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Case Report
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Muscular and collagenous cerebellar choristoma in a dog

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ABSTRACT

This report aims to describe the first case of muscular and collagenous choristoma in a dog. A 10-yr-old female mixed-breed dog presented with lateral recumbence, vocalization, positional vertical nystagmus, divergent strabismus, anisocoria, and status epilepticus. The clinical condition evolved to stupor and ultimately, death. Necropsy revealed a white mass causing an irregular increase in the volume of the cerebellar vermis. In histological analysis, a well circumscribed, unencapsulated mass was observed in the cerebellum, consisting of fibers of striated skeletal muscle and collagen fibers, mostly mineralized. Based on the histopathological and histochemical findings, a diagnosis of muscular and collagenous cerebellar choristoma was made.

Keywords: Choristoma; cerebellum; dog; neuropathology; immunohistochemistry

INTRODUCTION

Choristoma is the ectopic presence of histologically normal tissues [1]. Although it is an uncommon condition encountered in veterinary medicine, it is reported to affect all species of domestic animals and known to involve a number of different organs [1,2].

In humans, there are a few reported cases of choristoma, mostly occurring in the nervous system, with preferential involvement of the peripheral nerves [3]. There is one reported case of collagenous choristoma in a bovine, till date [2], however, there are no known reports describing similar lesions in the cerebellum of humans [4] or dogs.

This report aims to present a case of muscular and collagenous choristoma (macroscopic and microscopic features) in the cerebellum of an adult dog as a necropsy finding, and which was not related to the presenting neurological signs.

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Conflict of Interest
The authors declare no conflicts of interest.

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CASE PRESENTATION

A 10-yr-old, female, mixed-breed dog, weighing 30 kg, who was in lateral recumbence, presented with vocalization and episodes of generalized epileptic seizures which evolved over the last 12 h. According to the owner's report, the patient had never experienced epileptic seizures in the past.

On physical examination, the patient had generalized tonic-clonic epileptic seizures and induced vertical nystagmus. The pupils were anisocoric and unresponsive to light. The oculocephalic reflex was reduced, and there was divergent strabismus. Hematological and serum biochemical evaluations were within the normal parameters for the species. Serological analyses for toxoplasmosis and neosporosis revealed negative results.

After interrupting the epileptic seizure with intravenous administration of diazepam (0.5 $mg \cdot kg^{-1}$) (Santisa Laboratório Farmaceutico S/A, Brazil), the patient remained in lateral recumbence and stupor. On the same day, cerebrospinal fluid (CSF) was collected under general anesthesia, which revealed albuminocytological dissociation: 172 mg/dL of proteins and 2.2 mononuclear cells/mm^3 (reference range: < 25 mg/dL and < 5 cells nucleated/mm^3, respectively) [5]. Immunochromatography for detection of canine distemper virus antigen in CSF, as well as fungal and bacterial cultures, revealed negative results.

Immediately before and during CSF collection, continuous intravenous infusion of 20% mannitol (1 $g \cdot kg^{-1}$) (Fresenius Kabi Brasil Ltda, Brazil) and hyperventilation were performed because of a suspected increase in intracranial pressure (ICP) due to mass or inflammation [5]. Approximately 12 h after admission, the condition evolved to coma followed by death.

At necropsy, an irregular white mass was observed, in the vermis region of the cerebellum, which was well-circumscribed, had a sandy surface texture and measured approximately 0.5 cm in diameter (**Fig. 1**). Histological evaluation revealed a well-circumscribed, non-encapsulated mass consisting of striated skeletal muscle fibers interspersed with collagenized fibers, most of which were mineralized, characterizing a focally extensive mineralized collagenous and muscular choristoma (**Figs. 2** and **3**). No lesions outside the central nervous system were found.

