ACVR CT/MRI Society – Case of the Month
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This month’s case was provided by:
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History and Physical Exam Findings:
- 7 year old male neutered Bichon Frise
- 6-week history progressive vestibular signs, right-sided blindness, and behavioral changes. Vestibular signs temporarily improved after a dose of mannitol administered by an emergency clinician. One day prior to presentation, the patient developed a dull mentation and nystagmus.
- On physical exam the patient had a dull mentation. No other abnormalities were identified.
- On neurologic exam, the patient was dull but responsive, had a minimally ambulatory tetraparesis with a vestibular ataxia, and decreased hopping in all four limbs (worse on the right) with intact withdrawal reflexes. Cranial nerve exam revealed anisocoria (OS>OD), an inconsistent menace (particularly on the right), positional vertical nystagmus, ventral strabismus OD, and a right-sided head tilt.
- Neuroanatomic localization was right-sided cerebellovestibular with probable prosencephalon involvement given behavioral changes.

MRI Imaging Technique:
MRI System: 3.0T magnet (MAGNETOM Skyra, Siemens Medical Technologies, Malvern, PA)

Sequences (Brain):
- Sagittal T2W-imamges (TR 3000 ms, TE 102 ms, 2.5 mm slice thickness)
- Transverse T2W-images (TR 4200 ms, TE 93 ms, 2.5 mm slice thickness)
- Transverse T2W-fluid-attenuated inversion recovery (FLAIR) images (TR 8000 ms, TE 91 ms, 2.5 mm slice thickness)
- Transverse T1W-fluid-attenuated inversion recovery (FLAIR) images (TR 2100 ms, TE 9 ms, 2.5 mm slice thickness)
- Transverse Susceptibility Weighted Images (SWI) (TR 28 ms, TE 20 ms, 1.5 mm slice thickness)
- Transverse Diffusion Weighted Images (DWI) including ADC map (TR 6700 ms, TE 130 ms, 3.0 mm slice thickness)
- Transverse Proton Density (PD) with and without fat suppression (TR 2700 ms, TE 25 ms without and 44 ms with fat suppression, 3.0 mm slice thickness)
- Contrast: No intravenous contrast agent was administered.
**MRI Findings:**
Centered in the diploë of the left parietal and frontal bones, there is a well-defined expansile lesion that on T2W images is heterogeneous and predominantly hypointense, and on T1W images is homogeneous and mildly hypointense to skeletal muscle. The mass has a smooth, convex ventral margin and is associated with moderate compression of the left parietal and temporal lobes and left side of the thalamus, and mild compression of the left frontal lobe. The mass extends slightly to the right of midline within the diploë, and is associated with a mild leftward midline shift, transtentorial herniation, and caudal herniation of the cerebellum through the foramen magnum. Along the left dorsolateral aspect of the mass there is moderate locally extensive lysis of the outer table of the left parietal bone with extension of T2W-hypointense and hyperintense tissue into the left temporal muscles creating a mild mass effect. Both the mass and tissue extending into the temporal muscles are associated with strong, heterogeneous signal voids on the susceptibility weighted images. In the right parietal region, thickening of the T2W-hyperintense interface between the brain surface and calvarium is present, suggestive of meningeal reaction or involvement.

In the included cranial cervical spine, there is ill-defined T2W-hyperintensity within the cord dorsal to the central canal which is attributed to pre-syrinx edema formation secondary to cerebellar herniation.

The left medial retropharyngeal lymph node is mildly enlarged and peripherally hypointense in all sequences.

Based on the extra-axial location centered within the diploë, a primary bone neoplasm was prioritized (e.g. multilobular osteochondrosarcoma, osteosarcoma, fibrosarcoma, chondrosarcoma, hemangiosarcoma). A non-neoplastic etiology such as fungal osteomyelitis (e.g. blastomycosis) was considered less likely.
Case Outcome:
While under general anesthesia for imaging, the patient was hemodynamically unstable and had been given several doses of mannitol due to suspected elevated intracranial pressure. Given the poor prognosis, the owners elected humane euthanasia. A necropsy was permitted.

Diagnosis:
Extra-axial primary bone hemangiosarcoma (left parietal bone)
No evidence of distant metastasis

Grossly, firm attachments were present between the mass and adjacent meninges. There was histologic evidence of neoplastic cells bridging the dura and lining the subdural surface. Diagnosis of hemangiosarcoma was confirmed by positive immunoreactivity for Factor VIII related antigen on IHC staining.

Discussion:
Hemangiosarcoma (HSA), a malignant neoplasm of vascular endothelial origin, is commonly reported in the spleen, right atrium, skin, subcutis and liver. Primary bone HSA is however rarely reported, and accounts for less than 5% of all bone tumors. Several case reports describe primary vertebral HSA with extradural compression, however primary HSA of the skull is infrequently reported.

While intravenous contrast was not administered in this case, this could have been helpful to prioritize HSA, as a retrospective study of CT features of non-parenchymal HSA found that the presence of highly attenuating post-contrast foci (consistent with vascular channels) was a possible distinctive characteristic.

The prognosis for osseous HSA is poor, with <10% probability of surviving 1 year even if HSA is confined to one bone and completely surgically resected.
References:


