Case Report
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Caudal Occipital Malformation Syndrome in a 6-Year-Old Female Huacaya Alpaca

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Key words: Chiari syndrome; Conscious proprioceptive deficits; Foramen magnum; Suboccipital craniectomy; Syringohydromyelia.

A 58-kg, 6-year-old, female Huacaya alpaca presented to the Washington State University Veterinary Teaching Hospital with a history of gradual developing lethargy, reluctance to rise, anorexia, blindness, and respiratory distress. A presumptive diagnosis of dorsal displacement of the soft palate was made and a tracheostomy tube was surgically placed by the referring veterinarian. Supportive treatment included crystalloid fluids (lactated Ringer's solution) administered IV and antimicrobial (certiofur sodium, unknown dose) therapy. The tracheotomy tube was removed and the respiratory distress appeared to have resolved. For the 2 months prior, the alpaca had been housed at an out-of-state breeding facility. Marginal feed quality during this period was believed to be related to the alpaca's poor body condition score (1/5). The alpaca's vaccination (*Clostridium perfringens* C/D, *Cl. tetani* toxoids) and deworming (Ivermectin) history were deemed appropriate and current. The owner thought that the alpaca might be pregnant.

Upon examination, the alpaca was dull, lethargic (reluctant to rise when stimulated), and had not eaten for at least 1 day. The alpaca was thin (body condition score 1/5) and adequately hydrated. Vital signs (heart rate, respiratory rate, and temperature), capillary refill time, and mucus membrane color were normal. The alpaca urinated and defecated normally and had normal compartment 1 (C1) contractions upon auscultation. The tracheostomy site was patent, healing, and no abnormal respiratory sounds were asculted. Neurologic examination revealed an absent menace response in the left eye, with normal palpebral reflexes and pupilary light reflexes (direct and consensual). No other abnormal cranial nerve signs were detected. The alpaca held her head in an abnormally low position and moved her head slowly in response to stimuli. There were no signs of pain upon neck manipulation. Although it was difficult to maintain the alpaca in a standing position, conscious proprioceptive (CP) deficits were observed in all 4 legs. Flexor reflexes (thoracic and pelvic) and patellar reflexes were normal.

Abbreviations:

AST	aspartate aminotransferase
BUN	blood urea nitrogen
C1	compartment 1
C1-C6	cervical vertebrae 1–6
CBC	complete blood count
cELISA	competitive enzyme linked immunosorbent assay
COMS	caudal occipital malformation syndrome
CP	conscious proprioceptive
CSF	cerebral spinal fluid
EHV-1	equine herpes virus-1
FMD	foramen magnum decompression
KCl	potassium chloride
MRI	magnetic resonance imaging
PPN	partial parenteral nutrition
SDH	sorbital dehydrogenase

Abnormalities identified on clinical pathology included decreased serum activity of SDH and AST and serum concentration of cholesterol, BUN, and albumin, most likely a result of decreased feed intake. No fecal parasite ova were observed and serum bile acids were within the normal limits. Virus neutralization testing for EHV-1, West Nile virus, and cELISA for Bluetongue virus from blood were all negative. CSF protein concentration, cell count, and cell morphology were within normal limits.

Because of the continued anorexia, the alpaca was transfaunated with 2 L of rumen fluid and administered IV lactated Ringer's solution supplemented with KCl (20 mEqs/L) and Vitamin B complex (2 mls/L) at 1.3× maintenance (4 L/d). Ceftiofur sodium (1.1 mg/kg IV q12h) and flunixin meglumine (1.1 mg/kg IV q12h) were administered. Because of the dam's lack of milk production and substantial time spent in recumbency, the cria was weaned.

Magnetic resonance imaging (MRI) examination revealed malformation of the caudal occipital bone with herniation/caudal displacement of the caudal cerebellum through the foramen magnum. There was also a T2-weighted hyperintensity within the cervical spinal cord (from C1-C3) that became hypointense on the T1-weighted images consistent with syringohydromyelia (Fig 1). Follow-up neurologic examination findings were consistent with the changes found on MRI examination. CP deficits were most likely caused by the extensive syringohydromyelia in the cervical spinal

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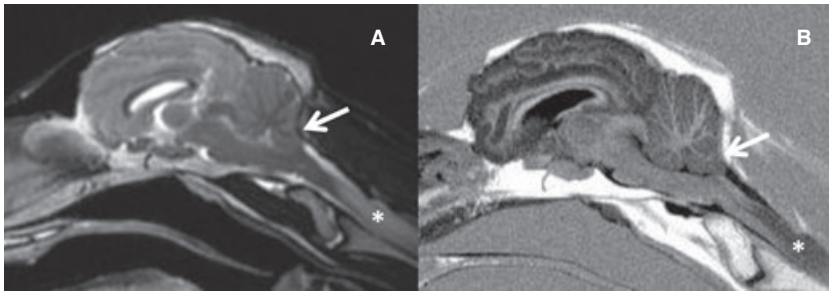

Fig 1. Sagittal T2-weighted (A) and T1-weighted Inversion Recovery (B) images of the brain and cervical spinal cord. There is a malformation of the caudal occipital bone causing caudal displacement of the cerebellum through the foramen magnum (arrow) and syringohydromyelia (asterisk) in the cervical spinal cord.

cord. The lack of menace response was likely caused by displacement of the cerebellum caudally through the foramen magnum. Options were provided to the owner and included medical management as well as surgical decompression of the occipital area. Though documentation of this condition or surgical outcome of this condition in alpacas was unavailable, the owners agreed to surgical decompression of the occipital/foramen magnum region.

The alpaca remained anorexic after the MR evaluation and up to the time of surgery, and was provided a noncommercial partial parenteral nutritional solution providing 1,322 Kcal/d (PPN) with frequent monitoring of blood glucose (q6h).