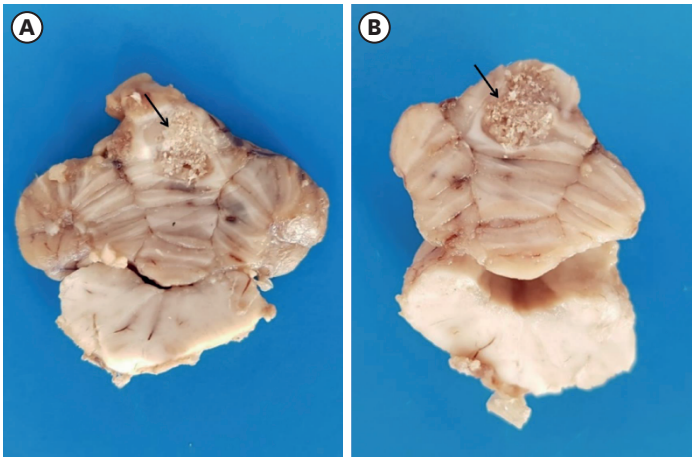

Fig. 1. Muscular and collagenous choristoma in a dog's cerebellum. Macroscopic aspect of the cerebellum on cranial (A) and caudal (B) cut surfaces with a well-circumscribed area of 0.5 cm in diameter, whitish, with a sandy appearance (arrow).

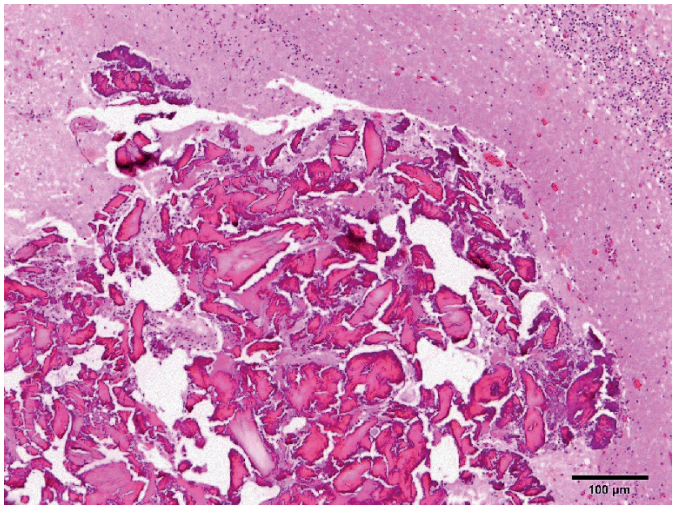

Fig. 2. Microscopic aspect of the cerebellum, showing a focal choristoma consisting of skeletal striated muscle fibers and collagenized fibers, amid the severely mineralized area (hematoxylin and eosin, bar = 100 μm).

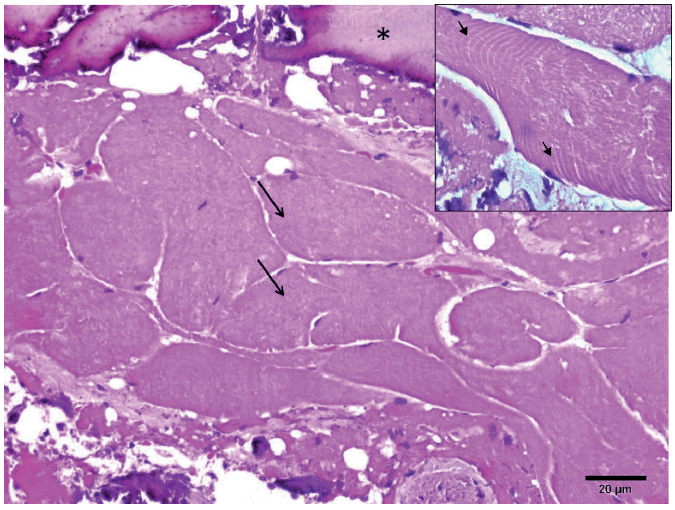

Fig. 3. Cerebellar choristoma. Higher magnification of muscle fibers (arrows) and partially mineralized collagenized fibers (asterisk) in the cerebellum (hematoxylin and eosin, bar = 100 μm). Inset: striations can be seen in muscle fibers (arrows).

Masson's trichrome staining of the histological sections of the decalcified tissue revealed differentiated collagen (stained in blue) and other tissues (stained in red) (**Fig. 4**). In addition, multifocal areas of hemorrhage were found mainly in the molecular, granular, and cerebellar meninges.

