Peritoneopericardial Diaphragmatic Hernia in an 8 year old Bloodhound

Stefanie M. Wong
Clinical Advisor: Dr. James Flanders
Basic Sciences Advisor: Dr. Linda Mizer

Senior Seminar Paper
Cornell University College of Veterinary Medicine
February 15, 2012

Keywords: dog, congenital, peritoneopericardial, diaphragmatic, hernia, surgery

Abstract

An 8 year old, male castrated Bloodhound presented to the Cornell University Hospital for Animals Soft Tissue Surgery service for evaluation of a chest mass and nine month history of a progressive cough. The mass was identified on radiographs taken by the referring veterinarian. On presentation, the dog had a mildly increased respiratory rate and effort. Cardiothoracic auscultation revealed decreased to absent heart sounds over the left thorax, with a point of maximal intensity on the right. The dog also had decreased to absent lung sounds ventrally. Based on evaluation of three-view radiographs, a diagnosis of a Peritoneopericardial Diaphragmatic Hernia (PPDH) was made. Abdominal exploratory and surgical correction of the hernia was performed. Incidental findings of multifocal splenic myelolipomas, diffuse vacuolar hepatopathy of the right medial lobe, and loss of architecture of the quadrate lobe were found on histopathology. Four months following surgery, the patient was doing well.

Introduction

Peritoneopericardial Diaphragmatic Hernia (PPDH) is the most common congenital pericardial defect in dogs and cats[1,2], as well as the most common congenital diaphragmatic defect in small animals[2]. It arises as a result of abnormal embryonic development of the diaphragm. The diaphragm forms from the fusion of three embryonic structures: the septum transversum, the caudal mediastinum and the pleuroperitoneal folds[3]. The septum transversum makes up the ventral portion of the embryonic diaphragm and will eventually become the central tendon. The central portion of the embryonic diaphragm, the caudal mediastinum, contains the esophagus and caudal vena cava. The

pleuroperitoneal folds are folds of tissue that arise from the dorsolateral margin of the body wall and extend into the peritoneal cavity. These folds expand to fuse ventrally with the caudal mediastinum and septum transversum to form the diaphragm.

There are several theories that propose how the abnormal embryogenesis of the diaphragm takes place to result in a PPDH. It is thought to arise from: (1) failure of the lateral pleuroperitoneal folds and ventromedial pars sternalis to unite during division of the coelom into abdominal and thoracic cavities[4], (2) faulty development of the dorsolateral septum transversum or rupture of a thin tissue membrane in this area allowing peritoneal and pericardial communication[5,6], or (3) prenatal injury to the septum transversum or to the fusion site of the septum transversum and pleuroperitoneal folds[7] . In the dog and the cat, PPDH cannot be acquired secondary to trauma, in contrast to humans. This results in a persistent opening between the pericardial and peritoneal cavities, allowing for cranial displacement of abdominal contents into the pericardial sac. This paper describes a case of a PPDH in a coughing dog, which was initially diagnosed as a thoracic mass.

Case History

An 8 year old, male castrated Bloodhound presented to Cornell University Hospital for Animals (CUHA) Soft Tissue Surgery service September 21, 2011 for evaluation of a thoracic mass. The patient initially presented to his primary care veterinarian in September 2011 for a progressive cough of nine months duration. On physical exam, the referring veterinarian could not hear the heart over the left thorax and took three-view thoracic radiographs, which showed a large opacity obliterating the majority of the

thoracic cavity. A Complete Blood Count and serum Chemistry were also run at this time, which showed a mild leukopenia and mild hyperglobulinemia. The bloodwork was otherwise normal. The patient was then referred to Cornell for further evaluation. The owners described his cough as a non-productive, dry, hacking cough. The patient had a history of allergic dermatitis and arthritis but was otherwise a healthy dog, with good appetite and energy level and no current medications.