The alpaca was anesthetized with 50 mg of propofol IV and maintained on isoflurane gas. A routine approach was made to the dorsal aspects of the foramen magnum. A suboccipital craniectomy was performed to decompress the foramen magnum. Fibrous soft tissues and thickened dura overlying the caudal cerebellum and brainstem were removed. A temporalis facial graft was placed over the craniectomy defect. The dorsal cervical musculature, subcutaneous tissues, and skin were closed in a routine manner.

The alpaca recovered from anesthesia and surgery without complications. Treatments consisting of PPN, fluids, antibiotics, and anti-inflammatory drugs were continued as previously described. Hydromorphone was administered for pain management (2.9 mg SC, q6h) for 2 treatments.

The alpaca became more alert, but remained anorexic over the next 3 days. She was maintained on PPN, fluids, antibiotics, and anti-inflammatory drugs. On the 3rd day after surgery, flunixin meglumine was reduced to 1.1 mg/kg q24h. The alpaca was also transfaunated with 2 L of rumen fluid.

Starting on the 4th day after surgery, the alpaca's strength and attitude improved sufficiently to the point where she was ambulating on her own. She became difficult to handle and started to spit when approached by the medical care team. Her menace response was normal and she was starting to nibble small amounts of hay. She received transfaunations of 1.5–2 L of rumen fluid every 3 days to supplement volatile fatty acids and re-establish C1 flora. Treatment with PPN and antibiotics were continued as described above. Flunixin meglumine was discontinued on day 10. By day 11, the alpaca was allowed outside to graze. From day 11 to day 28, she increased her intake and water consumption enough to start gaining weight. A CBC and chemistry panel were repeated on day 17. All abnormal serum biochemical and hematologic findings had returned to normal except for albumin serum concentration. This was attributed to the continued lack of protein consumption by the alpaca. On day 28, it was deemed that her feed consumption and water intake were enough to discontinue PPN and transfaunations.

The alpaca continued to eat and drink well, gain weight, and maintain hydration off of PPN and fluids for the next 3 days. A transabdominal ultrasound examination was performed and confirmed a live 45-day-old fetus. The alpaca was discharged from the hospital with instructions to monitor feed and water consumption. The alpaca carried her fetus to term and had a normal cria. The alpaca had no important clinical problems during the 2 years after surgery and has been sold and is reported to be healthy to date.

Discussion

Caudal Occipital Malformation Syndrome (COMS), also referred to by various names including Chiari malformation, is recognized in a number of species including humans. Certain breeds of dogs, particularly Cavalier King Charles Spaniel, have been described with a condition analogous to Chiari type 1 malformation of humans.[1] This syndrome typically consists of a malformed occipital bone with various skeletal abnormalities at the craniocervical junction, compression of the hindbrain and subsequent caudal herniation of the caudoventral cerebellum through the foramen magnum into the cervical canal. Formation of the syringohydromyelia is thought to occur because of disturbance of normal cerebral spinal fluid dynamics in the subarachnoid space of the vertebral canal causing a pressure differential between cranial and spinal compartments. The pressure differential causes "clefts or fissures" in the spinal cord. Over time, these clefts will conform into cavities filled with low-protein fluid within the spinal cord called syringohydromyelia.[2] The chronic bony compression at the foramen magnum and the

resultant turbulent CSF flow and pressure changes in this region is also thought to result in hypertrophy and fibrosis of the underlying meninges over time resulting in further nervous tissue compression.[3]

Dogs with COMS typically present as young adults before they are 4 years old. The most common neurologic presentations are cervical myelopathy and cerebellovestibular dysfunction,[2–5] similar to the alpaca presented here. MRI is the preferred imaging modality for diagnosing COMS and syringohydromyelia. Treatment for COMS can be both medical and surgical. Medical treatment for COMS includes analgesic drugs, corticosteroids, omeprazole, acetazolamide, and furosemide used to target inflammation and to decrease CSF production.[2] Surgical treatments commonly focus on foramen magnum decompression (FMD).[2] Surgical success is generally favorable for improvement in clinical signs, although relapse rates ranging from 8 to 30% have been reported in people.[2] Most of the relapses are suspected to be caused by excessive postoperative scar tissue formation at the FMD site.[5,6]

Similar malformations referred to as Chiari type II formation or Arnold-Chiari-like formations involving a small caudal fossa of the cranial cavity and an enlarged foramen magnum have been documented in lambs and calves.[7–9] These malformations are typically accompanied by other malformations such as spina bifida, arthrogryposis, and lumbar kyphoscoliosis.[9] The clinical presentation of the alpaca in this case more closely resembled the canine form of this disease than the bovine or ovine cases reported, with a later onset of clinical signs and no other grossly obvious congenital anomalies.

The clinical presentation excluding respiratory distress and MR findings is consistent with the diagnosis of a caudal fossa/foramen magnum abnormality with caudal displacement of the cerebellum in this animal. The respiratory distress observed by the referring veterinarian was presumed to be idiopathic dorsal displacement of the soft palate and probably unrelated to this condition. Surgical treatment was associated with improvement in the clinical condition. Based on this single observation, however, it cannot be definitely concluded that clinical improvement was exclusively associated with surgical decompression. Considering the clinical progression and decline before surgery, and the recovery after, it is clinically logical that the surgical treatment benefited the clinical condition. Based on this case observation, alpacas with similar clinical presentations should reasonably be evaluated for similar abnormalities. Previous to surgery, the owners were advised that the condition treated in their alpaca could be heritable. In hindsight knowing more about the heritability of this condition in dogs, we should have recommended that they not rebreed this animal or its offspring.

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Conflict of Interest: Authors disclose no conflict of interest.

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