In the histology of the thalamus region, areas of moderate to severe multifocal vacuolization were observed, mainly in the perineuronal location, with few interspersed neutrophils and areas of hemorrhage, characteristic of moderate multifocal neutrophilic encephalitis (**Supplementary Fig. 1**). Immunohistochemistry technique for the canine distemper virus (CDV-NP; VMRD, USA) was negative.

It was not possible to determine the cause of encephalitis and encephalic multifocal hemorrhages, which probably triggered the neurological signs observed at the time of the patient's admission to the veterinary hospital.

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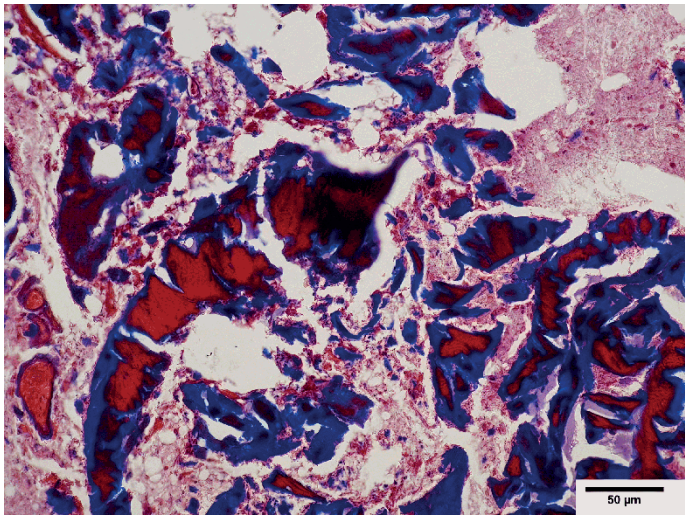

Fig. 4. Cerebellar choristoma. Collagenized fibers are stained in blue and other tissues are stained in red (Masson's trichrome, bar = 50 μm).

DISCUSSION

Choristoma must be histologically differentiated from hamartoma and teratoma [6,7]. A hamartoma consists of disorganized mesenchymal or epithelial tissue, but is located in an anatomically normal position [6]. While the teratoma is formed by disorganized tissue originating from at least two of the three embryonic leaflets (endoderm, mesoderm, and ectoderm), which also has an ectopic location [8]. Choristomas are congenital benign tumor-like growths that can be mistakenly confused with neoplasms [9] and, in most cases, are only diagnosed in adulthood [10]. These are not considered pre-malignant lesions; although there are limited reports on this topic, literature does not suggest further differentiation into a neoplasm [1]. The pathogenesis of choristoma has not been fully understood. It is believed that the process originates from ectopic pluripotent cells, and may contain several cellular elements or predominantly one type of tissue. Furthermore, during the process of organogenesis, some differentiated cells may be removed from their normal tissue structure, which then develop in a nearby location [11].

It is believed that the choristoma in the patient's cerebellum in our report was not the cause of the patient's clinical signs, as it was probably present throughout the dog's life and was incidentally discovered on necropsy. Similarly, the only case of collagenous choristoma in the cerebellum described in veterinary literature was found in a 5-yr-old cow that also did not present with neurological alterations [2].

In light of the macroscopic and microscopic findings of the lesion, a diagnosis of muscular and collagenous choristoma was established, as the presence of collagen in the brain parenchyma is abnormal [2]. Furthermore, on microscopy, the tissues observed were distinguished from neoplastic or metastatic lesions because of the well-differentiated appearance of the ectopic tissue. It believed that the extensive mineralization visualized in histology sections occurred because of the degeneration process of muscle fibers and collagen [12].

As for the acute clinical signs presented in our report, it is known that the risk of developing epileptic seizures associated with encephalitis is poorly understood, but seems to be related

to the cause of inflammation, the degree of cortical involvement, or even the inflammatory response mediated by cytokines [13]. The worsening of neurological signs that culminated into patient's death may have resulted from an increase in ICP, which, according to DeRisio [14], can occur in the case of inflammatory brain disease. Steroid-responsive arteritis meningitis was ruled out due to the absence of arteritis on histology, a characteristic finding of the disease [15].

Although the muscular and collagenous choristoma in the present case is a finding that is not associated with the animal's cause of death, it is important to describe such a lesion which was found at a location that does not match the presenting clinical signs. The description of a choristoma in a dog's cerebellum is important, as this type of anomaly is not considered in the list of possible diagnoses in advanced imaging examinations. It is also important to differentiate between this congenital alteration and a neoplasm, as this greatly impacts the treatment and patient's prognosis.