Clinical Findings

On physical examination, the patient was bright, alert and responsive. His mucous membranes were pink with a capillary refill time of < 2 seconds. He had lenticular sclerosis bilaterally and oral exam revealed moderate dental calculus and gingivitis. He had carpal hyperextension bilaterally, symmetrical pelvic limb muscle atrophy and crepitus in multiple joints. On cardiothoracic auscultation, he had decreased to absent heart sounds over his left thorax. His heart was auscultable on the right, with a point of maximal intensity over the right 5^{th} intercostal space. No murmurs or arrhythmias were heard and his pulses were strong and synchronous. The patient was mildly tachypneic and mildly dyspneic. His lungs could be ausculted dorsally, but he had decreased to absent lung sounds ventrally. No crackles or wheezes were detected. He had a soft, non-painful abdomen, with no masses or organomegaly. The remainder of his physical exam was within normal limits.

Consultation with the Imaging service regarding his radiographs was pursued. Closer examination revealed absence of the falciform fat, cranial displacement of the gastric axis

and liver and irregular shape and density of his thoracic “mass.” All of these were consistent with a Peritoneopericardial Diaphragmatic Hernia, with displacement of abdominal contents into the pericardium.

Problem List

1) Coughing
2) Tachypnea and dyspnea
3) Decreased to absent heart and lung sounds
4) Thoracic “mass” seen on radiographs

Differential Diagnoses

1. Peritoneopericardial Diaphragmatic Hernia
2. Respiratory disease
    a. Collapsing trachea – less likely due to signalment, normal tracheal diameter with no evidence of collapse on radiographs
    b. Laryngeal paralysis – less likely due to lack of noisy, stertorous breathing
    c. Neoplasia
    d. Infectious (bacterial/viral/fungal/parasitic pneumonia) – less likely due to lack of fever or systemic illness, no crackles on auscultation
    e. Inflammatory (chronic bronchitis) – less likely as no wheezes ausculted
    f. Trauma – less likely due to lack of history of trauma
    g. Toxin – less likely due to lack of history of toxin ingestion
    h. Vascular (Pulmonary Thromboembolism) – less likely as not acute onset

i. Foreign body

j. Pleural effusion – less likely due to lack of radiographic evidence

k. Pneumothorax, hemothorax, chylothorax – less likely due to lack of radiographic evidence

l. Pulmonary edema (non-cardiogenic) – less likely due to lack of history of drowning, electrocution, seizure, head trauma, smoke, sepsis

3. Cardiac disease

    a. Pericardial effusion – less likely due to irregular margin of pericardium, no other signs of pericardial tamponade

    b. Left-sided heart failure causing pulmonary edema – less likely due to lack of murmur/crackles on auscultation

Treatment

Abdominal exploration and surgical correction were discussed with the patient's owners. The decision was made to send him to surgery the following day. Repeat thoracic radiographs, abdominal radiographs and abdominal ultrasound were performed prior to surgery, confirming the presence of a PPDH. Abdominal ultrasound revealed the presence of the spleen in the pericardium, surrounding the heart on both the right and left sides. A splenic mass was identified near the head of the spleen. A standard ventral midline approach was made. A defect in the external rectus sheath was found adjacent to the xiphoid. On abdominal exploration, a large ventral defect in the muscular part of the diaphragm was found with greater omentum and the mesenteric attachments of the spleen extending caudally. The entire spleen was within the pericardial sac. Digital palpation

revealed two adhesions between the diaphragm and omentum, which were broken down using electrocautery. The rent was extended ventrally into the central tendon to allow better visualization and easier spleen removal. Caudal traction was placed on the spleen and it was removed from the pericardial sac, along with the omentum.

The spleen was examined - the splenic mass previously identified on ultrasonic examination was visualized. The spleen was subjectively large with multiple areas of nodular hyperplasia. Due to concern about possible neoplasia and compartment syndrome if left in due to its large size, a splenectomy was performed and the spleen was submitted for histopathology.

A stab incision was made into the pleura and extended to protect against pericardial tamponade that may result secondary to pericardial effusion. The caudal edges of the lungs could be visualized. They began to fully expand following removal of the spleen and were noted to be a healthy pink color and SpO2 measurements, which previously had been between 70-80%, went up to 100%.

