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Case Report

Ebstein anomaly in a dog

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Abstract
Ebstein's anomaly is a rare congenital cardiac abnormality characterized by an abnormal positioning of the cusps of the tricuspid atrioventricular valve. This study aimed to report the clinical and pathological aspects of the condition in an adult medium-sized mixed-breed dog, treated at a veterinary hospital in Brazil, through clinical evaluation, echocardiography, and post-mortem examination. This case highlights the importance of imaging exams for accurate diagnosis and the formulation of therapeutic strategies aimed at patient survival. To our knowledge, this is the first report of this congenital cardiac anomaly in a medium-sized mixed-breed dog.

Keywords: cardiology, congenital anomaly, veterinary medicine.

Introduction

Congenital heart anomalies are defects present from birth and often result in early death. However, in some cases, these abnormalities can be asymptomatic and remain undiagnosed until adulthood. Consequently, the percentage of adult dogs with subclinical congenital heart diseases may be significant (2). An example of a congenital heart condition is a right atrioventricular valve or tricuspid dysplasia, which accounts for approximately 2% to 8% of cardiac diseases in dogs (9). The breeds most affected include Labrador Retrievers, Great Danes, Bull Terriers, German Shepherds, and Dalmatians (12). Macroscopically, this condition may present as: focal or diffuse thickening of the valve leaflets, typically more pronounced at the leaflet edges; absence of valve leaflets, short or malformed chordae tendineae, and/or papillary muscles; direct fusion of the leaflets to the ventricular wall. These alterations may occur individually or concurrently (9,12).

Among cases of tricuspid dysplasia, Ebstein's anomaly stands out as a congenital defect first described in 1866 by Wilhelm Ebstein in a 19-year-old human patient (5). In dogs and cats, it is considered rare, accounting for only 2.9% of all congenital heart diseases in dogs (4). The anomaly occurs when the cusps of the right atrioventricular valve (tricuspid) are displaced more ventrally compared to the left atrioventricular valve (bicuspid or mitral), resulting in a functional division of the right ventricle into a "functional" portion and an "atrialized" portion. This change leads to right atrial enlargement and tricuspid regurgitation, which can, in some cases, cause signs of right-sided congestive heart failure (5,8).

The occurrence of Ebstein's anomaly is reported in an adult dog, highlighting the importance of clinical methodology combined with specific cardiological examinations, which are essential for a more accurate diagnosis.

Case description

During the clinical examination of a 5-year-old, 8 kg mixed-breed male dog at the Veterinary Hospital of the

Federal University of Paraná – Palotina Campus (UFPR – Palotina Campus), Brazil, cyanotic mucous membranes were observed. The heart rate was 156 bpm, and the systolic blood pressure was 110 mmHg. The dog had a history of a wound resembling a spider bite on the right pelvic limb, a corneal ulcer ten days after the suspected bite, diarrhea, and a neurological condition characterized by ataxia and distemper approximately two months prior. Ultrasonography revealed pleural effusion and ascites. A transthoracic echocardiogram was performed using right and left parasternal windows in standard two-dimensional, M-mode, and Doppler modes with a Philips Affiniti 50 ultrasound machine (USA). The examination showed that the tricuspid valve had an elongated parietal cusp relative to the septal cusp, with improper attachment to the papillary muscles, and the septal cusp displayed reduced mobility (Figure 1A). These findings resulted in moderate tricuspid insufficiency, leading to significant right atrioventricular dilation, moderate thinning of the interventricular septum, right ventricular systolic dysfunction, and signs of right-sided congestive heart failure, as observed through echodoppler (Figures 1B, C, D). The abnormalities detected in the tricuspid valve apparatus, combined with the associated hemodynamic complications, were suggestive of tricuspid valve dysplasia. The dog was hospitalized at the Veterinary Hospital of UFPR – Palotina Campus, where it underwent abdominocentesis and treatment with furosemide (2 mg/kg TID), pimobendan (0.25 mg/kg BID), and spironolactone (2 mg/kg BID). However, after seven days, the dog passed away and was sent for necropsy at the Veterinary Pathology Laboratory of the same institution.