We conclude that the description of a muscular and collagenous choristoma, even though rare, is important for it to be considered as a possible intracranial finding, especially when the lesion does not match the patient's clinical signs. The description of this type of lesion is also important in comparison with the possible differential diagnoses since it can be macroscopically confused with a neoplastic process.

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SUPPLEMENTARY MATERIAL

Supplementary Fig. 1

Moderate multifocal neutrophilic encephalitis, with mild multifocal vacuolization and moderate hemorrhage (hematoxylin and eosin, bar = 50 μm). Inset: higher magnification of degenerated neutrophils.

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REFERENCES

1. Cullen JM, Page R, Misdorp W. An overview of cancer pathogenesis, diagnosis, and management. In: Meuten DJ, editor. *Tumors in Domestic Animals*. 4th ed. Ames: Blackwell Publishing; 2002, 29.
2. Bianchi MV, Boos GS, Mello LS, Castro NB, Sonne L, Pavarini SP, et al. Collagenous choristoma at the cerebellar peduncle of a Nelore. Semin Cienc Agrar. 2015;36(31):2037-2040.
CROSSREF
3. Spencer A, Lee J, Prayson RA. Optic nerve choristoma. Ann Diagn Pathol. 2005;9(2):93-95.
PUBMED | CROSSREF
4. Welsh CT. Hamartomas and choristomas in the nervous system. Semin Diagn Pathol. 2019;36(1):62-70.
PUBMED | CROSSREF

5. Dewey CW, Da Costa RC, Ducoté JM. Neurodiagnostics. In: Dewey CW, Da Costa RC, editors. *Practical Guide to Canine and Feline Neurology*. 3rd ed. Ames: Wiley Blackwell; 2016, 61-86.
6. Kusewitt DF. Neoplasia e biologia tumoral. In: Mcgavin MD, Zachary JF, editors. *Bases da Patologia em Veterinária*. 5th ed. Rio de Janeiro: Elsevier; 2013, 291.
7. Thompson LD. Tumors of the ear. In: Fletcher CDM, editor. *Diagnostic Histopathology of Tumors*. 4th ed. Philadelphia: Elsevier Saunders; 2013, 2122.
8. Foster RA. Sistema reprodutor da fêmea e glândula mamária. In: Mcgavin MD, Zachary JF, editors. *Bases da Patologia em Veterinária*. 5th ed. Rio de Janeiro: Elsevier; 2013, 1101.
9. Rubin E, Farber JL. Neoplasia. In: Rubin E, Farber JL, editors. *Pathology*. 2nd ed. Philadelphia: J.B. Lippincott Company; 1994,146, 260.
10. Werner PR, Werner J. Avaliação histopatológica. In: Daleck CR, de Nardi AB, Rodaski S, editors. *Oncologia em Cães e Gatos*. 1st ed. São Paulo: Roca; 2008, 127.
11. Young TL, Büchi ER, Kaufman LM, Sugar J, Tso MO. Respiratory epithelium in a cystic choristoma of the limbus. Arch Ophthalmol. 1990;108(12):1736-1739.
PUBMED | CROSSREF
12. Hargis AM, Ginn PE. O tegumento. In: Mcgavin MD, Zachary JF, editors. *Bases da Patologia em Veterinária*. 5th ed. Rio de Janeiro: Elsevier; 2013, 995.
13. Michael BD, Solomon T. Seizures and encephalitis: clinical features, management, and potential pathophysiologic mechanisms. Epilepsia. 2012;53 Suppl 4:63-71.
PUBMED | CROSSREF
14. De Risio L. Structural epilepsy. In: De Risio L, Platt S, editors. *Canine and Feline Epilepsy: Diagnosis and Management*. London: CAB International; 2014, 101-206.
15. Burgener I, Van Ham L, Jaggy A, Vandevelde M, Tipold A. Chemotactic activity and IL-8 levels in the cerebrospinal fluid in canine steroid responsive meningitis-arteriitis. J Neuroimmunol. 1998;89(1-2):182-190.
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