Houston We Have A Praablem

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Introduction

Persistent right aortic arch (PRAA) is the most common vascular ring anomaly in dogs, especially German Shepherds. This disease is due to the right aortic arch failing to regress normally, entrapping the esophagus and trachea near the heart base on the right side. On the left and dorsally, the ligamentum arteriosum constricts the esophagus and the heart base is the ventral constricting structure. Due to the constriction of the esophagus by these three structures and the resultant megaesophagus, the most common sign of persistent right aortic arch is persistent regurgitation, often beginning at the time of weaning. Additionally, aspiration pneumonia is a common sequela in affected animals. Transection of the ligamentum arteriosum is the treatment of choice. Diagnosis can be achieved via radiograph- leftward deviation of the trachea on dorsoventral/ ventrodorsal view is both highly specific and sensitive for diagnosis of persistent right aortic arch. Overall, approximately 80% of pets who receive corrective surgery for PRAA have good resolution and owners who report satisfaction with the correction.

History and Presentation

A 7-week-old, female intact German Shepherd, Kimber, presented to MSU-CVM Small Animal Internal Medicine service on June 25, 2020. Her owners reported that she had been consistently regurgitating food since receiving the puppy, which was approximately 3 weeks prior to presentation. Initial physical exam revealed a weight of 4.70 kg and a body condition score of 4/9, a pulse of 108, respiration rate of 28 and a temperature of 101.7 F. She was bright, alert and responsive and mucous membranes were pink with a capillary refill time of less than 2 seconds. Peripheral lymph nodes were normal and the abdomen was soft and nonpainful on palpation. Radiographs performed on 6-25-2020 suggested a persistent right aortic arch and surgical correction was recommended. On July 8th Kimber presented on emergency for worsening regurgitation and weight loss. Since June 25 Kimber has been unable to keep food down despite being on a liquid diet. A few days prior to the July 9th presentation, Kimber has been vomiting and regurgitating constantly after getting into the trash. Physical exam revealed a weight of 4.60 kg, temperature of 101.4 F, pulse of 138 and respiratory rate of 48. Since June 25th, Kimber's body condition had depreciated noticeably. No fluid was seen on thoracic nor abdominal FAST scan. Final presentation occurred July 13th to the Small Animal Surgery Service for CT and surgical correction. At this time, Kimber's body condition had improved and she weighed 5.2 kg, Her temperature was 102.6 F, pulse was 92 and respiration rate 24.

Diagnostics

The diagnosis of PRAA begins with the patient's signalment. Breeds such as German Shepherds, Great Danes and Irish Setters are often affected. Additionally, these patients are young; the transition period between weening to solid food seems to be when a persistent right aortic arch becomes problematic. Patients with PRAA will likely be presenting for "vomiting" described by owners but will be determined to actually be regurgitation by a veterinarian. A lack of abdominal effort/ heaving will often differentiate the two. Additionally, PRAA patients may present with respiratory signs. These are generally the result of aspiration pneumonia or more rarely the presence of an additional vascular ring encircling the trachea. Patients with PRAA may be of poor body condition due to constant regurgitation as well, despite having appetites which are often ravenous.

Additional diagnostics which should be pursued includes a CBC and a chemistry in order to determine whether the afflicted pet is a good candidate for surgical correction. Abnormalities such as hypoglycemia and neutrophilia/penia can be seen due to poor nutrition and infection. These would require correction prior to surgery. Additionally, PCV, total protein and clotting ability need to be assessed.

Thoracic radiographs will reveal ventral deviation of the trachea in the cranial thorax on the lateral projection due to cranial megaesophagus. Additionally, and most importantly, the ventrodorsal projection will show a leftward deviation of the trachea in the cranial thorax which is very characteristic of a PRAA. This is due to the aorta developing on the right side of the trachea, instead of the left, as it should. Other finding on thoracic radiographs can include widening of the cranial mediastinum and cranial megaesophagus- filled with air and/ or ingesta showing characteristic constriction at the level of the heart base.

Once megaesophagus is determined, a CT angiogram can definitively determine the presence and type of vascular ring anomaly in the patient and what structure(s) is/are causing the esophageal constriction. Intravenous contrast is used during the CT scan in order to outline the vascular structures.

Kimber received thoracic 3 view radiographs for the first time on June 25th, 2020, revealing gas within the cranial thoracic esophagus. The presence of gas was responsible for dilation of the esophagus and the aorta being on the right side caused the leftward deviation of the cranial intrathoracic trachea. The cranial esophageal dilatation combined with the leftward deviation of the trachea indicated that a vascular ring anomaly is the most likely culprit.

Thoracic radiographs were repeated on July 9th due to concern for aspiration pneumonia. Radiographs revealed that the previously described ventral deviation of the cranial intrathoracic trachea and focal dilatation of the cranial intrathoracic esophagus had markedly progressed. There were no signs of aspiration pneumonia.

Kimber was referred to CT on July 13. Diagnostic CT imaging revealed the ascending aorta was lying on the right side, instead of the left as it should, as well as leftward deviation of the cranial intrathoracic trachea. Additionally, there was an aberrant left subclavian artery originating from the proximal descending aorta. The cranial intrathoracic esophagus, cranial to the region of the ligamentum arteriosum and the origin of the left subclavian artery was dilated. The brachiocephalic trunk was absent and the left and right common carotids and the right subclavian artery were originating adjacent to each other from the aortic arch, separately.

Based on our radiographic findings- ventral deviation of the trachea on the lateral projections and the leftward deviation of the trachea as well as the cranial megaesophagus on the

ventrodorsal view - we were confident in our diagnosis of a vascular ring anomaly in Kimber. The CT scan revealed additional abnormalities in Kimber allowing for a more ideal surgical plan to be made. The CT revealed the additional aberrant left subclavian artery. These findings definitively revealed the cause of Kimber's esophageal constriction and secondary cranial megaesophagus and regurgitation.