Macroscopic analysis revealed that the animal had a low body condition score (2/5), the oral and ocular mucous membranes were moderately cyanotic, in the right pelvic limb, a focally extensive area of discontinuity involving the skin, subcutaneous tissue, and musculature was noted, extending from the tarsus to the metatarsus, with bone

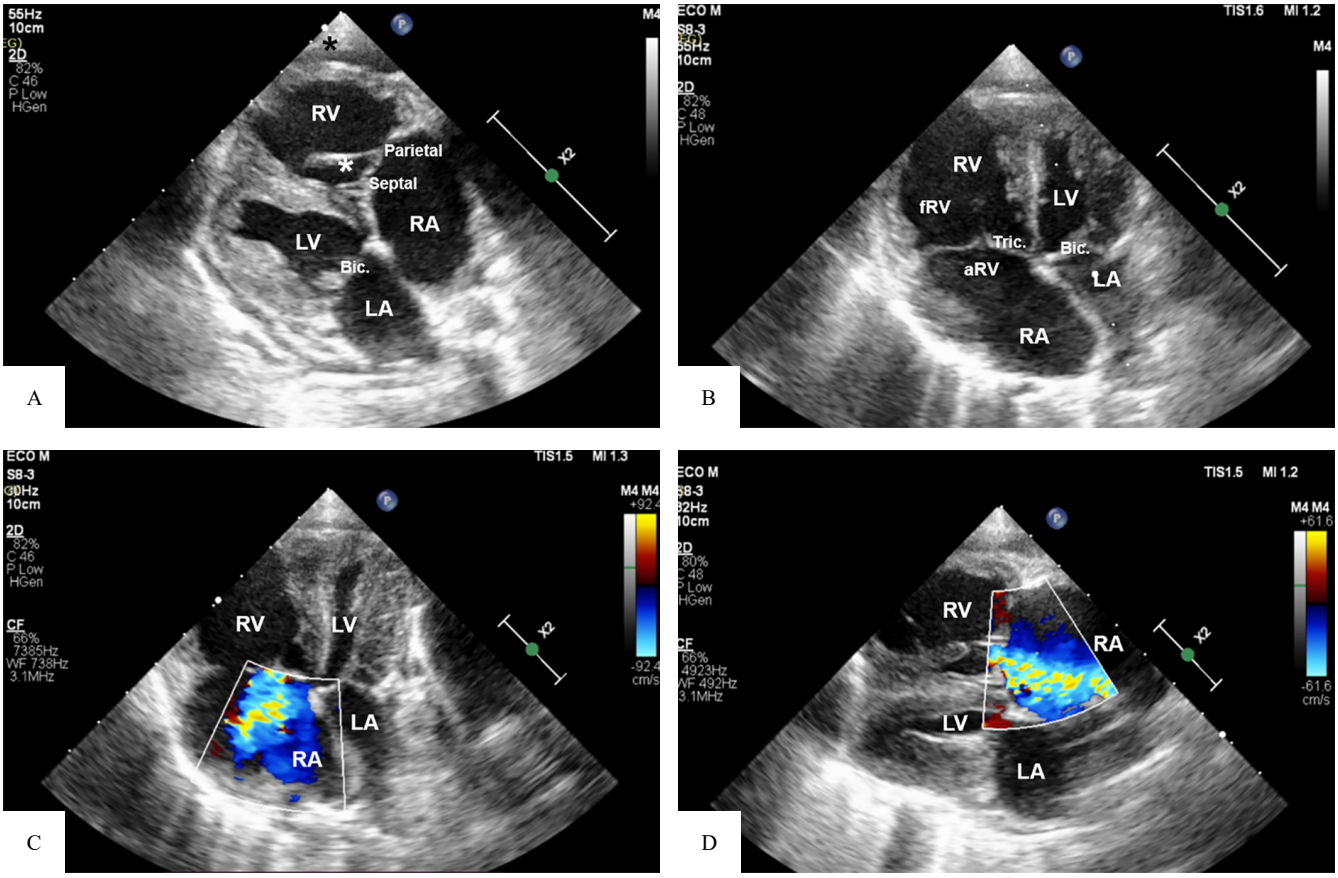

Figure 1. Echocardiographic images from different echocardiographic views. A. Longitudinal four-chamber view from the right parasternal window showing right atrioventricular remodeling with an elongated parietal cusp of the right atrioventricular valve (tricuspid) compared to the septal cusp. B. Apical four-chamber view from the left parasternal window. C. Echocardiographic image using color Doppler in the apical four-chamber view from the left parasternal window, demonstrating marked insufficiency of the right atrioventricular valve (tricuspid). D. Echocardiographic image using color Doppler in the longitudinal four-chamber view from the left parasternal window. RV - Right ventricle; RA - Right atrium; LV - Left ventricle; LA - Left atrium; White asterisk - Misattachment of the parietal cusp to the papillary muscles; Tric. - Tricuspid atrioventricular valve; Bic. - Bicuspid atrioventricular valve; aRV - Atrialized right ventricle; fRV - Functional right ventricle; Black asterisk - Small amount of pericardial effusion.

exposure and wound edges that were reddish proximally and gray to black distally. In the left eyeball, a mild focal area of corneal discontinuity was observed, with protrusion of material (descemetocele). There was hydrothorax (165 mL) and hydroperitoneum (5 mL), hepatomegaly and hepatic congestion, moderate splenomegaly, interstitial nephritis, hyperemic lungs with edema and congestion, as well as acute gastritis. The blood vessels of the leptomeninges were hyperemic. Upon opening the heart, the right atrium and auricle were dilated, with thin and distended pectinate muscles. Signs of blood regurgitation ("jet lesion") from the right