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Tricuspid valve dysplasia and Ebstein's anomaly in dogs: case report

[*Displasia da valva tricúspide e anomalia de Ebstein em cães: relato de caso*]

M.G. *Sousa*[1,4], D.G. *Gerardi*[1], R.O. *Alves*[2], A.A. *Camacho*[3]

[1]Aluno de pós-graduação - FCAV-UNESP
Via de Acesso Prof. Paulo Donato Castellane, s/n
14884-900 – Jaboticabal, SP
[2]Escola de Veterinária-UFG – Goiânia, GO
[3]Faculdade de Ciências Agrárias e Veterinárias-UNESP – Jaboticabal, SP
[4]Escola de Medicina Veterinária e Zootecnia-UFT – Araguaína, TO

ABSTRACT

Two cases of tricuspid valve dysplasia are reported. Dogs were presented for evaluation of weakness and ascites. In both cases, echocardiography disclosed tricuspid insufficiency and, in one of them, tricuspid leaflets also were displaced down into the right ventricle, substantiating Ebstein's anomaly. Medical therapy for congestive heart failure was initiated. One of the dogs suddenly died shortly after diagnosis was established. Although the other dog recovered much better initially, sudden death also occurred. Post-mortem examinations showed right atrioventricular enlargement, and thickened tricuspid leaflets. Clinical features, diagnostic methods and medical therapy are discussed in this paper.

Keywords: congenital heart disease, echocardiography, atrioventricular valve malformation

RESUMO

Dois casos de displasia da valva tricúspide são relatados neste trabalho. Os cães foram avaliados devido à fraqueza e presença de ascite. Em ambos os casos, o exame ecocardiográfico mostrou insuficiência tricúspide e, em um deles, a inserção dos folhetos da valva tricúspide encontrava-se deslocada para baixo do ventrículo direito, caracterizando a anomalia de Ebstein. A terapia medicamentosa para insuficiência cardíaca congestiva foi iniciada, mas um dos animais veio a óbito subitamente alguns dias após o diagnóstico. O outro cão, apesar de inicialmente ter apresentado melhora significativa do quadro clínico, apresentou morte súbita. A necropsia dos animais revelou dilatação atrioventricular direita e folhetos tricúspides espessados. As características clínicas, métodos de diagnóstico e terapia medicamentosa são discutidas neste artigo.

Palavras-chave: doença cardíaca congênita, ecocardiografia, malformação de valva atrioventricular

INTRODUCTION

Tricuspid valve dysplasia is a congenital malformation of the tricuspid leaflets, chordae tendineae, or papillary muscles, resulting in tricuspid insufficiency (Friedman, 1999). Although rare in dogs, the basal attachments of tricuspid valve may be ventrally displaced into the right ventricle ("right ventricular atrialization"), resulting in a further malformation named Ebstein's anomaly (Kittleson, 1998).

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*Autor para correspondência (*corresponding author*)
E-mail: marlos98@yahoo.com

It has been documented that dogs with tricuspid dysplasia commonly have absent or very short fused chordae tendineae and long thick septal leaflets adherent to the septum (Liu and Tilley, 1976). In human beings, however, several defects have been described to occur simultaneously, including focal or diffuse thickening of the valve leaflets, underdevelopment of chordae tendineae and papillary muscles, incomplete separation of valve components from the ventricular wall, and focal agenesia of valvular tissue (Becker et al., 1971). This wide spectrum of lesions is also speculated to occur in dogs (Sisson et al., 2000).

Most of the dogs affected with tricuspid dysplasia are large race purebred dogs. In a 10-year survey, the majority of dogs diagnosed with this abnormality by Kittleson (1998) were purebred and 25.8% were Labrador retrievers. To the authors' knowledge, no surveys have been done in Brazil to determine the incidence and prevalence of tricuspid valve dysplasia in dogs.

The diagnosis of tricuspid valve dysplasia is based on clinical signs and ancillary exams. Thoracic radiographs usually demonstrate cardiomegaly as a result of right atrium enlargement (Hoffmann et al., 2000). Kittleson (1998) reported that a massive right atrium enlargement in a young animal with right apical systolic murmur is generally pathognomonic for severe tricuspid dysplasia. Although not sensitive, high P waves, deep S waves in leads I, II, III, and a VF, and splintered QRS complexes may be observed on the electrocardiogram of dogs with tricuspid dysplasia (Kornreich and Moïse, 1997). Echocardiographic evaluation usually shows a markedly enlarged right atrium with right ventricular volume overload. Also, the left heart is often smaller than normal. Tricuspid leaflets may have little movement, appear to be very large, or be adhered to the interventricular septum or to the pappilary muscles (Hirschklau et al., 1977). Moreover, a turbulent regurgitant flow into the right atrium should be present during systole (Kittleson, 1998).

