

CLINICAL, MAGNETIC RESONANCE IMAGING, AND HISTOPATHOLOGIC FEATURES OF A HYPOGLOSSAL MALIGNANT PERIPHERAL NERVE SHEATH TUMOR IN A MALTESE DOG

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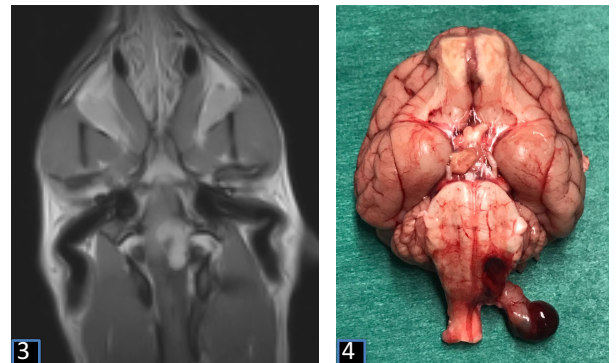
CASE DESCRIPTION

A three-year-old male Maltese dog presented with a one-week history of occasional neck pain and progressive tetraparesis. On neurologic examination the dog was obtunded and showed left pleurothotonus, vestibular ataxia, left proprioceptive deficits, and left eye ventrolateral strabismus. In addition, a left lingual hemiatrophy with ipsilateral deviation was evident (Fig. 1). Based on these findings, the neuroanatomical localization was consistent with a left brainstem lesion and the main differential diagnoses were neoplastic and inflammatory diseases.



DIAGNOSTIC IMAGING

Magnetic resonance imaging of the brain showed a well defined extra-axial mass (approximately cm. 1.5x2.0x0.6) located on the left side of the caudal fossa compressing the brainstem. Additionally, the mass extended extracranially through the hypoglossal canal as a tubular dumbbell-shaped mass. The mass was hyperintense on T2W images, isointense on T1W images, strongly and homogeneously enhancing after administration of contrast medium, except for a core of presumed necrotic tissue (Fig. 2 and 3, transverse and dorsal T1W p.c., respectively). Because of the poor prognosis the dog was euthanized on owner's request.



NEUROPATHOLOGICAL FINDINGS

At macroscopic examination, the mass appeared haemorrhagic, compressing the ventral surface of the medulla oblongata (Fig. 4). It extended into an enlarged hypoglossal canal assuming a tubular, pink-grey appearance. The left hypoglossal nerve was enlarged, with a multinodular appearance (Fig. 5). The most proximal part of the lesion was diffusely haemorrhagic and was responsible for the brainstem compression (Fig. 4). On transverse section, the left half of the brainstem revealed a large necrotic-haemorrhagic area extending from the floor of the fourth ventricle to the pyramis (Fig. 6). At histologic examination, the hypoglossal nerve was diffusely infiltrated by fusiform neoplastic cells arranged in Antoni A and Antoni B patterns (Fig. 7, EEx20). Nuclear atypia and pattern loss were evident at a high magnification (Fig. 8, EEx40) with up to 4 mitotic figures/HPF. Large areas of necrosis were present. Neoplastic infiltration did not occur neither in the brainstem, nor in the tongue. The histologic diagnosis was left hypoglossal MPNST associated with an haemorrhagic and necrotising lesion of the brainstem consistent with chronic compression.

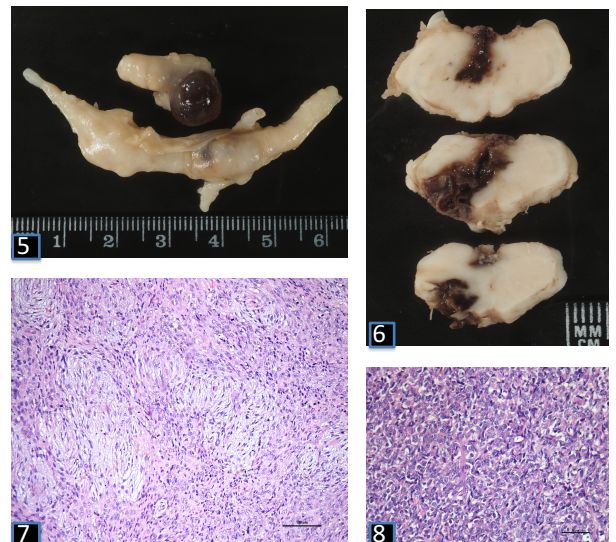


Fig. 1-8: See text

DISCUSSION

Malignant tumours arising from peripheral nerves or displaying differentiation along the lines of the various elements of the nerve sheath are collectively referred to as MPNSTs. Histologically, in human MPNST, the malignant nature of these tumours is associated with an infiltrative and cellular proliferation of atypical, mitotically active spindle cells. Their predilection for sites varies with the species. In dogs the most common site for MPNST is the nerve roots or nerves of the brachial plexus, while the cranial nerve more frequently involved is the trigeminal nerve. In humans, hypoglossal peripheral nerve sheath tumours are rare, only few cases are described in literature; and the malignant forms are even exceptional. Fifty per cent of cases displayed both an intra- and extracranial component, and are referred as dumbbell-shaped tumours. To our knowledge this is the first case report an hypoglossal MPNST in a dog. In our case the tumour was dumbbell-shaped, with both intra- and extracranial components and associated with characteristic clinical signs of hypoglossal tumour. The malignant histological appearance of the lesion occurred without infiltrative growth.