

Case Report

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Syringomyelia in a Newborn Male Simmental Calf

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Key words: Bovine; Malformation; Spinal cord.

Malformations of the spine and spinal cord are common in domestic mammals.¹ Among these, syringomyelia is defined as a cystic, fluid filled tubular cavity (syrinx) within the spinal cord extending over several segments. Syringomyelia occurs as a primary congenital malformation and is most commonly combined with neural tube defects. However, it can also be acquired after alterations in cerebrospinal fluid pressure (CSFP), following rupture of a hydromyelia, trauma, neoplasia, or inflammation. In chronic cases, the cavities can be lined with reactive astrogliosis. Syringomyelia has been most intensively studied in Cavalier King Charles Spaniels but occurs in other mammals.^{2–7} It was previously considered a rare condition in small animal medicine, but because of higher prevalences in certain dog breeds and the increased availability of magnetic resonance imaging (MRI), it has become a relatively common neurological diagnosis.⁸

Syringomyelia develops outside the central spinal canal and the cyst is lined by glia cells, whereas hydromyelia affects the central canal and is lined by ependymal cells.¹ The distinction between syringomyelia and hydromyelia is arbitrary; hydromyelia might extend into the spinal cord and form syringohydromyelia partially lined by ependyma⁹ and cavities might communicate with the central canal.¹⁰ At present, the term syringomyelia is generally accepted for all spinal cord cavitations containing cerebrospinal or similar fluids.⁸ A maldeveloped cerebellum with elongated cerebellar tonsils through the foramen magnum (Arnold-Chiari malformation) is seen in human infants and calves of many breeds.¹¹ Congenital malformations affecting the cerebellum, such as Chiari-like malformation with displacement of the cerebellum through the foramen magnum,

Abbreviations:

BHV-1	bovine herpes virus 1
BTV	bluetongue virus
BVDV	bovine viral diarrhea virus
CSFP	cerebrospinal fluid pressure
HE	hematoxylin and eosin
LFB	luxol fast blue
MRI	magnetic resonance imaging
SBV	Schmallenberg virus

or Dandy-Walker syndrome with cerebellar hypoplasia, cysts in the fourth ventricle and hydrocephalus often occur in combination with syringohydromyelia. Other congenital disorders affecting the spinal cord are described in the literature,¹² from spinal dysraphism,¹³ spina bifida and spinal dysraphism,¹⁴ segmental aplasia of the cord¹⁵ and segmental hypoplasia.¹⁶

Case History

An 8 day-old male Simmental calf was referred to the university veterinary hospital in Vienna by the Austrian Simmental Association. The calf originated from a dairy farm and was the fifth calf of a dairy type Simmental cow, which had been artificially inseminated. Both the bull and his semen tested negative for bovine viral diarrhea virus (BVDV) and bovine herpes virus 1 (BHV-1), but the bull tested positive for antibodies to Schmallenberg virus (SBV). All semen batches tested negative for SBV. The dairy herd in question has been proven free from BVDV, BHV-1, SBV and bluetongue virus (BTV) during routine herd surveillance (using PCR) carried out by the state veterinary authorities. The calf was suspected to suffer from a hereditary disorder as it appeared clinically healthy, except for the inability to move and position its hind limbs normally. The owner reported that the calf was in good condition at parturition; feed intake and movement of the front limbs were normal, however it was said to “bunny hop” on its hind limbs.

Clinical Findings

A full physical examination was performed on the day of arrival at the clinic. The Simmental calf weighed 47 kg on admission. Its mental status was normal (bright, alert, and responsive) and it reacted with appropriate movements of its head, neck, eyes, and ears to events in its vicinity. The body condition was somewhat less than expected, i.e. the animal was thin. The examination of all other organ systems revealed results within normal limits.

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Orthopedic and Neurological Examination

The calf was able to stand without assistance, and assessment of gait demonstrated that the front limbs were used normally but the hind limbs only moved in an asymmetric “bunny hopping” style. The calf had difficulty standing still and, in an attempt to balance its weight, it would spin around in a clockwise direction (Fig 1). However, it appeared to have normal mental status.

Palpation of the right hind limb did not reveal any abnormal findings. The calf could not flex the joints of the right hind limb, however, the right tarsal joint could be flexed passively. During locomotion, the right limb was abducted permanently in a craniolateral direction. The right thigh muscles were hypertonic. While the tail's muscle tone appeared normal, its movements in the proximal region, in particular, were decreased. Cranial nerves showed normal function. Skin sensitivity and pain responses in the interdigital space of all four feet were within normal limits. Skin sensitivity of the right thigh was found to be normal caudally, but severely reduced in the quadriceps region. Palpation of the vertebral column did not reveal any abnormalities, but the panniculus reflex was reduced caudally from L4.

Proprioception could not be tested on the right hind limb, because of a highly increased extensor tone, but conscious proprioception responses were close to normal for the remaining limbs. Following the clinical and neurological examinations, a diagnosis of a spinal cord lesion at the level of the caudal lumbar spine was made.

