Case Report

Imaging diagnosis and clinical presentation of a Chiari malformation in a Thoroughbred foal

A. Lempe*, M. Heine†, B. Bosch‡, K. Mueller§ and W. Brehm

Large Animal Clinic for Surgery; †Large Animal Clinic for Theriogenology and Ambulatory Services; ‡Department of Small Animal Medicine; and §Institute of Pathology, University of Leipzig, Leipzig, Germany.

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Summary

A newborn Thoroughbred foal was presented to the clinic with ambiguous neurological deficits, spinal anomalies and a soft tissue swelling dorsal to the lumbar vertebral column. The foal was alert but unable to rise and stand. With radiography, ultrasonography, computed tomography (CT) and magnetic resonance imaging (MRI) a lumbar dysraphic anomaly, cerebellar herniation and coincidental skeletal abnormalities were documented. Finally, a meningomyelocele was defined and, in context with the cerebellar herniation through the foramen magnum, the foal was diagnosed to have a Chiari malformation. The MRI examination corresponded best with the post mortem findings. Although 3-dimensional imaging methods have been considered superior regarding full and detailed assessment of the congenital malformation, radiography and ultrasonography also provide essential information to diagnose dysraphic lesions at reduced costs and efforts. A Chiari malformation should be considered as a differential diagnosis in foals with neurological deficits.

Introduction

Cleft anomalies and dysraphism are rare congenital conditions in the equine species caused by an embryonic failure of vertebral fusion (Copp and Greene 2010). Spina bifida is the most commonly used term describing such malformations. A more accurate definition refers to the extent of spinal cord protrusion through the cleft. In horses meningomyelocele and meningeal cysts of the cervical and thoracic segment combined with dorsal or less obvious ventral protrusion have already been described (Leathers et al. 1979; Harmelin et al. 1993; Dolge 1996; Rivas et al. 1996; Jacobsen et al. 2007; Rendle et al. 2008; Gerhauser et al. 2010). Due to the variation of cleft anomalies and other spinal deformities (Lerner and Riley 1978; Kirberger and Gottschalk 1989; Ryan et al. 1992; Johnson et al. 1997; Wong et al. 2005) clinical signs are inconsistent including general paresis, ataxia, ‘dog-sitting’ position and delayed or nonexistent response to reflex tests. The majority of affected foals were diagnosed by necropsy at only a few weeks or even months when neurological signs had increased.

A number of species, including calves (McFarland 1959; Grays 1973; Ohfuji 1999) sheep (Potter et al. 2010), dogs (Chesney 1973; Wilson et al. 1979; Ruberte et al. 1995; Shamir et al. 2001) and cats (Plummer et al. 1993) can demonstrably develop spina bifida. However, the uncommon condition of a meningomyelocele in association with cerebellar herniation through the foramen magnum has only been described in calves (Frauchiger and Fankhauser 1952; Grays 1973; Cho and Leipold 1977; Madarame et al. 1993; LeClerc et al. 1997; Testoni et al. 2010; Zani et al. 2010) and one dog (Schmidt et al. 2008). This complex congenital disorder has similarities with human Chiari malformation type II and, to date, no descriptions exist referring to this condition in the equine species.

The following report describes the first case of a foal with Chiari malformation using radiography, ultrasonography, MRI and CT in order to identify and to characterise the involved structures noninvasively. The imaging diagnosis which can be derived from such techniques was assessed and compared with the findings of the post mortem examination.

Case details

History

A few hours old female Thoroughbred foal was referred to the clinic for evaluation of a distinct swelling dorsal over the lumbar column and to clarify its inability to rise or stand. The

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owner reported no noticeable problems during the mare’s pregnancy and, after a gestation period of 319 days, parturition had been uneventful. The foal presented was the third foal of the mare without repeat breeding; earlier foals were born healthy on time. A soft swelling at the foal’s dorsal lumbar region had been present since birth. At no point in time had the foal been able to rise or to stand but showed temporary movement of the front limbs.