Pathophysiology

During embryonic maturation, there are six pairs of aortic arches that undergo metamorphosis in order to form the major vessels of the thorax, neck and head. Vascular ring anomalies are the abnormal embryonic development of these aortic arches around structures in the thorax such as the esophagus and trachea. In the normal embryo, the arterial circulation in the thorax is comprised of paired ventral aortas and paired dorsal aortas and an intercommunicating series of six paired aortic arches that surround the primordial esophagus and trachea. The paired ventral and dorsal aortas eventually fuse caudally to form the heart and descending aorta, respectively.

Theoretically, the left fourth arch should enlarge to form the normal adult aortic arch. However, when the right fourth aortic arch enlarged and becomes the functional adult aorta instead, a persistent right aortic arch occurs. The left arch will instead form into the proximal portion of the left subclavian artery. Because the aorta is on the opposite side of the esophagus as the pulmonary artery, when the ductus arteriosus is replaced by the ligamentum arteriosum, a constricting band of fibrous tissue is present over the esophagus.

Constriction of the esophagus causes megaesophagus and results in regurgitation and poor body condition in these patients.

Treatment

Treatment of persistent right aortic arch is always surgical. Megaesophagus can be managed medically, however, this is not sustainable. Poor body condition and aspiration pneumonia make it impossible for these patients to live long without correction. The surgical procedure consists of ligating and transecting the ligamentum arteriosum and breaking down adjacent fibrous tissue as well as other aberrant/ constricting structures (i.e., the aberrant left subclavian artery) and ensuring that the esophagus is then free from constriction. After surgical correction, the patient is managed as a megaesophagus patient initially and then slowly transitioned to a more normal diet. Prognosis for surgical correction is excellent in most patients (Avg 80% return to normal function) and is inversely correlated with patient age, i.e., younger patients receiving surgical correction have a better prognosis. However, post-operative management is key to ensuring a good outcome.

Kimber's surgical operation began in right lateral recumbency and the site was aseptically prepped with chlorhexidine scrub and draped. The initial plan was to thoracoscopically ligate and transect Kimber's PRAA and aberrant left subclavian artery. We were unable to adequately/ safely access the ligamentum arteriosum safely because of Kimber's small size and opted to convert to an open procedure. A 4^{th} intercostal space thoracotomy was used. The cranial left lung lobe was retracted caudally and packed off with moist laparotomy sponges. Exposure to the mediastinum was obtained dorsal to the heart and the vagus and phrenic nerves were identified. Additionally, the esophagus, along with the aorta was also identified. A combination of sharp and blunt dissection was used in order to dissect through the

mediastinum and reach the ligamentum arteriosum. Liga- Clips were utilized to ligate the ligamentum arteriosum. The aberrant left subclavian artery was also ligated, however, using 2-0 silk braided suture at the bifurcation of the left subclavian and the ligamentum arteriosum. In order to obtain hemostasis at this point, vascular clamps were utilized. Next, a tube was passed into the esophagus to identify any additional constricting tissue. All identified fibrous tissue found to be constricting the esophagus was then carefully dissected. The thorax was then lavaged with warm saline and the cranial lung was repositioned. Finally, a thoracostomy tube was placed and the thorax is closed. Nocita was placed subcutaneously for post- operative local analgesia. Anesthesia and recovery were uneventful.

Case Outcome

Kimber was discharged to go home on July 15th and faced several challenges post operatively. Initially Kimber was reported to have been doing well on her diet (1/2 cup dry food blended with 1 cup water) but was still underweight. In August, owners tried to increase her feed intake up to 1 cup of dry food blended with 1 cup of water but it proved to be too much too fast. The increased food volume was unable to adequately pass through the esophagus and Kimber aspirated. She was sent to the emergency room in Hattiesburg, MS where veterinarians reported that Kimber was unable to keep food down and recommended an esophageal ballooning procedure. Owners were hesitant to return to Mississippi State CVM. We recommended that the esophagus be scoped by the Internal Medicine Service in order to determine whether the esophagus had strictured, or whether the dilatation that was present was going to be permanent. Owners were advised to return in September for a follow up. They did not schedule an

appointment or return. The owners report that Kimber passed away in February after consuming daffodils.

Conclusion

Persistent right aortic arch is a vascular ring anomaly that affects young dogs primarily, especially during the weaning period. Chronic regurgitation will be the primary sign of the presence of esophageal disease/ disorder. Radiographs will show very characteristic signs of vascular ring anomaly, especially if it happens to be a persistent right aortic arch (the most common vascular ring anomaly in dogs). These signs, including megaesophagus on the lateral view and leftward deviation of the trachea in the cranial thorax on the ventrodorsal view, are pathognomonic for a persistent right aortic arch. Further imaging should be pursued in order to detect any other vascular anomalies, such as an aberrant left subclavian artery (the second most common vascular ring anomaly) and to make plans for corrective surgery, as in Kimber's case. Corrective surgery can be done thoracoscopically but often consists of a left 4^{th} intercostal space thoracotomy in order to access the ligamentum arteriosum and aberrant left subclavian artery and to ligate them, freeing the esophagus from constriction. In Kimber's case, complete thoracoscopic resolution proved to be impossible due to her size, however, the ligamentum arteriosum and aberrant left subclavian artery were successfully ligated via left lateral thoracotomy and anesthetic recovery was uneventful. Kimber's issues during the months following her surgery would likely have been prevented through better management practices; in most cases, the prognosis for persistent right aortic arch surgical repair is excellent, as is return to function.

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