Diagnostic Imaging

Lateral radiographic views were taken of the spine, showing no abnormalities. Economic considerations

limited the range of diagnostic imaging techniques available and MRI was not feasible in this case. Because of poor prognosis, the calf was euthanized and a thorough necropsy examination was performed.

Pathological Findings

On necropsy, no malformations, fractures, or luxations of the vertebral column were detected. Tissues, with particular focus on the CNS, were removed and fixed in 10% neutral buffered formalin, embedded in paraffin-wax, sectioned, and stained with hematoxylin and eosin (HE) and luxol fast blue (LFB) for histology. The formalin-fixed spinal cord was transversally sectioned at each segment and examined for macroscopic lesions. The cervical, thoracic, and lumbar segments up to L4 were without abnormalities. Between L5 and S2 there was a clearly visible cavitation (5 mm deep and 2 mm wide) located in the midline of the dorsal area of the spinal cord (Fig 2). Microscopic examination revealed multiple malformations of the lumbar spinal cord over several segments. A further cystic cavity was detected in the dorsal funiculi ranging from L5 (Fig 3a) to S2, accompanied by smaller cavitations in the adjacent white matter. Neither the central cavity nor the smaller cavitations showed signs of inflammation (e.g. gliosis or spheroids). The central cavity was confined by a thin line of connective tissue. The smaller cavitations could be observed bilaterally at L5, L6, S1-S2 and unilaterally between L6-S1 and at S2. There was no direct communication detectable between the central canal and the syringomyelia. The dorsal contour of the spinal cord appeared irregular because of the cavitations. Additionally, hypomyelinated areas were detected in the dorsal root entry zone.



Fig. 1. The calf losing balance and trying not to fall, not able to bend the right hind limb.



Fig. 2. Macroscopic view of the spinal cord at L5 with the clearly visible cystic formation (syringomyelia) in the dorsal funiculi; caudo-cranial view.

The shape of the central canal changed, becoming only rudimentary and virtually disappearing between L6 and S1, (Fig 3b), before returning to normal between S1-S2. When present, the inner lining of the central canal appeared normal. Furthermore the ventral median fissure disappeared completely between L6 and S1 (Fig 3b) and reappeared at S1-S2. The gray substance with its dorsal and ventral horns was clearly distinguishable by means of LFB at L5 (Fig 3a). However, between L6-S1 (Fig 3b) only the dorsal horn was clearly visible. In this segment, dysplasia of the gray matter was detectable in the form of neurons lying within the normal white substance. Despite the indistinguishable outline of the ventral horn, the ventral motor neurons appeared to be in situ. The remainder of the spinal cord, brain, sciatic nerves and muscles were unremarkable. Additional necropsy findings included milk in the omasum and abomasum, a mild focal interstitial nephritis and a severe alveolar lung edema.

Discussion

This report presents a newborn born Simmental calf with the inability to move its hind limbs in a normal manner. The cause of this locomotory abnormality was found to be linked to a syringomyelia and presence of multiple smaller cavitations located close to the dorsal horns of the spinal cord. The clinical signs of syringohydromyelia reflected the location of the lesion in the caudal spinal cord and corresponded with previously reported clinical signs.¹⁷ From clinical and neurological examinations, as well as basic diagnostic imaging techniques, it is difficult to differentiate between hydromyelia and syringomyelia. Advanced diagnostic techniques such as MRI are useful, with necropsy and histology essential for a definitive diagnosis.

Cavitations and hypomyelination close to the dorsal horn ranging from L5 to S2 in this calf might have negatively affected the dorsal roots of the lumbar nerves resulting in a reduced level of skin sensitivity on the dorsolateral plane of the hind limb and an absence of the panniculus reflex at that level. The markedly