As documented in the literature elsewhere, tricuspid valve dysplasia is not a rare congenital heart defect in dogs. It is unresolved, however, how often this abnormality occurs in Brazil. The aim of this paper is to report two cases of tricuspid dysplasia in dogs. In one of them, the congenital disease also involved Ebstein's anomaly.

CASE REPORT

Two dogs were individually referred for evaluation of anorexia, weakness, and ascites. The first dog was a 5-month-old female English Cocker Spaniel presented with a 15-day history of abdominal swelling and exercise intolerance. On physical examination, it was lethargic, tachycardic, and had pale mucous membranes. Thoracic auscultation revealed a loud 4/6 holosystolic heart murmur, which was better heard over the right apex. Owing to severe ascites, abdominal palpation did not permit delimitating internal structures. The second dog was a 4-month-old male mongrel dog presented with an 8-day history of ascites, weakness, lethargy, and exercise intolerance. Pale mucous membranes were also observed on physical examination, as well as tachycardia, hypokinetic femoral pulses, and ascites. A 2/6 holosystolic murmur was also heard over the tricuspid area. Although ascites was present, the borders of enlarged liver and spleen were identified on abdominal palpation.

In both cases, tricuspid dysplasia was suspected due to age, development of severe ascites, exercise intolerance and the loud heart murmur over the tricuspid area. However, ancillary exams were requested to confirm the suspected congenital heart disease.

A complete blood count and serum biochemistry were performed. Interpretation of clinicopathologic data revealed mild anemia ($3.6x10^6$ erythrocytes/μl), thrombocytopenia ($95x10^3$ platelets/μl), hypoproteinemia (4.8g/dl), hypoalbuminemia (2.1g/dl), and increases in serum alanine aminotransferase (136.2U/l) and alkaline phosphatase (505.8U/l) in case 1. In case 2, anemia ($4.3x10^6$ erythrocytes/μl), mild leukocytosis (21,800 leucocytes/μl) and thrombocytopenia ($135x10^3$ platelets/μl) were observed. As seen in case 1, the biochemical profile showed increased serum alanine aminotransferase (665.0U/l) and alkaline phosphatase (232.0U/l). However, no changes in total protein and albumin existed.

Abdominal fluid was tapped to make the animal more comfortable and to provide fluid samples for physical and cytologic examination. A modified transudate was yielded in both cases. A bloody fluid with specific gravity of 1.017 and cellularity of 2,200 cells/µl was observed in Case 1, whereas in Case 2, a serous-bloody fluid with specific gravity of 1.015 and 25 cells/µl, mostly consisting of neutrophils.

Thoracic radiographs were taken, showing right atrial and ventricular enlargement in lateral and ventrodorsal views. In case 1, the electrocardiogram revealed sinus tachycardia (196bpm), increased duration of P wave (63ms) and QRS complex (60ms), right electrical axis shift, and suppression of R wave amplitude (0.57mV). In case 2, increased P wave duration, deep S waves, and right axis deviation were observed, as well as sinus tachycardia (180bpm). In both cases, a massive right heart enlargement was observed in two-dimensional echocardiograms. The right paraesternal short-axis view demonstrated a markedly enlarged right ventricle (Case 1: 2.38cm; Case 2: 2.57cm). Right ventricular overload volume pushed the ventricular septum towards the left ventricle and caused the septum to move paradoxically (Fig. 1-A). On right paraesternal long-axis view and left four-chamber apical view a massive right atrial enlargement was also observed, as well as a mildly hyperechoic tricuspid valve with little movement. A large tricuspid regurgitation (Case 1: 3.2m/s; Case 2: 2.7m/s) was present during systole on continuous-wave Doppler (Fig. 1-B). Only in Case 2, tricuspid leaflets were displaced ventrally into the right ventricle, resulting in a downward displacement of the right atrium (Fig. 1-D). This finding characterized Ebstein's anomaly in this case. Fractional shortening and ejection fraction were within the normal range in both animals.

Dogs were given furosemide (3mg/kg tid), enalapril (0.5mg/kg bid), and spironolactone (0.4mg/kg sid). The Cocker Spaniel recovered well one week after therapy was started. Ascites completely disappeared and the dog presented nearly normal activity. Although much better, the dog described in Case 2 suddenly died four days after the beginning of medications. The animal described in Case 1 was re-evaluated one week later for readjustment of diuretic dose (furosemide was reduced to 1.5 mg/kg sid). This animal would be submitted to four other follow-up examinations at 10-day intervals. However, 48 days after the first presentation, sudden death occurred without any signs of decompensation.
At postmortem examination, mild bloody ascites, right ventricular eccentric hypertrophy, dilatation of right atrium and vena cava, short chordae tendineae, and a notched, thickened tricuspid valve (Fig. 1-F) were seen in Case 1. Tricuspid leaflets of Case 2 were ventrally displaced into the right ventricle, thereby resulting in "right ventricular atrialization". Also, a markedly hypertrophied and dilated right atrium and ventricle were observed, as well as a shortened, notched, thickened valve.