ventricle to the right atrial endocardium were observed near the openings of the venae cavae and the fossa ovalis (Figures 2A and B), characterized by mild endocardial thickening and roughness. The fibrous ring supporting the three cusps (parietal, septal, and angular) of the tricuspid valve was positioned ventrally relative to the bicuspid valve (Figures 2A and B). The cusps displayed thickened edges, resembling a "hockey stick" appearance (Figure 2C).

The parietal cusp of the tricuspid atrioventricular valve had three distinct papillary muscles, giving rise to five primary chordae tendineae with an average length of 7.3 mm (± 1.9 mm) (Figure 2A). One of these muscles was located basally, near the interventricular septum, and sent out one primary chorda tendinea and four secondary chordae. The second papillary muscle, situated near the trabeculae carneae, had a typical appearance and emitted three primary chordae tendineae. One of these primary chordae gave rise to three secondary chordae, each subdivided into two tertiary chordae. The other two primary chordae each emitted two secondary chordae. Lastly, the third papillary muscle of the parietal cusp had two heads. One head emitted one primary chorda tendinea and four secondary chordae attached to the parietal cusp (Figure 2C). The other head emitted one primary chorda tendinea with two secondary chordae and a longer primary chorda attached to the base of the angular cusp (Figure 2C).

Another papillary muscle was observed in the angular cusp, emitting four primary chordae tendineae. This muscular projection was firmly attached and located at the base of the interventricular septal wall. Two of these chordae had secondary chordae with a circular nodule of an average diameter of 1.3 mm (± 0.2 mm) (Figure 2C). In total, there were six primary chordae tendineae with an average length of 9.4 mm (± 3.6 mm). In the septal cusp, seven small basal papillary muscles were identified, collectively emitting 13 primary chordae tendineae with an average length of 3.5 mm (± 1.0 mm) (Figure 2B).

