Diagnostic Exercise

From the CL Davis/SW Thompson Foundation

Case :**265**; Month: **July**; Year: **2024**

Title: Congenital vascular hamartoma in the spinal cord of a neonatal foal

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Clinical history: A three-day-old Arabian filly with a history of not being able to stand unassisted after birth and lack of control over the right front and right hind limbs. The filly had inadequate proprioceptive responses, with absent pain/withdraw answers, and failed to respond to medical therapy. Due to the poor prognosis, euthanasia was elected, and a full necropsy was performed.

Necropsy findings: Paramedial section of the whole spinal canal revealed spindle shape, dark-red, friable masses measuring ~ 8 cm and 4 cm long at the level of C4-C5 and T7-T8, respectively. These masses invaded approximately half of the spinal cord diameter, which was ~ 25% thicker than normal (Fig 1).

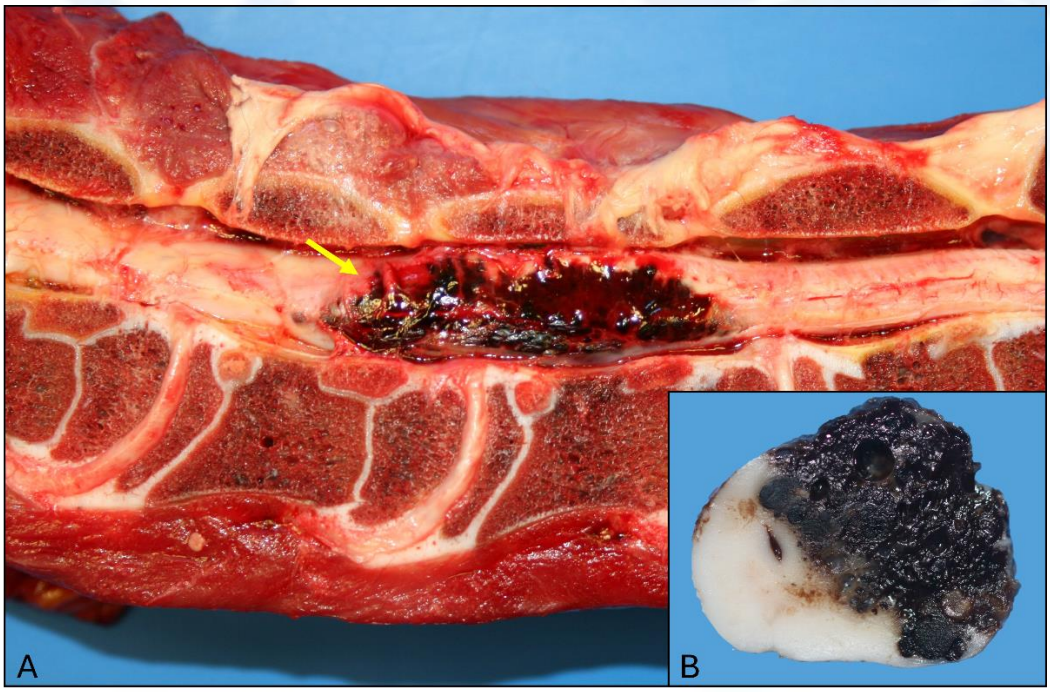

Figure 1. Longuitudinal section of the cervical spinal cord. **A.** There is a dark-red friable mass effacing/replacing the neuroparenchyma (arrow). **B.** Transverse section of spinal cord. Focally extensive mass with multiple cavities, involving more than 70% of the tissue (this specimen was fixed in formalin before being photographed).

