What Is Your Diagnosis?

In collaboration with the American College of Veterinary Radiology

Figure 1—Left lateral (A) and dorsoventral (B) thoracic radiographic images of a 4-year-old spayed female Boston Terrier with a grade VI/VI systolic right apical heart murmur, grade III/VI diastolic left apical heart murmur, intermittent arrhythmia, respiratory distress, and lethargy.

History

A 4-year-old spayed female Boston Terrier was referred to the Louisiana State University Veterinary Teaching Hospital for evaluation of a heart murmur, respiratory distress, and lethargy. On physical examination, a VI/VI systolic right apical heart murmur, III/VI diastolic left apical heart murmur, and an intermittent arrhythmia were auscultated. Auscultation of lung sounds was obscured by the heart murmurs, and respiratory rate was moderately increased with mild effort. Femoral pulses were weak and synchronous with the heart sounds. Mucous membranes were pink with a capillary refill time of < 2 seconds. Thoracic radiography was performed (**Figure 1**).

Formulate differential diagnoses, then continue reading.

Diagnostic Imaging Findings and Interpretation

Thoracic radiography revealed a severely enlarged cardiac silhouette with predominantly left-sided

cardiomegaly (**Figure 2**). Straightening of the caudal cardiac silhouette with a large soft tissue convexity in the area of the left atrium, resulting in dorsal displacement of the intrathoracic trachea, was noted. On the dorsoventral view, there was a soft tissue bulge in the area of the left auricle and rounding in the region of the right ventricle. Furthermore, the pulmonary veins were mildly distended. A severe, generalized, interstitial pulmonary pattern that obscured the margins of the vasculature was noted in all lung lobes with bronchocentric pattern in the caudal lung lobes. A diagnosis of left-sided congestive heart failure (CHF) was made; differential diagnoses included mitral valve dysplasia, degenerative mitral valve disease, infective endocarditis, patent ductus arteriosus, ventricular septal defect, and subaortic stenosis.

On 2-D echocardiography (**Figure 3**), the left ventricular chamber dimensions and wall thicknesses were within reference limits. The left atrium was severely dilated and the left atrial diameter-to-aorta diameter ratio was 3.8 (4.6 cm/1.2 cm) with normal being < 1.5. The mitral valve leaflets were thickened, dysplastic, and severely stenotic. The leaflets appeared to be tethered to dysplastic papillary muscles via short and thick chordae tendineae. The

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Figure 2—Same images as in Figure 1. The cardiac silhouette is severely enlarged. It has a large soft tissue bulge in the region of the left atrium, resulting in severe dorsal displacement of the intrathoracic trachea (asterisk), and a soft tissue bulge is in the region of the left auricle (arrows). There is a severe interstitial pattern in the caudal lung lobes that obscures the vasculature margins.

Figure 3—Right parasternal long-axis echocardiographic images without (A) and with (B) color flow Doppler ultrasonography and right parasternal short-axis color M-mode echocardiographic image of the left apex at the level of the mitral valve (C) of the dog described in Figure 1. A—There is doming of the mitral valve leaflets (dagger) during diastole, indicating stenosis. B—Turbulent mitral inflow (double dagger) during diastole is also present. C—There is concordant motion of the anterior and posterior mitral valve leaflets and interrogation of the mitral valve shows turbulent diastolic flow (arrowheads).

right atrium and ventricle were subjectively normal. The tricuspid valvular leaflets were mildly thickened with no additional apparent structural abnormalities. Color flow Doppler echocardiography revealed

severe mitral valve regurgitation and turbulent mitral valve forward flow. Moderate tricuspid regurgitation, mild pulmonic insufficiency, and trace aortic insufficiency were also present. Turbulent, high-velocity mitral inflows (averaging 2.92 m/s; 34.11 mm Hg) were observed. Aortic flows were normal. Thoracic radiography and echocardiography findings were consistent with left-sided CHF secondary to severe mitral valve stenosis. The final diagnosis of mitral

valve stenosis with concurrent pulmonary hypertension and left-sided CHF was made based on these findings.

Treatment and Outcome

Furosemide (2 mg/kg, IV) was administered during the appointment, and continued treatment with furosemide (2.3 mg/kg, PO, q 12 h) was prescribed for the management of CHF. Additionally, treatment with spironolactone (1.5 mg/kg, PO, q 12 h), enalapril (0.4 mg/kg, PO, q 12 h), and clopidogrel (2.3 mg/kg, PO, q 24 h) were initiated. Avoidance of high-sodium food and treats and transition to a diet (Hill's Prescription diet h/d, Hill's Pet Nutrition Inc) with low sodium, high levels of L-carnitine and taurine, balanced levels of protein and phosphorus, and antioxidants were recommended. Additionally, recommendations included a renal panel in 1 to 2 weeks, echocardiography in 3 months, and continued monitoring for persistent cough, tachypnea, dyspnea, exercise intolerance, and syncope.