Discussion

The etiology of Ebstein's anomaly, a type of tricuspid valve dysplasia, is still not fully elucidated. However, this condition is known to be associated with an autosomal dominant inheritance linked to chromosome 9, which is more prevalent in Labrador Retriever dogs (10). The absence of cardiac signs in patients usually occurs in about 59% of cases of subclinical dogs diagnosed incidentally, with suspicion raised during cardiac auscultation (5). Thus, many animals affected by the anomaly have remained undiagnosed (10),

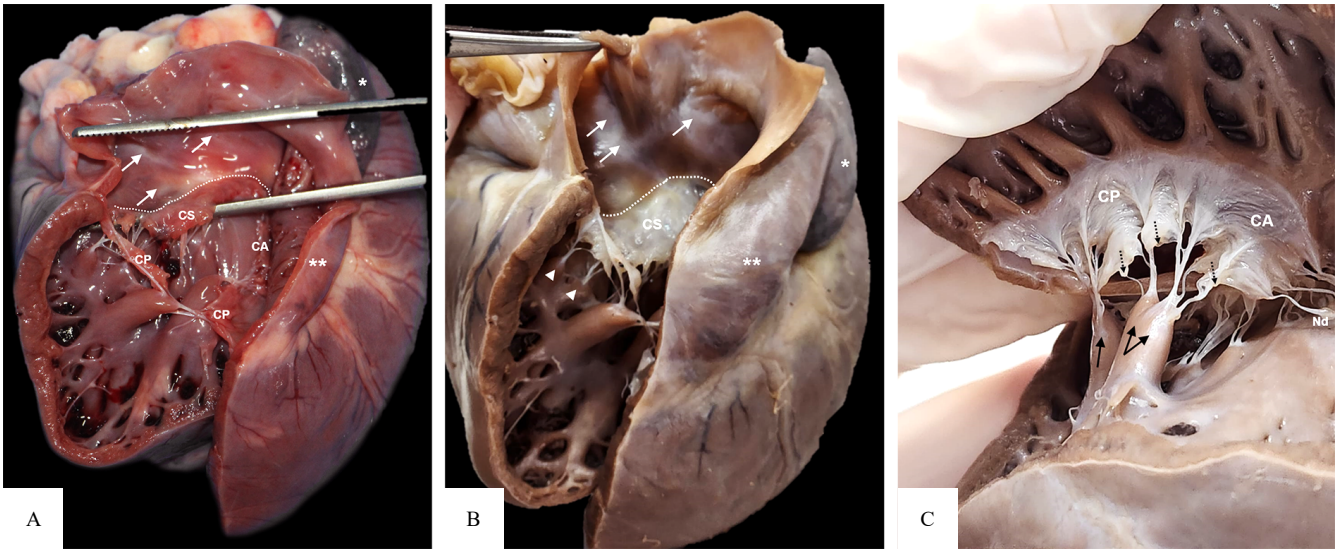

Figure 2. Opening of the right atrium and ventricle. A. Heart dissected in its natural state. B. Heart preserved in 10% formaldehyde. C. Image of the interior of the dissected right ventricle. White arrows- points of blood regurgitation on the endocardium ("jet lesion") of the right atrium; One asterisk- dilated right auricle; Two asterisks- dilated right atrium; Dashed line- level of the right atrioventricular valve; CS- septal cusp; CP- parietal cusp; CA- angular cusp. Arrowhead- basal papillary muscle at the interventricular septum. Dashed arrow- thickened tips of the cusps resembling a "hockey stick." Black arrow- typical papillary muscle; Double black arrow- papillary muscle with two heads; Nd- nodule on the chordae tendineae.

as occurred in the present clinical case, in which the cardiac condition was identified only through complementary exams.

On physical examination, signs consistent with right-sided congestive heart failure were observed, as seen in all 32 dogs analyzed in another study in which such murmurs were classified as grade 4 or grade 5 (5). Thus, from the clinical picture, differential diagnoses should include tricuspid dysplasia, tricuspid valve endocarditis, tricuspid valve prolapse, degenerative valve disease, right ventricular cardiomyopathy, pulmonary valve insufficiency (8), as well as mitral valve myxomatous disease and dilated cardiomyopathy (10). Identifying individuals affected by this congenital condition is essential due to the reproductive implications it may entail (10).