Clinical examination

On admission, the foal was in lateral recumbency but alert and showed paroxysmal flailing movement of both thoracic limbs. The head could only be lifted for short periods. Turning to a sternal position to stand up was impossible for the foal due to the inability of any spontaneous coordinated movement of the hind limbs. Absence of the withdrawal reflex with nociceptive deficiency of the pelvic limbs was noticed. Generally, the foal had a good body condition, and temperature, pulse and respiratory rates were within the normal limits. The owner could not provide information on whether the foal had defaecated or urinated normally.

A soft, fluctuant swelling approximately 4 x 5 cm rising approximately 3 cm above the skin was present at the dorsal lumbar region. The skin over the swelling was covered with focal ulcerations at the dome (Fig S1). Palpation seemed painful to the foal. The swelling had a fluid-filled consistency but the bulging dimensions impeded palpation of the vertebrae beneath. Additionally, a severe lateral displacement along the thoracic vertebral column was noted suggesting a scoliosis malformation.

Diagnostic imaging

Radiography

Radiographs (FCR 5000) were obtained in laterolateral and dorsoventral projections. The images showed a severe C-shaped scoliosis (concave side left) of the thoracolumbar segment and distinct homogenous soft tissue opacity dorsal to the 3rd to 6th lumbar vertebrae. Evaluation of the integrity of the particular vertebrae was hindered by the 2-dimensional superimposition and oblique projection. However, the spinous processes appeared abnormally thickened and misshapen in this region (Fig 1). Due to the severe scoliosis and suspected dysraphic lesion the owner requested euthanasia. Imaging examination was continued immediately after death.

Ultrasonography

Subsequently, the swelling was clipped and prepared for further ultrasonographical examination in transverse and longitudinal orientation using a 10 MHz linear transducer. Transverse ultrasonograms demonstrated a symmetrical split formation of the underlying spinous processes and the dorsal arch of L3 to L6 allowing ultrasonographically access to the soft tissue content. The depth of the cavity measured 5 cm from the skin to the layer of well defined hyperechoic, homogenous tissue ventrally. In transverse orientation the width measured 2 cm ventrally, diverging to 5 cm dorsally. The bone cavity and swelling contained an irregularly arranged tissue accumulation of moderate echogenicity and varying amounts of fluid-like anechoic areas, bordered by a narrow hyperechoic tissue layer (Fig 2). Considering the defect as a cleft formation of the vertebral column, the hyperechoic tissue layer was characterised as meningeal protrusion surrounding ascending parts of the spinal cord, nerve filaments and cerebrospinal fluid.

Computed tomography (CT)

Computed tomography (Brilliance CT 6, Philips Whole body-CT Mx8000 IDT) was performed in noncontrast images and 3D-reconstruction to visualise the thoracic and lumbar vertebral column with the foal in ventral recumbency. On CT images the lateral deviation of the thoracic vertebral column was clearly visible, involving T14

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and extending to L1. Additionally, there was a clockwise rotation of the vertebral bodies of T15 to L1 from 10–50° around the central spinal axis. The dorsal arches, including the spinous processes from L3–L6 were not fused; they appeared almost symmetrically divided, leaving a cleft of approximately 3 cm maximum. The dorsal arches of the sacral vertebrae were complete but their spinous processes were also nonfused. Additional findings were a fracture of the 18th rib on the left side and a fusion of 3 rib structures on the right side articulating with T17. The right 18th rib was not visible (Fig 3).

