Chiari-Like Malformation with Syringomyelia in the Small Breed Dogs: Prognosis Evaluation after Surgical Management (Case Report)

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Abstract: Small breed dogs were visited for the evaluation of neurologic deficits. On neurologic examination, all dogs were showed hyperesthesia, pain, neck stiffness or ataxia. After diagnosis by magnetic resonance imaging (MRI) dogs had cranial cervical decompression with durotomy. These dogs had the durotomy patched with autologous subcutaneous fat. The clinical signs of all dogs improved postoperatively. This case report demonstrates to evaluate success of cranial cervical decompression for management of canine chiari-like malformation with syringomyelia (CM/SM).

Key words: Canine • Syringomyelia • Canine Chiari-Like Malformation • Cervical Decompression • Prognosis

INTRODUCTION

Syringomyelia (SM), frequently found in association with myelomeningocele, is an extremely serious condition in which fluid-filled cavities develop within the spinal cord near the brain as a consequence of obstruction to cerebrospinal fluid (CSF) movement [1-4]. It is also known as “neck scratcher’s disease”, because one of its common signs is scratching in the air near the neck. Increasingly recognized in dogs, one of the most common causes is chiari-like malformation-defined as decreased caudal fossa volume with caudal descent of the cerebellum and often the brainstem, into the vertebral canal. The condition is characterized by a mismatch between the caudal fossa volume and its contents, the cerebellum and brainstem. Consequently, there is insufficient space for the neural structure, which is forced caudally. This causes obstruction of the foramen magnum and the pressure wave of CSF emanating from the head during arterial pulsations [1-6]. The pathogenesis of syringomyelia remains controversial. There is increasing agreement that the syrinx fluid is not CSF and is most likely extracellular fluid that coalesces within the central canal and spinal cord substance as a consequence of abnormal pressure differentials between the spinal cord and subarachnoid space [6]. Although many congenital defects are recognizable before birth, at birth [7], SM seldom can be detected in young dogs, as symptoms of it usually are not evident before the age of six months or years later. Pain is the most important clinical sign of the disorder. Pain may be difficult to localize on clinical examination as it is often intermittent. Symptoms may vary widely among different dogs, but the earliest sign often is that the dog feels hypersensitivity in its neck area, causing in some an uncontrollable urge to scratch at its neck and shoulders. Then usually follows severe pain around its head, neck and shoulder, causing it yelp or scream [8, 9]. As the disease progresses, it destroys portions of the spinal cord and is so painful that the affected dog may contort its neck and even sleep and eat only with its head held high. The dog’s legs may become progressively weaker, so that walking becomes increasingly difficult. Some dogs deteriorate to the point of paraparesis or paralysis [1-3, 10]. Surgical therapy has been recommended to improve the dog’s quality of life and to retard clinical and radiographic progression of the syrinx [6, 11, 12]. The most common procedure is cranial cervical decompression (Also described as suboccipital...
decompression or foramen magnum decompression) in which most of the supraoccipital bone and dorsal laminae of the atlas are removed (With or without durotomy) to decompress with 4 small breed dogs which had surgical management of CM/SM in these cases. Diagnosis was made by neurologic examination and MRI of the brain and cervical spinal cord. CSF analysis was not performed because of the perceived risk in dogs with a cerebellar herniation and wide syringomyelia (Fig. 1). Dogs were positioned in sternal recumbency and the head was flexed. A dorsal approach was made to the supraoccipital bone and atlas. The rhomboideus, splenius and occipital muscles were reflected using a periosteal elevator. Hemorrhage was controlled using bipolar electrocautery. A supraoccipital craniectomy and partial C1 laminectomy were performed with bone removed until egg-shell thickness using burr and then removed with rongeurs. Stay sutures (8-0 polyglactin 910) were placed in the dura. Using the stay suture to identify and support the outer meninges, the dura and subarachnoid membranes were incised in a cruciate shape because this allowed the best visual decompression of cerebellum and medulla. The resulting kite-shaped defect in the meninges was either left open or covered with autologous subcutaneous fat (Fig. 2 and 3). After surgical management, dogs were administered 2 µg/kg/hr fentanyl for 12 hours and 25 µg fentanyl patches for 48 hours. Also dogs were administered oral cefalexin (20-30 mg/kg twice daily), carprofen (2-4 mg/kg daily) for 7 days. Surgery was associated with a low morbidity. All dogs made a quick recovery after surgery and were able to exercise normally within 1 week. One dog was slightly hypermetric and ataxic the day after surgery; this resolved within 72 hours. One dog was euthanatized because of necrotizing meningoencephalitis 4 months after surgery but at that time maintained an improved status. Successful surgical management could be defined as a procedure with low mortality and morbidity where postoperatively, the syrinx has resolved, the patient is free of clinical signs and the syrinx does not recur. Like previous reports, this study suggests that cranial cervical decompression is a safe procedure if performed by surgeon with appropriate neurosurgical training. Intraoperative complications were minimal and confined to meningeal vessel bleeding which was rapidly controlled with bipolar electrocautery and hemostatic aids. Surgery was associated with low morbidity; all dogs had a quick recovery and were able to exercise normally within 1 week after surgery. However, although 100% of the dogs had postoperative improvement, this surgical procedure seemingly did not result in syrinx resolution (Fig. 4). Similar findings were described by Rusbridge et al. [2], the reason for failure of this surgical technique to adequately correct the syringomyelia is unclear [8]. In the previous reports, possible explanation is that once formed, a large syrinx may self perpetuate because normal CSF flow cannot be re-established through the narrowed subarachnoid space or possibly because the dogs with chronic syringomyelia is not compliant enough to allow syrinx collapse and resolution. Modifications and improvements to the surgical technique may be possible. Previously, many authors described a technique of cranial cervical decompression combined with a titanium polymethylmethacrylate plate over the bony defect [11, 13]. It is unknown whether this technique will be more successful.

Fig. 1: MRI features of the dog with COMS. Syrinx in fourth ventricle and C1-5 cervical spinal canal (arrows) were noted on the midsagittal T2 weighted images (A). Transverse T1 (B) and T2 (C) weighted images at the level of the third cervical vertebrae revealed syrinx with an enlarged central canal.
Fig. 2: Before cranial cervical decompression (A). Cranial cervical decompression (B). The dura and arachnoid meninges have been incised. The spinal cord and the cerebellar vermis can be visualized through the meningeal defect.

Fig. 3: 3-D CT scan of pre-operation (A) and post-operation (B).

Fig. 4: Transverse CT scan image of the brain preoperatively (A) and 3 months postoperatively (B). The dog was clinically improved at the time of the CT scan; however, the enlarged ventricle did not resolve significantly.
An alternative method of managing syringomyelia is direct shunting of the cavity. However, in humans, this is not a preferred management technique for CM/SM as long-term outcome is poor because of shunt obstruction and/or spinal cord tethering [14]. In this report, cranial cervical decompression surgery for CM/SM has low mortality and morbidity and in dogs with pain the procedure can improve quality of life; however, syrinx collapse seemingly does not occur and resolution may be related to other factors involved in the development and/or persistence of syringomyelia. Surgical management may be regarded as not resolve the syringomyelia, but prevent the deterioration of syringomyelia. Therefore, the timing of surgical management is very important for the good prognosis. Further, based on these dogs, it is suggested that to properly evaluate the success of any surgical technique for CM/SM that postoperative MRI scans be performed because clinical improvement does not necessarily imply radiographic improvement.

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REFERENCES