cerebellum.
Positional nystagmus	Vestibular system (central, peripheral, or cerebellar). Horizontal nystagmus is more consistent with peripheral disease; however, induction with a change in head posture is more consistent with central disease.
Postural deficits right thoracic limb and both pelvic limbs	In combination with the vestibular signs, this is indicative of a central lesion, less likely cerebellar given the complete absence of postural reactions.
Vestibular ataxia	Vestibular system (central or peripheral). Unlikely to have a cerebellar location because of absence of hypermetria, dysmetria, or truncal ataxia.

Likely location of 1 lesion

Right portion of the brainstem affecting the vestibular system.

Etiologic diagnosis—The primary differential diagnoses for this dog included meningoencephalitis (infectious or inflammatory), neoplasia (meningioma, lymphoma, glioma, ependymoma, or choroid plexus neoplasm), acute vascular event, or cyst. The diagnostic plan included a CBC, serum biochemical analysis, urinalysis, thoracic radiography, and abdominal ultrasonography (to evaluate for evidence of compressive, inflammatory, or infectious disease), brain MRI (with and without gadolinium contrast administration), and CSF analysis (to evaluate for inflammatory, infectious, or neoplastic disease).

Diagnostic test findings—Results of the CBC, serum biochemical analysis, thoracic radiography, and abdominal ultrasonography were within reference limits. The dog was anesthetized, and MRI of the brain was performed with and without IV administration of gadolinium contrast agent.[a] Transverse T1-weighted images (before and after contrast agent administration), T2-weighted gradient echo and fluid attenuated inversion recovery (FLAIR) images, and sagittal T2-weighted and sagittal and dorsal plane T1-weighted images after contrast administration were obtained. Within the fourth ventricle, a well-marginated, round, ring-enhancing mass was identified. The lesion measured 9 X 8.5 X 10 mm and was hyperintense on T2-weighted images and hypointense on T1-weighted images (compared with the appearance of gray matter) with heterogeneous signal intensity on FLAIR images (**Figure 1**). The mass displaced the cerebellum dorsally. The lateral ventricles, third ventricle, and mesencephalic aqueduct were moderately distended, consistent with obstructive hydrocephalus. Analysis of a CSF sample (collected at the cerebellomedullary cistern) revealed high total protein concentration (107 mg/dL; reference interval, < 20 mg/dL) and high total nucleated cell count (375 cells/μL; reference interval, < 5 cells/μL). Cytologic interpretation of these findings was neutrophilic pleocytosis. The dog was discharged from the hospital and treated orally every 12 hours with prednisone (0.2 mg/kg [0.09 mg/lb]), ciprofloxacin (10 mg/kg [4.5 mg/lb]), clindamycin (6.5 mg/kg [3 mg/lb]), and fluconazole (4.3 mg/kg [2 mg/lb]). A urine sample was negative for *Blastomyces* antigen; *Cryptococcus* antigen was not detected in a serum sample. Anti-*Neospora caninum* IgG was not present in the CSF sample. Therefore, administration of all medications except prednisone was discontinued. On the basis of the diagnostic imaging findings, signs of inflammatory processes in the CSF, and the lack of positive results following infectious disease testing, a presumptive diagnosis of a cyst with second-

