Brief Communication


Hydrocephalus, Syringomyelia, and Spinal Cord Angiodysgenesis in a Lhasa-apso Dog

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Abnormal cavitations of the central nervous system generally are named by using the idiom “syrinx-” and they occur most frequently in the spinal cord as syringomyelia. The underlying event usually is a malformation of the central nervous system caused by arrested development before gray and white matter have completed differentiation. Acquired syringomyelia as a post-traumatic event also has been described in man. Syringomyelia formation in man also may be caused by hydrodynamic disorders of the cerebrospinal fluid where pulsations of the fluid lead to abnormal distension and disruption of the spinal canal. Circulation disorders within the spinal cord are another important cause of syrinx formation. We describe syringomyelia in a Lhasa-apso dog associated with a developmental disorder of both the central nervous system and the spinal blood vessels. While the former status exists from the fetal period, the latter is a progressive disorder, leading to a clinical appearance of the entire developmental disorder only at advanced age.

The 8.5-year-old dog was presented because of severe ataxia, rolling over, and a disturbed sensorium. The dog became entangled in its leash and had a subluxation between C5 and C6. At necropsy, the cervical, thoracic, and lumbar-sacral regions had large hemorrhages and hematomas. There was a marked hydrocephalus (fig. 1) with reduction of the frontal cortex to a diameter of about 0.5 to 0.8 cm. The corpus callosum was absent. In the basal forebrain there were diverticula bilaterally located within the cortical matter. The sylvian aqueduct was distended markedly from the mesencephalic region. The cerebellum did not cover the roof of the fourth ventricle, but was dislocated anteriorly. There was an extension of the spinal central canal between the cervical segments 1 to 5 and from the cervical region C5 to lumbar segment L2. There was always a complete closure of the cord, although the dorsal neural tissues were thinned to a small rim beyond the meningeal membranes.

Histologically, the ventricular lining of the forebrain had scattered subependymal fibrosis and some hemorrhages. At many locations the border against the ventricular system was a thin rim of fibrotic tissue with only a few, locally concentrated astrocytic reactions. There were multiple clefts and cavitations in the basal hippocampus as well as in the suprachiasmatic and preoptic nuclei (fig. 1). Occasionally these clefts were surrounded by fresh hemorrhagic spots. There was no clear demarcation of the brain parenchyma against the clefts. The cerebellar lingula and the lobulus centralis were absent. The fourth ventricle was covered only by a thin membrane which continued bilaterally into a small pin of protruding brain tissue (fig. 2; “B”). The inner side of this membrane consisted of a continuous layer of ependymal cells—the upper side was part of the pia mater.

At the C1 nd C2, the spinal cord had a syrinx within the dorsal cord, separated by thin layers of tissue from both the arachnoidal space and the central canal. The gray matter of the cord was well-developed, as were the dorsal and ventral nerve trunks, which were surrounded by well-formed pachymeningeal tissue. From C3, the cavitation of the cords extended more laterally, thus severely affecting the formation of the central gray matter (fig. 3). At the same time the syrinx was in direct continuity with the central canal. The most conspicuous finding, especially between C2 and C6, was an abundance of tortuous, vascular tissue within the cavitations (figs. 3, 4). This tissue was comprised mainly of thick-walled veins with many capillaries and undefined vessels, conglomerated within dilated fibrotic tissue. These vessels often bordered on the clefts, which extended to the arachnoid space at the level of C3. The parenchyma immediately beneath the cavities was edematous, with hemorrhages and some necrosis. There was a complete lack of reactive inflammation or any demarcation.

This is the first report of simultaneous occurrence of both a central nervous system malformation and a vascular malformation in an animal. The malformation of the cerebellar vermis and hypoplasia of the roof of the fourth ventricle would be characteristic for Dandy-Walker malformation, but since no abnormalities of the tentorium were found, this entity only can be suggested. The Dandy-Walker malformation often is linked to nonopening of the cerebellar foramina and to a subsequent development of hydrocephalus. The finding of a vascular malformation is rare. Our diagnosis was based on the appearance of significant masses of tortuous vascular tissue within the syrinx cavities. A similar finding, with the exception of a few sporadically appearing vessels never was described in hydrocephalus or in syrinx formation of the spinal cord. Thus, we do not regard these vascular peculiarities only as rudiments of a rarefying process.