DISCUSSION

Although not rare, several retrospective studies have shown that the occurrence of tricuspid dysplasia ranges from 7.0% to 7.5% of all congenital heart defects (Tidholm, 1997; Baumgartner and Glaus, 2003). However, prevalence and breed associations may exhibit geographical variations.

Tricuspid valve dysplasia has been reported in various dog breeds, including Old English Sheepdogs, Great Danes, Weimaraners, German Shepherds, and Irish Setters (Liu and Tilley, 1976). In Labrador Retrievers, this defect has even been determined to be a heritable disorder (Famula et al., 2002). The cases of tricuspid dysplasia reported here included an English Cocker Spaniel and a mongrel dog. To our knowledge, this disease was not previously diagnosed in English Cocker Spaniels. However, mixed-breed dogs have been reported to be affected by this defect (Kittleson, 1998).

Ebstein's anomaly has been reported to occur in dogs (Cave, 2001). However, some authors advocate that the diagnosis of this anomaly demands simultaneous intracardiac pressure and electrode studies to document ventricular complexes and right atrial pressure in the lowermost portion of the right atrium (Sisson et al., 2000). The diagnosis of Ebstein's anomaly in one of the cases described here (Case 2) was supported by the echocardiographic identification of the downward displacement of the tricuspid septal leaflet on the apical four-chamber view. This approach was recommended by Hirschklau et al. (1977) and Friedman (1999), who reported that invasive studies are rarely necessary to diagnose such cardiac anomaly.