Echocardiography, considered the confirmatory technique for various congenital anomalies (8), enabled confirmation of the diagnosis of tricuspid valve dysplasia and suspicion of Ebstein's anomaly. The definitive diagnosis of this anomaly was concluded through the association of clinical signs, imaging exams, and macroscopic findings with evidence of a more ventral insertion of the tricuspid valve cusps compared to the level of the bicuspid valve, which is one of the most important confirmatory criteria for congenital disease (5).

The prognosis of this condition is quite variable due to the possibility of other concomitant cardiac or systemic alterations (5). Although this anomaly is more common in large-breed dogs, there are reports of its occurrence in smaller breeds, such as the French Bulldog (8) and the Beagle (11). However, this is the first report of Ebstein's anomaly in a medium-sized mixed-breed dog in the veterinary medical literature.

The present report is in accordance with the observation that most dogs affected by this cardiac condition can live for many years and that death does not necessarily occur as a result of heart disease. Although, in the adult phase, the anomaly is frequently associated with severe morphological alterations, a good medium-term survival is observed (5). The presence of thickened cusps tips with an appearance resembling a "hockey stick" was similar to that described for the condition by Sudunagunta et al. (2020), in contrast, the alterations in several cusps of the atrioventricular valve differ from what is normally pointed out in the scientific descriptions that highlight that the septal cusp is the most affected (3) as well as the size of the animal attended, considered as medium-sized.

Morphologically, right atrioventricular valve dysplasia can be erroneously caused as mitral valve myxomatous disease or confused with dilated cardiomyopathy, especially in patients who present marked cardiac remodeling and systolic dysfunction. However, the correct identification of congenitally affected individuals is crucial, as it can have significant implications for reproduction (10). This concern becomes relevant in the present case, since the animal was 5 years old and could have already procreated. The arching of the right atrioventricular valve cusps was presented in the documented case; however, this alteration can also occur in situations of dilated cardiomyopathy. This change can result, for example, from a remodeling of the left ventricle, which leads to a rounded geometry of this chamber, to the abnormal alignment of the papillary muscles and chordae tendineae, to the dilation of the mitral annulus, to the poor coaptation of the cusps, and to the angled appearance (13). Therefore, patients with mitral dysplasia may present primary abnormalities of the valvular apparatus, including the tension of the cusps, which can further aggravate the arching (6). This situation is very similar to that observed in the dog reported herein when analyzing the heart macroscopically during its necropsy.

In medicine, the treatment of Ebstein's anomaly involves surgical intervention, together with the clinical management of the symptoms of right-sided heart failure (7). On the other hand, in veterinary medicine, surgical procedures are not frequently performed due to technical limitations (8). However, it is important to emphasize that there are reports of cases with positive evolution in dogs with mitral valve dysplasia that underwent surgical repair or valve replacement. Thus, with advances in the safety and availability of these interventions, together with early detection and accurate diagnosis of cardiac alterations, many dogs may present more detailed clinical results and longer survival (2, 14, 15).

In conclusion, to our knowledge, this is the first report of Ebstein's anomaly in a medium-sized mixed-breed dog. The right atrium and auricle were dilated, with thin and distended pectinate muscles. There were signs of blood regurgitation from the right ventricle to the right atrial endocardium near the openings of the venae cavae and the fossa ovalis. Additionally, the fibrous ring supporting the three cusps of the tricuspid valve was positioned ventrally in relation to the bicuspid valve. This case highlights the importance of imaging exams for accurate diagnosis and the formulation of therapeutic strategies aimed at patient survival. These findings emphasize the need for increased awareness of congenital heart anomalies in general veterinary practice and suggest that future research should focus on the prevalence, diagnostic criteria, and long-term outcomes of Ebstein's anomaly in dogs.

Conflict of Interest

The authors declare no competing interests.

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