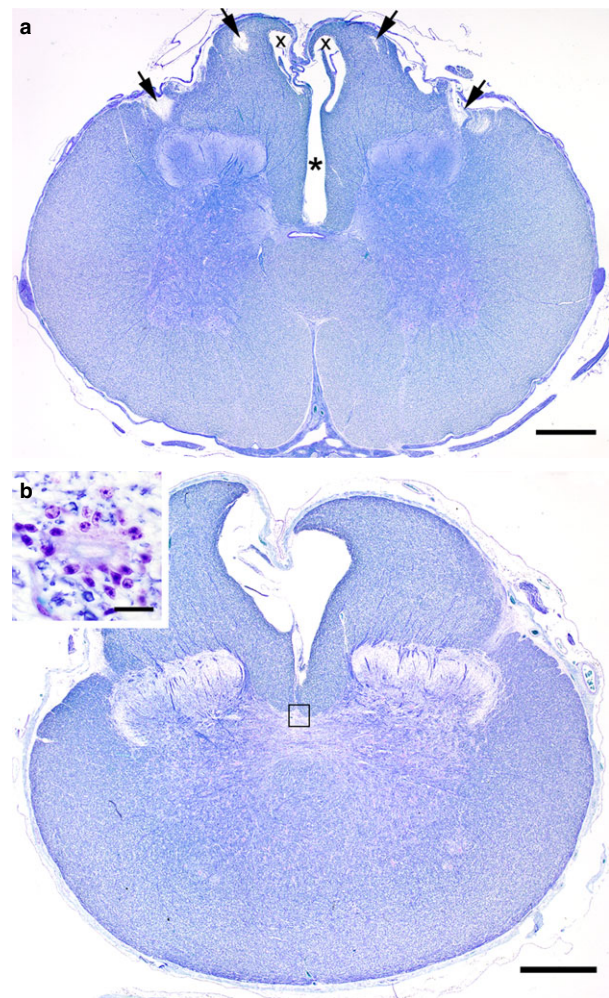


Fig. 3. (a) Spinal cord at L5 with the centrally located syringomyelia (asterisk) and the bilateral cavitations (crosses and arrows). No communication detectable between the central canal and the syringomyelia. The dorsal and ventral horn of the gray matter and the ventral fissure are clearly distinguishable (LFB, 12.5 \times ; bar = 2 mm; caudo-cranial view). (b) Spinal cord at L6-S1 with the centrally located syringomyelia. The dorsal horn of the gray matter is clearly visible, whereas the outline of the ventral horn is not distinct. The ventral fissure is missing. The inset shows the rudimentary central canal in detail (LFB, 12.5 \times , bar = 2 mm; inset: 600 \times ; caudo-cranial view).

reduced ventral median fissure and central canal at the level of L6 to S1, as well as the presence of the syringomyelia from C5 to S2 could suggest the involvement of intrasegmental tracts from one side to the other. Flexor-extensor inhibition, which is essential for normal locomotion,¹⁷ could be affected by such tracts, resulting in the asymmetric bunny hopping gait seen here. In this case, however, the increased extensor tone was more severe in the right hind limb.

The likelihood of a traumatic event, which has caused spinal injuries, was ruled out in this case, as no bone fractures or other abnormalities such as epidural or intraspinal hemorrhage were observed in the radiographs nor at necropsy. Furthermore, the histological

examination did not show edema, neuronal cell shrinkage, axon swelling, necrosis, gliosis or capillary proliferation, which are commonly described as a consequence of trauma.¹⁸ In hindsight, the authors would recommend ultrasound investigations for the practitioner in general practice as this method of diagnostic examination is readily available and is highly likely to have provided further evidence of syringomyelia in a living animal.^{19,20} Furthermore, myelography and MRI^{6,21} would have been appropriate as further diagnostic imaging tools in valuable living animals, but were not carried out in this case because of economic constraints.

These findings, combined with the fact that the physical signs were observed as soon as the animal was able to stand and were noted on its first attempts to walk, suggest that the calf suffered from a congenital disorder of syringomyelia.

Virus infections are known to play a substantial role in neural tube abnormalities.¹¹ In cattle, cerebellar and vertebral malformations induced by BVDV *in utero* are well-documented.²² However, no evidence of SBV, BVDV, and BTV were found in the calf, dam, or herd, and no vaccinations against these viruses had been administered on the farm in question. Syringomyelia as described in the literature is often combined with additional disorders such as Dandy-Walker syndrome, scoliosis, cleft palate,²² Arnold-Chiari like malformation,⁷ dicephalus or spinal dysraphism.²³ It is an abnormal spinal cord development,¹ that includes – among others - hydromyelia, aplasia, or hypoplasia of the spinal cord, diplomyelia, aberrations of the central canal and of dorsal or ventral septa. These changes can be associated with vertebral malformations leading to spina bifida or myelomeningocele. Without skeletal abnormalities it can be difficult to clinically distinguish these abnormalities from other causes of spinal cord diseases.²⁴ Whereas, in this case, there were no vertebral malformations, the syringomyelia was combined with abnormalities of both the central canal and the ventral fissure. Although the central canal was significantly narrowed over several segments, there were no signs of a subsequent dilatation or hydromyelia elsewhere as might be expected because of a raised CSFP.

Genealogical information obtained from the cattle breeders' association with respect to the calf's heritage failed to indicate any recent inbreeding in the pedigree; however the common degree of inbreeding in domestic cattle breeds²⁵ has generally increased the likelihood of phenotypic expression of recessive traits and therefore the occurrence of genetic defects. Clinical signs seen in this calf were similar to those exhibited by Tyrolean gray calves suffering from degenerative axonopathy, which is reported to appear at the age of 4 to 6 weeks. Furthermore, there were no signs of axonal degeneration, of swelling and chromatolysis of neurons in the brainstem, the spinal cord or sciatic nerves.²⁶ A spontaneous mutation event, which affected cerebral development, could also have caused this phenotypical presentation. As a genetic cause of the syringomyelia cannot be ruled out at this time, it is very important that calves with similar clinical

problems are reported to the relevant cattle breeders' associations, as well as diagnostic and research centers, such as university clinics, to facilitate future analyses of bovine pedigrees.

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Supporting Information

Additional Supporting Information may be found online in Supporting Information:

Video S1. Video of the affected calf showing the bunny hopping gait.

Video S2. The calf had difficulty standing still and, in an attempt to balance its weight, it would spin around in a clockwise direction.