Comments

Mitral valve dysplasia is a congenital malformation of the mitral valve apparatus where pathology of leaflets, chordae tendineae, and papillary muscle causes failure of the mitral valve to properly close. Large breeds, such as Great Danes, Golden Retrievers, German Shepherd Dogs, Dalmatians, and Newfoundlands, are predisposed to mitral valve dysplasia.¹ The dog of the present report had mitral valve stenosis, which is a specific type of mitral valve dysplasia that can be congenital or acquired and is a rare, complex narrowing of the mitral valve orifice.¹ Mitral valve stenosis has been reported in Bull Terriers, Newfoundlands, and other breeds.²

Clinical signs of mitral valve stenosis are similar to those in older dogs with degenerative mitral valve disease: exercise intolerance, respiratory signs, and atrial arrhythmias. Mitral valve stenosis may cause a diastolic murmur at the left apex, where duration, not intensity, of the murmur may correlate with the degree of stenosis.³ This is consistent with a III/VI diastolic left apical murmur found in this dog. This patient also had a VI/VI systolic right apical murmur that may have obscured a systolic left apical murmur typically heard with mitral regurgitation.

Anatomic pathology of mitral valve stenosis shows the affected valvular leaflets were thickened, stiff, and nodular. The chordae tendineae may be thickened, short, and sometimes fused with hypertrophied papillary muscles.² A subvalvular ring or hypoplastic valves may be present.⁴ Mitral valve stenosis is characterized by a failure of the leaflets to properly diverge, resulting in pressure overload of the left atrium. Diastolic mitral regurgitation may be secondary to some cases of mitral stenosis. Systolic mitral regurgitation, or leakage of blood from the

left ventricle back into the left atrium due to poor connection between the mitral valve leaflets, can be caused by several pathological conditions. Myxomatous mitral valve degeneration (MMVD) is the most common cause of mitral regurgitation and secondary left atrial dilation in dogs and is characterized by stretching and thickening of the chordae tendineae and nodules on the thickened valvular leaflets. Mitral valvular bacterial endocarditis is extremely rare, but it is another acquired cause of left atrial dilation and is characterized by fibrin, a platelet or agglomerate forming vegetative lesions on the valves. Changes caused by endocarditis can vary from mild thickening of 1 or more valvular leaflets to a proliferating mass. The dog of the present report had thickened, dysplastic, and severely stenotic mitral valve leaflets. The valvular leaflets were tethered to dysplastic papillary muscles by thick and shortened chordae tendineae, similar to those seen in MMVD and endocarditis. This emphasizes the importance of echocardiography in revealing a stenotic mitral valve to distinguish between diseases that cause mitral valvular defects.

Diagnosis of mitral valve stenosis is not well described in veterinary medicine and is based on echocardiographic and catheterization findings associated with visualization of the valve apparatus structure and function, and demonstration of high-velocity diastolic regurgitation across the mitral valve. In most cases, survey thoracic radiography is an inaccurate method of diagnosing congenital heart defects in dogs, due to variation in thoracic conformation among patients, inconsistent positioning, variation of views from different phases of respiration, and lack of external cardiac changes that represent chamber enlargement.⁵ Survey radiography does not allow visualization of internal cardiac structures or assessment of function. The most consistent feature of mitral valve stenosis on thoracic radiography is severe enlargement of the left atrium. However, left atrial enlargement on thoracic radiography may occur in many conditions, and echocardiography is integral for the diagnosis of mitral valve stenosis. Pulmonary vein distension and evidence of left-sided CHF may also be present. Left ventricular size has been reported to be increased, normal, or decreased, so size is not considered as a component of mitral valve stenosis.³ Echocardiography is the most sensitive modality for diagnosing mitral valve stenosis. Two-dimensional echocardiography reveals malformation and narrowing of the mitral valve apparatus and dilation of the left atrium. Doming of the leaflets toward the left ventricle during diastole, incomplete diastolic separation of the mitral leaflets, and concordant diastolic motion of the septal and parietal mitral leaflets confirm mitral stenosis. Color flow Doppler echocardiography reveals high-velocity mitral inflows and diastolic mitral regurgitation, as described in this patient. Treatment most commonly consists of medically managing CHF. Patients with

only mild to moderate malformations may do well for several years. With the severity of this case, the prognosis was poor and likely end stage. To our knowledge, only 1 case of successful surgical valve replacement or reconstruction with cardiopulmonary bypass specific to mitral valve stenosis has been reported⁴ and is therefore not well studied; nonetheless, open-heart surgery may be an option for some patients.⁴

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