30th Annual Symposium of the European Society of Veterinary Neurology

Helsinki, Finland - September 2017, 21th-23th

CLINICAL, MAGNETIC RESONANCE IMAGING, AND HISTOPATHOLOGIC FEATURES OF HYPOTHALAMIC NEURONAL HAMARTOMA IN A YOUNG VIZSLA

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CASE DESCRIPTION

A one-year-old female Vizsla presented for progression of neurological signs noted since adoption at 3 months of age. Neurological examination showed depressed mental status, compulsive pacing, left head tilt and circling, positional horizontal nystagmus, left eye positional ventral strabismus and ipsilateral proprioceptive deficits. Every 2 weeks left continuous compulsive circling lasting 48 hours. A left brainstem and/or forebrain localization was suspected. Differential diagnoses included congenital anomaly, neoplastic and inflammatory lesion.

DIAGNOSTIC IMAGING

Magnetic Resonance Imaging (MRI) of the brain showed an ill-defined intra-axial mass in pituitary and hypothalamic regions. The pituitary fossa appeared enlarged and flattened and the pituitary gland was right laterally displaced. The mass was mildly and heterogeneously hyperintense to gray matter on T2-weighted images (Fig.1,2), isointense to gray matter on precontrast T1-weighted images. No postcontrast enhancement (Fig.3). The owner refused any further investigation and elected euthanasia.

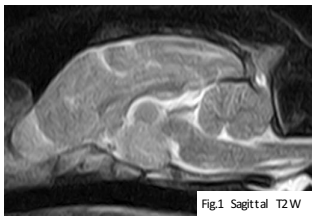
Fig.1 Sagittal T2W

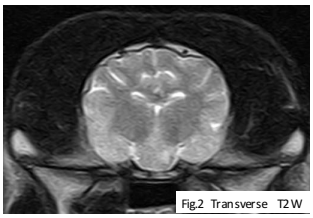
Fig.2 Transverse T2W

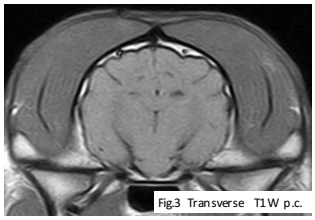
Fig.3 Transverse T1W p.c.

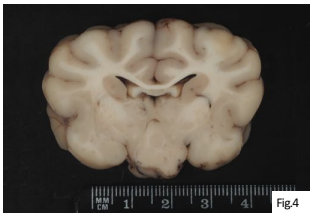
Fig.4

NEUROPATHOLOGICAL FINDINGS

After removal of the brain, the pituitary fossa appeared enlarged and both cavernous sinuses were clearly visible (Fig.5). A symmetric mass resembling normal cerebral tissue covered the ventral surface of the hypothalamus, caudally to the optic chiasm, and mesencephalon. The pituitary gland was located into the rostral border of the mass (Fig.4,6).

Histologically, the mass was composed of abnormally distributed normal neurons and glia, with a prevalence of neuronal elements. LFB-PAS stain confirmed the region was disorganized. Discrete nodular foci of abnormally distributed neurons interspersed with glial cells were present, with a few large ganglion-like ballon cells (Fig.7). Compared to the hypothalamic area of a healthy 12-month-old dog, the GFAP-positive glial cells were scattered most diffusely. Synaptophysin immunoreaction revealed a more crowded network of synapse-associated proteins. Immunohistochemistry for phosphorylated neurofilaments revealed disorganised tracts of axons (Fig.8). Based on histological findings, a hypothalamic neuronal hamartoma was diagnosed.

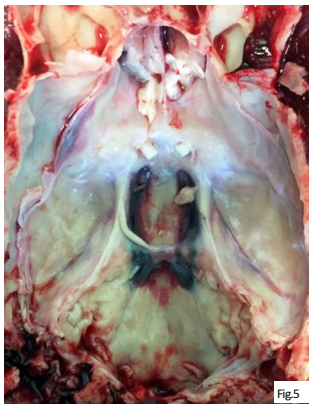
Fig.5

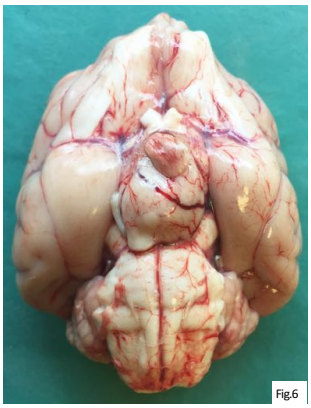
Fig.6

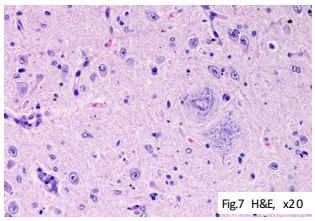
Fig.7 H&E, x20

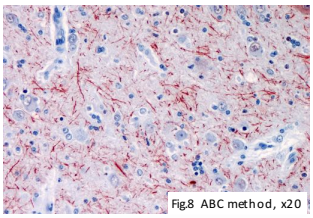
Fig.8 ABC method, x20

DISCUSSION

Hypothalamic hamartomas (HH) are rare, tumor-like malformations that occur during fetal development and are present at birth. They differ from neoplasms since they are not autonomous and they grow in proportion to normal brain growth, and consequently their relative size to the rest of the brain is the same for the lifetime of the patient. Hamartomas are non-progressive lesions and do not expand, spread or metastasize to other locations. In canine nervous system, vascular, neuronal and peripheral nerve fibers hamartomas have been described; to our knowledge, this is the first report describing the MRI features of a hypothalamic neuronal hamartoma in a dog.