The central tendon was reapposed with a simple continuous pattern. The edges of the portion of the muscular diaphragm were debrided and apposed back together with an interrupted cruciate pattern. A red rubber tube connected to a stopcock and syringe was placed in the pleural cavity to restore negative pressure. After no more air could be aspirated back, it was removed. The liver was examined and the left liver lobes, especially the quadrate lobe, appeared to be hypoplastic – they were small, pale, firm and

rubbery. By contrast, the right liver lobes appeared to be hypertrophied, extending over mid-abdomen to the left kidney. They were cobblestoned with a grayish red tinge. Biopsies were taken of the quadrate and right medial liver lobes and submitted for histopathology. The remainder of the abdomen was examined and no other abnormalities were found. The abdomen was closed in a standard fashion.

The patient recovered uneventfully from anesthesia and was placed in the ICU. Overnight he became anxious, tachypneic and dyspneic. Multiform Ventricular Premature Complexes (VPCs) and 1st and 2nd degree heart block were observed on continuous electrocardiogram (ECG) with a normal heart rate. An arterial Gaslyte was performed, which revealed hypokalemia (2.84, reference range 3.9-5.1) and hypoxemia (PaO2 67.6 mmHg, reference range 95-100 mmHg). The IV fluids were supplemented with potassium and he was placed on nasal oxygen, which resolved his cardiac arrhythmia.

The following morning, the patient was quiet, alert and responsive. He had a normal respiratory rate and effort. Repeat arterial Gaslyte off oxygen revealed resolution of the hypokalemia with a persistent hypoxemia (PaO2 79.1 mmHg). Crackles were ausculted over his right and left lung fields, so thoracic radiographs were taken. Thoracic radiographs showed no overt abnormalities, with no pulmonary edema or aspiration pneumonia seen. Overnight, pulse oximeter measurements were 99% on room air and no further cardiac arrhythmias were seen, so nasal oxygen and continuous ECG were discontinued.

The patient remained in the ICU for observation for two more days. On Day 4 post-op, a grade III/VI left-sided cardiac murmur was ausculted. In order to further evaluate the murmur, a Cardiology consultation was performed. Echocardiogram revealed both mitral and triscupid regurgitation, but no evidence of endocarditis. A repeat Complete Blood Count was performed, which was within normal limits. The following day he was discharged into the care of his owners.

Outcome

Histopathology of the spleen revealed multifocal aggregates of adipocytes surrounded by myeloid and erythroid precursors, consistent with multifocal myelolipomas. The quadrate lobe, which had been grossly small, pale, and firm in consistency, had diffuse absence of normal hepatic architecture. Occasional bile ducts and arteriolar profiles were present, surrounded by fibrous connective tissue. A reticulin stain, which stains reticular fibers within hepatocellular basement membranes, revealed moderate to marked hepatocellular parenchymal collapse surrounding the portal areas and central veins. These changes were proposed to most likely be secondary to compression of venous blood flow to the quadrate lobe over a very long period of time. The right medial lobe, which had been grossly large and abnormal in texture and color, was noted to show changes consistent with a moderate degenerative vacuolar hepatopathy.

Four months following his discharge, the patient was doing great. According to his owners, he was able to go on longer walks and was able to breathe much more

comfortably. He no longer had any coughing and had an improved energy and appetite at home.

Discussion

Dogs with Peritoneopericardial Diaphragmatic Hernias can live with this abnormality for many years and remain asymptomatic. Often, it is a completely incidental finding[4,6,9,10]. Patients are often diagnosed early in life, but a significant portion of patients is identified later, at eight years of age or older[8,10]. Although trauma cannot cause a PPDH, affected animals may be identified following a traumatic incident that causes a sudden shift of the abdominal contents into the pericardium, resulting in an acute onset of clinical signs. Alternatively, abdominal contents may move in and out of the pericardium periodically, causing intermittent presentation of signs.

Clinical signs are dependent on what abdominal contents herniate into the pericardium, with the most common clinical signs being gastrointestinal (anorexia, weight loss, vomiting, diarrhea) and respiratory (coughing, dyspnea, tachypnea)[2,8,11-14]. The most common herniated items are the liver and gallbladder, followed by spleen, intestine and stomach in decreasing frequency[2,8,13,14]. The severity of these signs range from non-existent to life threatening. Other potential clinical findings may include bowel sounds on thoracic auscultation as well as signs of cardiac tamponade and right-sided heart failure. With asymptomatic dogs, common physical exam findings are muffled heart and lung sounds or a thin abdomen[2,8,13].