The leading pathological condition is, undoubtedly, the hydrodynamic disorder of the central nervous system fluid, associated with a significant hydrocephalus internus. Long persisting hydrocephalus is known to lead, by the force of the increasing hydrodynamic pressure to a massive dilatation of the spinal cord central canal (hydrromyelia) and also to syrinx formation. Due to the lack of reactive processes within the spinal cord and in view of the occurrence of fresh hemorrhages, the acute traumatic (and strangulative) injury

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Fig. 1: Coronal section of forebrain; marked hydrocephalus and absence of corpus callosum. Two large cavitations (C) situated bilaterally in area of globus pallidus. Bar = 1 cm.

Fig. 2: Fourth ventricle covered only by thin membrane composed inside by an ependymal cell layer (EP) and outside by the pia mater (PM). Bilateral thin bridges (B) of neural tissue. Mallory. Bar = 0.1 cm.

Fig. 3: Spinal cord section at C5. Extension of syrinx into lateral cord; communication with central canal. Numerous islets of vascular tissue (V) within cavitation. Diminution of dorsal horns. Wilder. Bar = 0.5 cm.

Fig. 4: Spinal cord section at C4. Boundary (arrow) between syrinx and neural tissue. Central cavity filled with connective tissue containing numerous tortuous vessels (V). Masson’s trichrome. Bar = 50 μm.

of the dog was a most serious aggravating factor in the disease. An alternate pathogenesis of the cord syringes emerges, in our opinion, from the vascular disease. The underlying etiology seems to be a congenital status of “vessel wall dysoria,” which presumably affects the veins more selectively than the arteries. This is interpreted as the outcome of an insufficient involution of the less-differentiated vessels of the cerebro-medullovasculos area in fetal development. This maldevelopment initially is made evident by the abnormal varicosity of the relevant veins. The life-long strain on these vessels leads to insudation, fibrinous emboli, probably also inflammation, and finally —concomitant with proliferative processes of the intima—to a distinct fibrosis of the vessel walls.

In accordance with similar observations in man, the vascular malformation induces a necrotizing myelopathy of angiodyssgenetic origin. In human neurology this entity is known as the Foix-Alajouanine syndrome. Initially there would have been edema, then necrosis of the pre-existing spinal cord syrinx walls and, as a result, an enormous extension of the cavities in later life. This progressive disorder even involved the central canal, which gave rise to hydrosyringomyelia.

References
10 GARDNER, W.J.: Hydrodynamic mechanism of syringo-
Ganglioneuroma of Vater's Papilla and Extrahepatic Cholestasis in a Dog

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Ganglioneuroma\(^4\) or ganglioneurofibroma\(^1\) is a benign neoplasm composed of nerve fibers and mature ganglion cells. The principal tissue consists of interlacing fascicles of axons and their accompanying Schwann cells\(^6\) as well as fibroblasts.\(^1\) The ganglion cells are distributed in an irregular fashion or are arranged in compact clusters. Satellite cells usually are absent, but occasionally are arranged normally.\(^1,4\) This neoplasm is uncommon in animals;\(^3\) only three reports of ganglioneuromas in the dog were found.\(^2,3,5\)

A five-year-old castrated male bobtail dog was presented with episodes of apathy, decreased endurance, vomiting, and jaundice for one year. Physical examination revealed no abnormalities except jaundice.

Hematocrit, leukocyte count and differentiation, total protein and electrophoresis were within normal range. Blood coagulation, as indicated by prothrombin time and fibrinogen level, also was normal. Alkaline phosphatase (4100 IU/l; normal range 46-104 IU/l), serum glutamic pyruvic transaminase (1760 IU/l; normal range 12-36 IU/l), serum aspartate aminotransferase (135 IU/l; normal range 10-26 IU/l) and gamma glutamyl transferase (60 IU/l; normal range 3-14 IU/l) were elevated markedly. Total bilirubin was increased to 126 \(\mu\)mol/l (normal range <3.4 \(\mu\)mol/l) and 88% was conjugated.

After physical examination and the laboratory results, a liver biopsy was done percutaneously using the Menghini aspiration technique.\(^6\) Histological examination of the biopsy revealed portal fibrosis, extending in the surrounding liver parenchyma, with ductular proliferation, neutrophils, ceroid-laden macrophages and few lymphocytes and plasma cells. The lobular parenchyma had only minimal changes, i.e., some hemosiderin in Kupffer cells, and few neutrophils in the sinusoids. Although no cholestasis was seen histologically, the portal changes warranted the diagnosis of extrahepatic cholestasis.

Investigative laparotomy was done under general inhalation anesthesia (Halothane\(^6\), [Profield Surgicals A.G., Zug, Switzerland], nitrous oxide, oxygen). Lesions were confined