Magnetic resonance imaging (MRI)

Magnetic resonance imaging was performed to characterise the soft tissue using a 0.5 Tesla scanner (Gyroscan T5-NT)³. The foal was placed in dorsal recumbency on the table and scanned in T1- and T2-weighted spin and gradient sequences. Images were obtained in sagittal, transverse and dorsal planes. Tissue characterisation in T1-weighted sequences was made in relation to the longissimus dorsi muscle. Tissue of isointense signal intensity extended through the dorsal opening of affected vertebrae, representing grey and white matter. The isointense, inhomogenous tissue originating from the epidural space was enlarged in its dorsoventral dimensions. The cavity contained hypointense cerebrospinal fluid and was bordered by isointense meninges and skin which could not be visualised separately. In T2-weighted sequences the cerebrospinal fluid and the tissue of the spatium epidurale showed increased signal intensities compared to T1-weighted sequences. The demarcation of the epidural space beneath the nonfused vertebrae was clear as there was signal difference to the hypointense, ventral part of the meninges (Fig 4a). Due to the cross-sectional sections along the arching protrusion, assessment of the origin and definition of the spinal cord in transverse planes was limited. Differentiation of grey and white matter within the spinal cord was not possible in the affected region (Fig 4b). Magnetic resonance imaging examination of the cranium revealed a herniation of the cerebellum through the foramen magnum into the spinal canal. Diagnosis of a malformation resembling a Chiari type II malformation was made, founded on the previously described protrusion of the spinal cord and meninges at the lumbar region. In addition, a dilatation of the cranial spinal canal was apparent on sagittal images indicating a hydromyelia (Fig 4c).

Post mortem examination

On post mortem examination a severe scoliosis about 20 cm in length was diagnosed involving both the thoracic and lumbar segment (T13-L3) of the vertebral column.
Additionally, the lumbar vertebral column had a complete dysraphic malformation dorsally from the 4th to 6th vertebrae (spina bifida aperta) with bullous protrusion of meninges and parts of the spinal cord (meningocele). The cavity was filled with liquor spinalis (Fig 4a). After longitudinal sectioning of the cranium herniation of the cerebellar vermis through the foramen magnum was obvious and therefore the imaging diagnosis of a Chiari malformation type II could be confirmed (Fig 4c).

**Discussion**

Spina bifida is classified as spina bifida aperta (also cystica) defined by a protrusion of spinal tissue through the cleft vertebræ or as spina bifida occulta where the spinal cord tissue remains in place. Depending on the protruding tissue, a spina bifida aperta can be divided into 3 subtypes. In case of a meningocele only the meninges bulge out to form an apparent cyst (Leathers et al. 1979; Doige 1996) whereas a meningomyelocele is characterised by the passage of meninges and parts of the spinal tissue including nerve cords through the bone cleft (Harmelin et al. 1993; Rivas et al. 1996; Jacobsen et al. 2007). In human medicine this type is the most common among all spinal defects (DeMarco et al. 2011). Myeloschisis represents the most severe subtype by exposing spinal tissue without a covering of skin, and to date this condition has not been reported in horses.