Thoracic radiographs showing generalized cardiomegaly, dorsal displacement of the trachea, and inability to distinguish the border between the caudal cardiac silhouette and ventral diaphragm often increase the index of suspicion. With intestinal displacement, gas-filled loops of bowel may be seen silhouetting with the pericardium. Cranial displacement of abdominal organs, particularly the liver and stomach may be seen. Ultrasound is now often used instead of contrast studies and can be used to confirm the diagnosis of a PPDH, as dogs with PPDH may appear radiographically normal[8]. Small hernias of fat or omentum only can be difficult to diagnose with any imaging technique[15]. Echocardiography may be indicated, as PPDH has been found concurrently with other congenital cardiac conditions, such as aortic stenosis, patent foramen ovale, ventricular septal defect and pulmonic stenosis[2,8,13]. Bloodwork consisting of a Complete Blood Count and serum Chemistry are often unremarkable. The most common abnormality seen is an elevated alanine aminotransferase (ALT), seen with liver herniation[13].

Surgical correction of the defect is recommended, as the risk always exists for entrapment of stomach and small intestine. Concurrent congenital midline abnormalities, umbilical hernias being the most common, can be corrected at that time. Sternal abnormalities have also been reported[8,13]. Conservative treatment may be chosen by owners due to lack of clinical signs associated with the PPDH, advanced age at diagnosis, concurrent major disease, or morbidity and mortality associated with anesthesia and surgery[2].

Myelolipomas are rare, benign tumors composed of a mixture of well-differentiated adipose and hematopoetic tissue. Myelolipomas associated with PPDH have been

documented in cats[16,17]. It is hypothesized that their formation is a metaplastic change in response to chronic hypoxia caused by entrapment within the pericardial sac[17-20]. Myelolipomas are often reported in geriatric dogs, suggestive of a relationship between their development and advanced age. They are most commonly seen in the adrenal glands, spleen and spine[21-24] and are often incidental findings[21].

The liver changes seen in this patient are likely the result of compromised hepatic perfusion associated with the altered orientation of the abdominal viscera and vasculature due to the herniation of the spleen into the pericardial sac. Although vacuolar hepatopathies are often associated with a variety of diseases such as adrenal, hepatobiliary, cardiac, neurologic, gastrointestinal, immune-mediated, endocrine and renal disease[25], due to the absence of any other abnormalities on our physical exam, bloodwork and gross examination of the abdominal organs, it is most likely that our patient's hepatic changes are the result of his PPDH.

Prognosis

Long-term prognosis following surgery is excellent for return to normal function. Intraoperative complications include hypotension, hypoventilation, hypoxia and ventricular premature contractions (VPCs). These are most often seen with hepatic herniation, with release of free radicals following reperfusion of underperfused hepatic parenchyma[2]. Perioperative complications are often minor and self-limiting. Minor complications reported include transient hyperthermia in cats, incisional inflammation, and seroma formation. Possible major perioperative complications include pleural

effusion, oxygen dependency and pulmonary edema[2,13]. Reexpansion pulmonary edema is often seen after correction of traumatic diaphragmatic hernias with rapid reexpansion of chronically atelectatic lung. Although this has been seen in association with PPDH correction[2], it was not seen in this patient.

References

1. Berry, CR. Recognition of congenital heart disease in the adult dog and cat. In: Bonagura, JD, ed. *Kirk's Current Veterinary Therapy XII. Small Animal Practice.* 12th ed. Philadelphia: WB Saunders Co, 1995, 833-836.
2. Reimer, SB, Kyles, AE, Filipowicz, DE, Gregory, CR. Long-term outcome of cats treated conservatively or surgically for peritoneopericardial diaphragmatic hernia: 66 cases (1987-2002). *J Am Vet Med Assoc* 2004; 224, 728-732.
3. Noden, DM, De Lahunta, A. *Embryology of Domestic Animals: Developmental Mechanisms and Malformations*. 1st ed. Baltimore: Williams and Wilkins, 1985, 287-290.
4. Baker, GJ, Williams, CS. Diaphragmatic pericardial hernia in the dog. *Vet. Rec.* 1966; 78, 578-583.
5. Bolton, GR, Ettinger, S, Rousch, JC. Congenital peritoneopericardial diaphragmatic hernia in a dog. *J Am Vet Med Assoc* 1969; 155, 723-730.
6. Finn, JP, Martin, CL. Diaphragmatic pericardial hernia. *J Small Anim Pract* 1969; 10, 295-300.
7. Clinton, JM. A Case of congenital pericardio-peritoneal communication in a dog. *Vet Radiol Ultrasound* 1967; 8, 57-60.