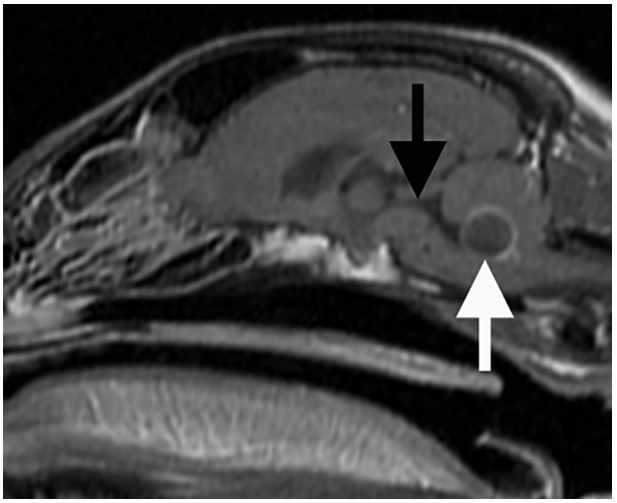

Figure 1—Sagittal plane T1-weighted image (obtained after contrast agent administration) of the brain of a 4-year-old Shetland Sheepdog that had sudden-onset ataxia of 12 hours' duration. Notice the round lesion in the fourth ventricle with mild rim enhancement (white arrow) and mild dilation of the ventricular system rostral to the lesion (black arrow).

ary inflammation and obstructive hydrocephalus was made 7 days following presentation.

Comments

The dog of the present report underwent repeated CSF analysis 5 weeks after the initial diagnosis. At that time, marked improvement in the dog's condition was noted; therefore, the prednisone dosage was gradually tapered and administration was discontinued 10 months after the initial diagnosis. An examination at 16 months after the initial diagnosis revealed abnormalities similar to those identified initially, with the presence of hypermetria of the right thoracic limb suggesting cerebellar involvement. Prednisone had been intermittently administered by the owner but treatment was restarted at a higher dosage (0.4 mg/kg [0.18 mg/lb], PO, q 12 h). Thirty-two months after the initial diagnosis, the dog was found laterally recumbent and was euthanized.

At necropsy, abnormalities were limited to the CNS and liver. Hepatic histopathologic findings were consistent with previous corticosteroid administration. Gross examination of the CNS revealed a gelatinous substance to the right of the cerebellum, consistent with fluid from a ruptured cyst, and a mass lesion on the ventral left aspect of the cerebellar vermis. Histologic evaluation of the mass revealed a central cavity surrounded by a single layer of tall ciliated columnar epithelial cells. These cells were positive for S-100 and cytokeratin and negative for glial fibrillary acidic protein. There were several areas of mild papilliferous epithelium folds, surrounded by vascularized dense collagenous connective tissue attached to the leptomeninges. Large numbers of lymphocytes and neutrophils were present in the folds. The external side of the cyst had papilliferous proliferations of a single layer of cuboidal cells consistent with choroid plexus. The cerebellum was moderately displaced dorsally with a localized area of necrosis and a moderate number of gitter cells within the neuropil. Findings were consistent with an ependymal cyst in the fourth ventricle and cerebellar necrosis.

Ependymal cysts are reported rarely in the veterinary and human medical literature.[1,2] In people, ependymal cysts arise most commonly from the lateral ventricle[3] but have been reported in the fourth ventricle and cerebellopontine angle.[4–8] In humans, ependymal cysts often have no associated clinical signs; however, when patients are affected clinically, the signs reflect supratentorial disease (eg, seizures or motor deficits) or infratentorial disease (eg, cranial nerve VII, VIII, or IX dysfunction).[9] The dog of the present report had central vestibular (cranial nerve VIII) dysfunction initially, with cerebellar signs developing later. As for this dog, CSF abnormalities have been noted previously for a dog with an intracranial cyst and were considered to be secondary to leakage of cystic fluid or compression of surrounding tissues.[10] The dog of the present report had a fair to good prognosis as evidenced by an almost 3-year survival period following diagnosis and intermittent administration of prednisone. Ependymal cysts are rare in dogs; however, an ependymal cyst should be considered a differential diagnosis for dogs with a cystic lesion at the level of the fourth ventricle.

Footnotes

a. Signa Advantage (1.0 T), GE Healthcare, Milwaukee, Wis.

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This feature is published in coordination with the American College of Veterinary Internal Medicine on behalf of the specialty of neurology. Contributors to this feature should contact Dr. Helen L. Simons (800-248-2862, ext 6692) for case submission forms. Submissions will be sent to Dr. Karen Kline, DVM, DACVIM, for her review, except when Dr. Kline is an author.