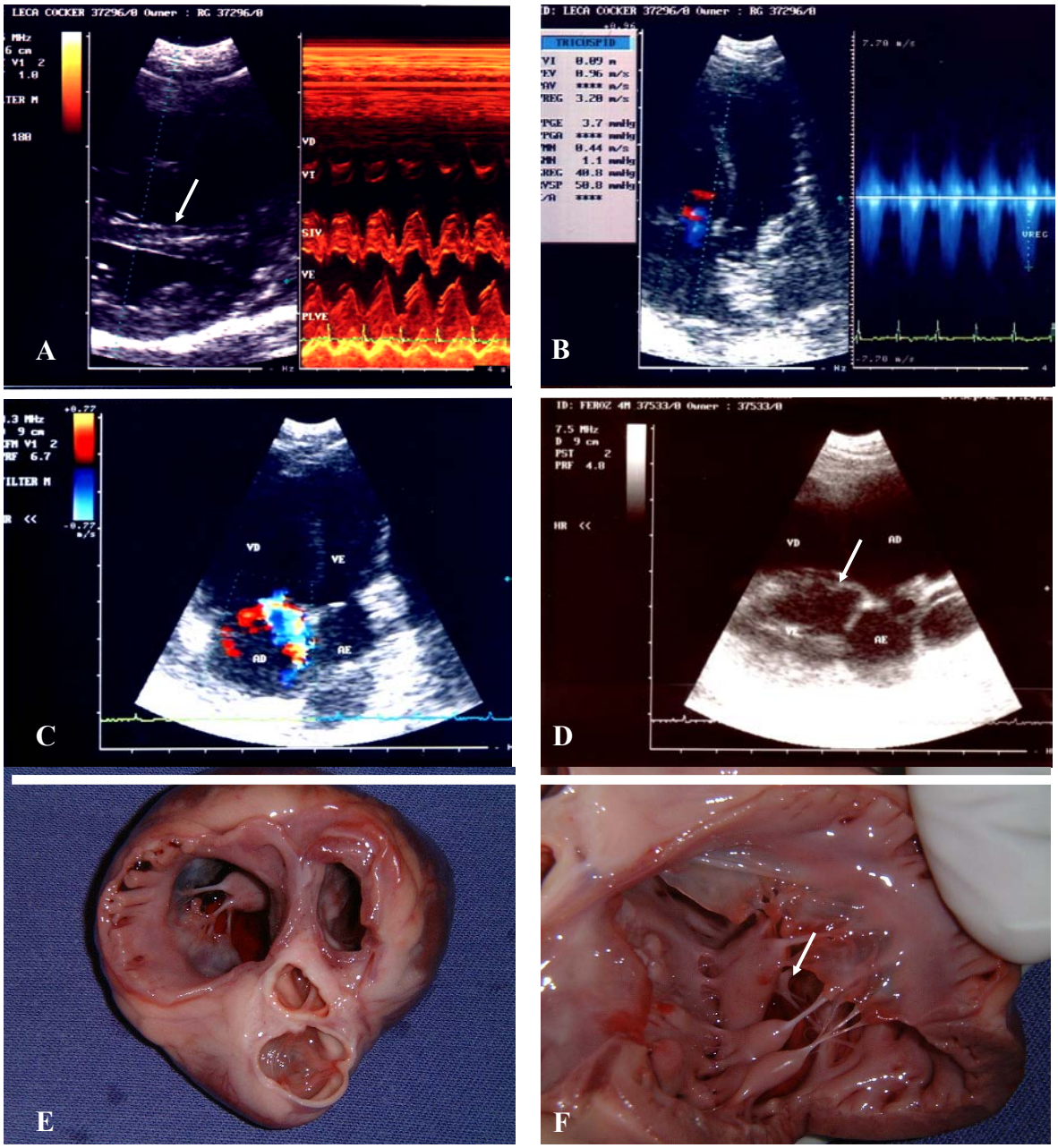

Figure 1. Representative findings in dogs with tricuspid valve dysplasia. (A) Two-dimensional and M-mode echocardiography shows a markedly enlarged right ventricle and paradoxal movement of the septum (arrow) in Case 1; (B) Continuous-wave Doppler in Case 1 shows a large turbulent flow into the right atrium; (C) Color Doppler was performed on Case 2 showing severe tricuspid regurgitation into right atrium. Also, severely enlarged right atrium and ventricle is seen on this apical four-chamber view; (D) The ventrally displaced tricuspid septal leaflet (arrow) is seen on this right paraesternal long-axis view, characterizing Ebstein's anomaly in Case 2; (E) and (F) Pathology specimens of Case 1 showing a marked dilatation of right heart and the abnormally attachment of papillary muscle to the notched thickened leaflet (arrow) with no intervening chordae tendineae.

Both cases were referred due to development of ascites, lethargy, and weakness. This is in agreement with several reports that described those signs related to right congestive heart failure and exercise intolerance (Sisson et al., 2000). Moreover, cardiac auscultation of both dogs revealed holosystolic heart murmurs over the right apex. This is considered to be the hallmark of tricuspid insufficiency (Chetboul et al., 2004) and, associated with those signs and age at presentation, led to the suspect of tricuspid valve dysplasia.

The diagnosis was based on clinical signs and ancillary exams. Right atrial and ventricular enlargement was observed on thoracic radiographs. As reported by Hoffmann et al. (2000), a markedly enlarged right atrium in young dogs with right apical systolic murmur is usually a consequence of tricuspid valve dysplasia. Besides right-axis deviation in both dogs and deep S waves in one of them, there were no further electrocardiographic evidences of right heart enlargement. Because electrocardiography is not so sensitive in detecting chamber enlargements, the absence of high P waves did not rule out right atrial enlargement (Kittleson, 1998). Although common, neither atrial arrhythmias nor splintered QRS complexes were seen in these dogs (Kornreich and Moïse, 1997). Sinus tachycardia, however, was recorded and is probably related to the activation of neurohumoral responses to overwhelm the leakage of blood into the right atrium and the decrease in pulmonary stroke volume (Kittleson, 1998). Investigators have demonstrated that the most striking finding on echocardiography is right atrial enlargement (Chetboul et al., 2004). In fact, both cases had severely enlarged right atrium and ventricle. On right paresternal short-axis views, right ventricular volume overload resulted in paradoxal movement of the septum and an abnormally small left ventricle (Friedman, 1999). Alterations of the tricuspid valve seen in these cases were similar to others reported in the literature that included thickened and notched leaflets with little movement as the most commonly observed abnormalities (Cave, 2001). The diagnosis of tricuspid dysplasia was confirmed by Doppler interrogation of tricuspid valve, in which a large turbulent jet into the right atrium during ventricular systole was observed (Friedman, 1999).

Although some animals might tolerate this defect for a long time (Hoffmann et al., 2000), medical therapy was instituted because right heart failure had already developed in both dogs. Therefore, the dogs were put on furosemide and enalapril to overcome compensatory mechanisms. Due to right heart failure and probably increased plasma aldosterone concentration, spironolactone was also given to competitively inhibit the action of aldosterone on distal tubular cells, thereby increasing fractional excretion of sodium and water.

CONCLUSION

This report reinforces the necessity of a careful auscultation in young dogs with ascites and a history of weakness and exercise intolerance. Tricuspid malformation should be suspected whenever a heart murmur is heard over the right apex and signs of right-sided heart failure develop.

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