A previous report of a miniature colt with a cervical meningomyelocele and hydrocephalus has already suggested a Chiari malformation, but there was no proven evidence because of the lack of diagnostic imaging of the head and necropsy (Rivas et al. 1996). In our case, MR examination allowed the determining depiction of cerebellar herniation to diagnose a Chiari-like
malformation noninvasively. Even though the final diagnosis was based on MR findings of a foal recently subjected to euthanasia, such an examination is also practical in foals under general anaesthesia. The associated meningomyelocele was protruding dorsal from the lumbar vertebrae, easily assessable with other imaging modalities. Radiographic indication of spina bifida in the presented case was the appearance of nonfused spinous processes in conjunction with a radiopaque mass dorsal to the suspected cleft. If possible, a ventrodorsal or dorsoventral orientation is preferable in young foals as in laterolateral views both parts of the spinous process superimpose. However, additional complex malformation of the spinal column can also lead to superimposition of the dysraphic segment. Contrast myelography may have resulted in increased informative value but due to the oncoming diagnostic procedures it was not performed. Regarding the classification of the dysraphic lesion, ultrasonography was useful to evaluate soft tissue involvement and furthermore to exclude other causes of swellings along the column such as haematoma, seroma, neoplasia or vertebral fractures. In comparison to the post mortem findings this method is considered to be more sensitive than radiography. One report documented the cerebellar displacement in a calf ultrasonographically at the craniocervical junction but due to the different equine anatomy the approach through the foramen magnum is impractical in horses (Testoni et al. 2010). Three-dimensional imaging techniques are advantageous for visualising complex vertebral malformations. Both, CT and MRI provided specific information and were equally important to describe the multiple malformations of this case in detail. Computed tomography revealed the degree of vertebral anomaly, the extension of the scoliosis in connection with vertebral rotation and the abnormal fusion of 3 ribs, findings which were incompletely documented during radiography. Most likely the fracture of the left 18th rib originates from parturition or a post natal event. The conclusive diagnosis was based on MR-images. This imaging technique depicted the lumbar dysraphic lesion in close agreement with the pathological findings by showing the exact protrusion of spinal cord tissue and meninges, ascending nerve filaments, bulging tissue of the epidural space and accumulation of cerebrospinal fluid. T2-weighted sequences provided best detail. The combination of a spina bifida with other skeletal abnormalities is a previously described phenomenon (Cho and Leipold 1977; Madarame et al. 1993; Ruberte et al. 1995; Doige 1996; DenoiX 2005; Jacobsen et al. 2007; Gerhauser et al. 2010). Thus it is advisable to precisely evaluate a neonate or yearling with skeletal deformity and/or neurological signs as there is a wide range of differential neurological diseases in young horses (MacKay 2005; Johnson 2010); eventually the prognosis deteriorates when spina bifida has been diagnosed. In dogs, attempts to correct spina bifida surgically are described (Fingeroth et al. 1989; Shamir et al. 2001) but in horses no such interventions have been documented as being successful (Rivas et al. 1996). For the equine species an appropriate surgical technique to treat cerebellar herniation is currently not available. The MR examination is regarded as essential to distinguish foals with a fatal Chiari-like malformation from those only with vertebral abnormalities and to diagnose less obvious dysraphism such as spina bifida occulta, a meningomyelocele, ventral or cervical meningomyelocele intra vitam.

The occurrence of spina bifida in man is closely associated with folic acid deficiency but genetic distribution, maternal age and diet also play a role (Copp and Greene 2010; DeMarco et al. 2011). The influence of retinoic acid excess and a zinc deficiency may also lead to an increased prevalence of congenital dysraphism (Yasuda et al. 1986; Dubrulle and Pourquie 2004; Rendle et al. 2008). Oral supplementation of the B-vitamin folic acid reduces the incidence of spina bifida in man but a primary folate deficiency in horses is virtually impossible considering their natural diet. However, it could be demonstrated that secondary folate deficiency appeared in horses after administrating sulphadiazine and pyrimethamine to treat equine protozoal myeloencephalitis (Toribio et al. 1998; Piercy et al. 2002). Unfortunately we could not receive any information regarding possible medical treatment during gestation of the mare in our case. In general, the mare and her foal were in good body condition; none of the earlier foals of the mare had shown signs of a developmental disease. A breed disposition can not be derived from literature.

In conclusion, a Chiari malformation is a congenital disease also occurring in the equine species, the prognosis of which deteriorates when diagnosed. MRI is an important diagnostic modality to distinguish foals with this fatal disease from those with neurological disorders for other reasons. Although 3-dimensional imaging methods were superior to assess the dysraphic anomaly in detail, conventional radiography and ultrasonography were inexpensive alternatives that have proven to be sufficiently conclusive in practice.

Authors’ declaration of interests
No conflicts of interest have been declared.

Manufacturers’ addresses
1Fuji Photo Film GmbH, Düsseldorf, Germany.
2GE Healthcare Technologies, Solingen, Germany.
3Philips Healthcare, Hamburg, Germany.

References


Supporting information

Additional supporting information may be found in the online version of this article. Fig S1: Photograph of a Thoroughbred foal a few hours old showing a distinct swelling at the dorsal lumbar region. There is also a scoliotic malformation of the thoracolumbar vertebral column visible.

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