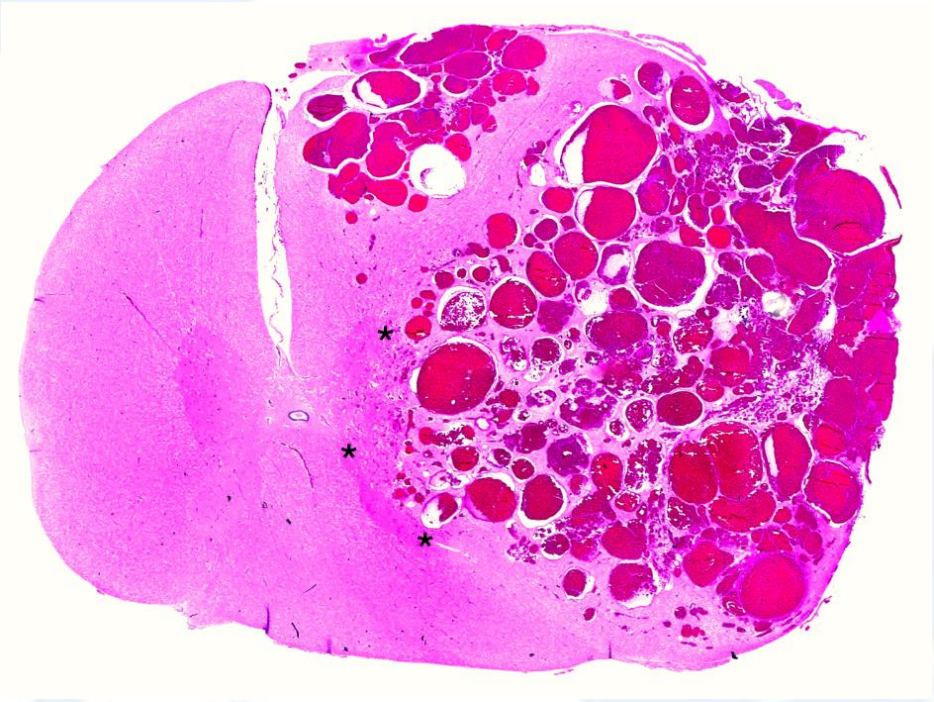

Figure 2. Spinal cord, subgross view. A focally extensive, infiltrative, and non-encapsulated vascular proliferation effacing aproximately 70% of the neuroparechyma is observed. The mass compreses the surrunding grey and white matter (asterisks). HE.

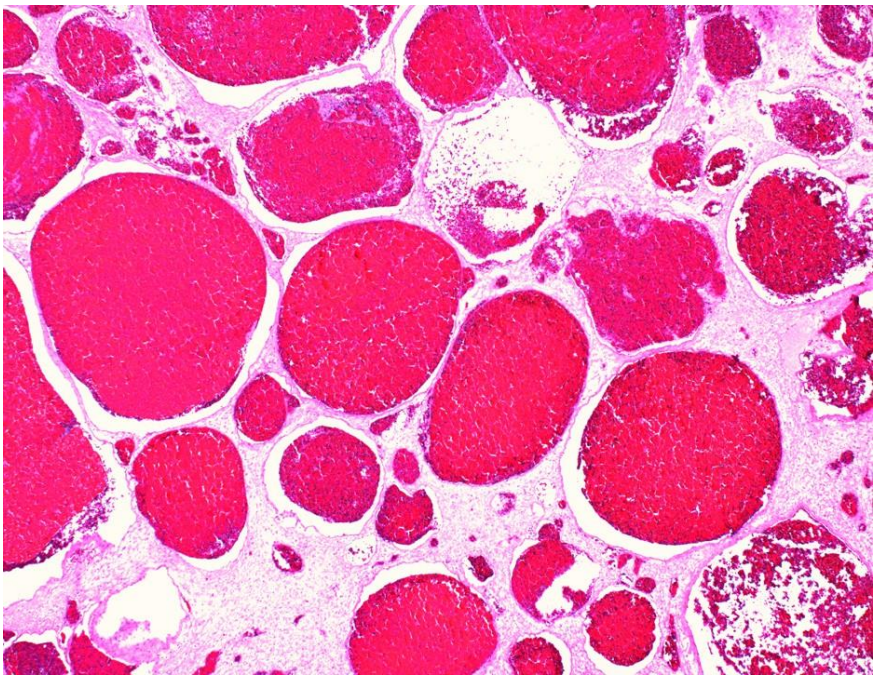

Figure 3. Spinal cord. Higher magnification of Fig. 3. The mas is composed of numerous profiles of well-diferentiated, variable sized, blood filled vascular chanells. HE.