8. Evans, SR, Biery, DN. Congenital peritoneopericardial diaphragmatic hernia in the dog and cat: a literature review and 17 additional case histories. *Vet Radiol Ultrasound* 1980; 21, 108-116.
9. Atkins, CE. Suspect congenital peritoneopericardial diaphragmatic hernia in an adult cat. *J Am Vet Med Assoc* 1974; 165, 175-176.
10. Weitz, J, Tilley, LP, Moldoff, D. Pericardiodiaphragmatic hernia in a dog. *J Am Vet Med Assoc* 1978; 173, 1336-1338.
11. Reed, JR. Pericardial diseases. In Fox, PR, ed. *Canine and Feline Cardiology*. 1st ed. New York: Churchill Livingstone, 1988, 495-518.
12. Hunt, GD, Johnson, KA. Diaphragmatic, Pericardial and Hiatal Hernia. In: Slatter, D, ed. *Textbook of Small Animal Surgery*. 3rd ed. Philadelphia: Saunders, 2003, 471-485.
13. Banz, AC, Gottfried, SD. Peritoneopericardial Diaphragmatic Hernia: A Retrospective Study of 31 Cats and Eight Dogs. *J Am Anim Hosp Assoc* 2010; 46, 398-404.
14. The Merck Veterinary Manual website. Peritoneopericardial Diaphragmatic Hernia (PPDH). Available at: http://www.merckvetmanual.com/mvm/index.jsp?cfile=htm/bc/11112.htm. Accessed January 14, 2012.
15. Kienle, RD, Thomas, WP. Echocardiography. In: Nyland, TG, Mattoon, JS, eds. *Small Animal Diagnostic Ultrasound*. 2nd ed. Philadelphia: WB Saunders Co, 1995, 412-414.

16. Gourley, IM, Popp, JA, Park, RD. Myelolipoma of the liver in a domestic cat. *J Am Vet Med Assoc* 1971; 158, 2053-2057.

17. Schuh, JCL. Hepatic Nodular Myelolipomatosis (Myelolipomas) associated with a peritoneo-pericardial diaphragmatic hernia in a cat. *J Comp Pathol* 1987; 97, 231-234.

18. Wouda, RM, Chalkley, MD, Fraser, AR, Moses, PAE. Hepatic myelolipoma incarcerated in a peritoneopericardial diaphragmatic hernia in a cat. *Aus Vet J* 2010; 88, 231-235.

19. Thamm, DH. Hepatobiliary tumors. In: Withrow, SJ, MacEwen, EG, eds. *Small Animal Clinical Oncology*. 3rd ed. Philadelphia. WB Saunders Co, 2001, 327-334.

20. Liptak, JM, Dernell, WS, Withrow, SJ. Liver Tumors in cats and dogs. *Compend Contin Educ Pract Vet* 2004; 26, 50-57.

21. Al-Rukibat, RK, Bani Ismail, ZA. Unusual presentation of splenic myelolipoma in a dog. *Can Vet J* 2006; 47, 1112-1114.

22. Newman, SJ, Inzana, K, Chickering, W. Extradural myelolipoma in a dog. *J Diagn Vet Invest* 2000; 12, 71-74.

23. Tursi, M, Iussich, S, Prunotto, M, Buracco, P. Adrenal myelolipoma in a dog. *Vet Pathol* 2005; 42, 232-235.

24. Zimmer, MA, Stair, EL. Splenic myelolipomas in two dogs. *Vet Pathol* 1983; 20: 637-638.

25. Sepesy, LM, Center, SA, Randolph, JF, Warner, KL, Erb, HN. Vacuolar hepatopathy in dogs: 336 cases (1993-2005). *J Am Vet Med Assoc* 2006; 229, 246-252.