Necrotizing leptomeningeal vasculitis associated with a compressive meningioma in a cat: a rare paraneoplastic syndrome

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Abstract

A 17-year-old cat with a compressive meningioma was found to have an intradural, severe, necrotizing vasculitis, spatially un-associated with the neoplasm. Paraneoplastic vasculitis has been reported in two cases in the human literature associated with meningiomas. This is the first report of such an association in a domestic species.

Introduction

Vasculitis is broadly classified as primary or secondary,1 and, due their relative frequency and clinical significance, a specific classification system exists for categorizing primary vasculitides in humans. They remain, however, largely idiopathic in nature.2,3 Secondary vasculitides are generally associated with an infectious agent, or, non infectious causes such as autoimmune conditions or direct drug interactions.3 One uncommon association with secondary vasculitis is malignancy. Paraneoplastic vasculitis, most usually associated with hematological malignancies such as leukemias, is most commonly cutaneous in manifestation.4 Cases of concurrent vasculitides associated with solid neoplasms are less well described - when reported, these are typically pulmonary, prostatic, colonic, renal or breast related carcinomas.5,6 Vasculitis associated with malignancy of the nervous tissue is extremely rare. In the case of meningiomas, two cases have been reported in the human literature: a single report associated with a subdural spinal meningioma,7 and a second case involving an intracranial meningioma.8 The veterinary literature tends to classify vasculitis in a fundamentally different manner, the principal categories being infectious versus non infectious.1 Of these, infectious causes are by far more numerous and include a number of significant diseases commonly encountered in veterinary species. In the cat, for example, Feline Corona Virus classically manifests as a vasculo-centric inflammatory response.9 Non infectious causes of vasculitis in animals are relatively poorly defined, however, a number of well known conditions are described which have analogues in the human literature. These include beagle pain syndrome, which is well described. Beagle pain syndrome is often associated with anti-neutrophil cytoplasmic antibodies (ANCA) and is considered to be similar to the human condition, Kawasaki disease.10-13 The closely related steroid responsive meningitis / arteritis has many characteristics of an immune mediated disorder, however, it is considered idiopathic.14,15

To date, there has been no publication in domestic or companion animals of vasculitis associated with malignancy.

Case Report

A 17-year-old domestic short haired cat presented to the Atlantic Veterinary College with a two day history of lethargy, weakness, anorexia and head pressing. The animal had a 4 year history of hyperthyroidism, which had been successfully controlled. During physical examination, anisocoria was noted, with left pupillary dilation. The animal continually paced, was non responsive to external stimuli, lacked a menace response on the left side, and failed to demonstrate a left sided direct or consensual papillary light reflex. Additionally, there were significant proprioceptive deficits in all four limbs and a lack of placing responses in the forelimbs. A preliminary diagnosis of cerebral hemorrhage, thromboembolism or central nervous neoplasia were all considered differential diagnoses, and the owners elected euthanasia.

At necropsy, the animal was in reasonable body condition, and minimal findings were found outside of the nervous system. There was a 2 cm diameter soft tissue protrusion on the ventral surface of the left piriform lobe, with compression of the cerebral cortex. At the level of the caudal brain stem and proximal cervical spinal cord, there was a focally expansile subdural hemorrhage. Histologically, the mass on the piriform lobe consisted of a focally expansile mass of spindle to irregular shaped cells, forming irregular whorls and bundles. The cells were moderately variable, with no mitoses noted. At the margins of the mass, there was a distinct zone of compression of the cerebral cortex, and a focal line of hemorrhage. The findings were consistent with a meningioma. The brain stem and spinal cord junction contained an extensive zone of subdural hemorrhage, associated with severe fibrinoid necrotizing vasculitis of the dural blood vessels (Figure 1). The tunica intima of multiple vessels was replaced by a retraction, fibrinoid band. The remnant tunica media and adventitia, and surrounding connective tissue, were significantly infiltrated with abundant neutrophils, lymphocytes and plasma cells with lesser macrophages. Immunohistochemistry was performed for Feline Corona Virus, Toxoplasma gondii, Rabies and West Nile Virus antigens, with no positive findings.

Discussion

The presentation of this case, clinically, was strongly supportive of a focal, centrally located lesion, and was consistent with the demonstrated meningioma. The acute nature of presentation, however, was somewhat unusual. There was no evidence nor history of any traumatic incident. It is likely that the severe necrotizing vasculitis and focally extensive subdural hemorrhage had a significant effect on the presentation. The most commonly occurring infectious cases were excluded, and commercially available FIV and FeLV tests were found to be negative.

The vascular changes were histologically similar to those found in cases of idiopathic canine polyanerteritis, which frequently presents with cranial cervical cord distribution.12 The advanced age of this cat, however, was in distinct contrast to the distribution of that condition, which is more common in far younger dogs.14 The presence of severe vasculitis, spa-
tially at some distance to the meningioma, was considered to be consistent with a paraneoplastic origin.

Within human medicine, paraneoplastic vasculitides are uncommon, accounting for less than 5% all cases. The majority of cases are found concurrently with hematological malignancies; however, solid malignancies are described. The presence of vasculitis associated with a meningioma has been reported in two single case reports in the human literature, and as such, is exceedingly rare. In the two human cases, there was no spatial relationship between the tumours and the location of vasculitis. In the first case, an intracranial meningioma was associated with vasculitis of the aortic arch. In the second, a spinal benign meningioma located at T1 was associated with vasculitis of the coronary artery.

Paraneoplastic conditions are not unknown in the veterinary literature. Recently, myokymia was reported in association with an intracranial meningioma. Vasculitis, however, in conjunction with a solid neoplasm has to the knowledge of the authors, not been reported in any non human species.

References