Follow-up questions:

(1) Histologic findings: Spinal cord. Aproximately 70% of the neurparencyma including grey and white matter is effaced by a well demarcated, unencapsulated mass composed of numerous, varieble sized vascular channels. The vascular structures are lined by a single layer of well differentiated endothelial cells, and surrounded by loosely arranged collagenous stroma (Fig. 3). No mitotic figures are observed. The parenchyma surronding the vessels is compressed (Fig. 2).

(2) Morphological diagnosis: Spinal cord (C4-C5 and T7-T8). Congenital vascular hamartoma with compression of the surrounding grey and white matter/neuroparenchyma.

(3) Most likely differential diagnosis: Hemangioma

Comments:

Hamartoma is a benign focal disorder consisting of overgrowth of normal mature tissue in a specific organ. Vascular hamartomas (VA) are considered vascular malformations (8). They can develop in any part of the body, and they are often referred to as "angiomas" by some authors. These lesions are considered a disordered

overgrowth of mature tissue that shows a coordinated growth with the adjacent tissue. Most cases are congenital (7,8), as observed in our case, where the lesion was detected during the first few days of life. However, VA have also been reported in adult animals, possibly due to their slow and progressive growth (7,9). Although VA are not malignant, they may cause symptoms depending on their size, location, and the impact on adjacent structures. In this case, the location in the spinal cord prevented the animal from standing and walking, which prompted euthanasia.

In horses, VA has been described in limbs, ovary, heart, and intestine (2,3,6,7,9). These lesions can cause lameness (2,9), may rupture or become injured, leading to bleeding, and have also been associated with cardiovascular dysfunction (3). A case involving the white matter of the medulla in a foal was described, causing dyspnea, left side facial nerve paralysis and collapse (1).

Grossly, cutaneous hamartomas may appear as swollen areas where blood circulation is compromised. In joints, these lesions could be attached to tendons and muscles by a firm fibrous capsule, enclosing friable vascular tissue (2,9). The size of these tumors can vary significantly, ranging from a few millimeters in diameter (3) to over 20 cm (9). In our case, the filly had hemiparesis of her right limbs because of the compression of the spinal cord by the VA.

Histologically, VA are characterized by abnormal blood vessel proliferation. Some vessels may exhibit thin or multiple layers of endothelial cells surrounded by hemorrhagic granulation tissue. Multiple thrombi and inflammatory cells, predominantly neutrophils, may be noted (2,3,6,7,9). In the case presented here, however, no inflammation or granulation tissue was observed. Immunohistochemistry markers such as vimentin, smooth muscle actin, and factor VIII can be performed (1,3), although the morphology in HE-stained sections is usually straight forward to reach a diagnosis.

Differential diagnoses include other benign vascular proliferations such as hemangioma, hemangioendothelioma, telangiectasis, angiokeratoma and arteriovenous hemangioma (8). VA of the nervous system compress the neuroparenchyma and as a result, swollen axons and dilated myelin sheath may be noted. (4,5,8,10).

The hemangiomas present a significant diagnostic challenge due to the histologic similarities to VA. Both lesions exhibit vascular channels; however, hemangiomas tend to be more hyalinized, tightly packed, sometimes thrombosed, and separated by varying amounts of preexisting neuroparenchyma, hemosiderin and mineralization (4). A key distinguishing feature is the presence of a leptomeningeal plaque in hemangiomas, which extends along the bridging vasculature and invades the adjacent neuroparenchyma. Additionally, meningioangiomatous tissue forms "collars" around parenchymal blood vessels. While both hemangiomas and VAs are vascular malformations, hemangiomas are typically more destructive and neuroinvasive, occasionally exhibiting behavior similar to that of malignant neoplasms (4,5,8,10).

Other vascular malformations, such as telangiectasia, are characterized by abnormally dilated vessels that may be congenital or acquired. Unlike hamartomas,

telangiectasia is not true vascular proliferation but rather a dilatation of preexisting vessels.

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