



Case report

Arrhythmogenic Right Ventricular Cardiomyopathy in Two Dogs

Letícia Batelli de Oliveira¹, Gustavo Henrique Lima de Siqueira¹, Luciana Sonne², Fabiano José Ferreira de Sant'Ana¹, Janildo Ludolf Reis Junior^{1*}

¹Laboratório de Patologia Veterinária (LPV), Universidade de Brasília (UnB), Brasília, Distrito Federal, Brazil.

²Setor de Patologia Veterinária, Universidade Federal do Rio Grande do Sul (UFRGS), Porto Alegre, Rio Grande do Sul, Brazil

* Corresponding Author: LPV, UnB, 70910-900, Brasília, DF, Brazil. E-mail: janildo@unb.br

Submitted February 06th 2014, Accepted March 14th 2014

Abstract

Arrhythmogenic right ventricle cardiomyopathy (ARVC) is a rare primary heart condition recognized in dogs. It is an inherited disease more commonly described in Boxer dogs. Clinically, ARVC is characterized by ventricular tachyarrhythmias and histopathologically by fibrofatty replacement, mostly on the right ventricle. This report describes the gross and histopathological findings from a female adult Boxer and from a male adult mixed breed dog, presenting signs of heart failure and marked infiltration of adipose tissue replacing cardiomyocytes, predominantly in the epicardium and myocardium of the right ventricle.

Key Words: cardiomyopathy, ARVC, right ventricle, fibrofatty replacement.

Introduction

Arrhythmogenic right ventricle cardiomyopathy (ARVC) is a rare primary heart condition recognized in dogs and humans characterized by partial or total fatty or fibrofatty replacement of right ventricular and atrial wall and, less frequently, of the interventricular septum and left ventricular wall. It is believed ARVC is transmitted by an autosomal dominant trait in Boxer dogs. However, it has also been described in other breeds, such as Labrador Retriever and Dachshund. It is more commonly reported in animals ranging from 6 and 10 years old. No predilection for sex has been described (3, 5, 6).

The most commonly reported clinical findings of ARVC is arrhythmia, specifically ventricular premature complexes (VPCs), and those changes due to congestive right heart failure. Sudden death without clinical signs has also been reported (4). Family history of the disease, ventricular tachyarrhythmia, history of syncope or exercise intolerance are important clinical data to guide the diagnosis. This latter is achieved by the typical gross and histopathological findings of right ventricular and atrial wall of ARVC (5).

Grossly, animals with ARVC may present dilation and pallor of the right ventricle (RV) chamber. No changes in heart weight or wall thickness of the RV or left ventricle (LV) are observed. Microscopically, there are two presentations of ARVC. The first is characterized by diffuse adipose tissue infiltration in the RV myocardium, extending from the epicardium to the endocardium. The second is the fibrofatty form, which consists of diffuse to multifocal areas of adipose tissue infiltration associated with proliferation of fibrous connective tissue. In either forms LV or right and left atria may be mildly affected. Multifocal infiltration of lymphocytes with cardiomyocyte individual necrosis of the RV free wall is rarely described in ARVC animals (2).

This report describes the clinical and pathological findings of two dogs, a female Boxer and a male mixed breed dog with ARVC.

Case report

Two dogs were submitted to the Laboratory of Veterinary Pathology (LVP) at the University of Brasília (UnB), Brazil, for necropsy. The first dog (dog 1) was a 13-year-old female Boxer with history of anorexia,

abdominal distension and tachypnea for four days. The radiographic exam revealed pleural effusion and ascites, which drained 2.5 liters from the abdominal cavity. During examination the dog had a cardiopulmonary arrest with unsuccessful resuscitation procedure. The second dog (dog 2) was a 15-year-old male, Labrador Retriever mixed breed dog. This dog was abandoned at the Center for Zoonosis Control (CZC) with history of weakness. The dog was euthanized at the CCZ prior submission to necropsy. Both dogs had samples from the heart and other tissues collected, fixed in buffered 10% formalin and submitted for routine histological processing. Sections from multiple organs were stained with hematoxylin and eosin (HE) and Gomori's trichrome (heart sections).

Dog 1 had pale external mucosae. The heart was enlarged with areas of petechiae and suffusions in both ventricles. The RV was markedly pale (Figure 1).

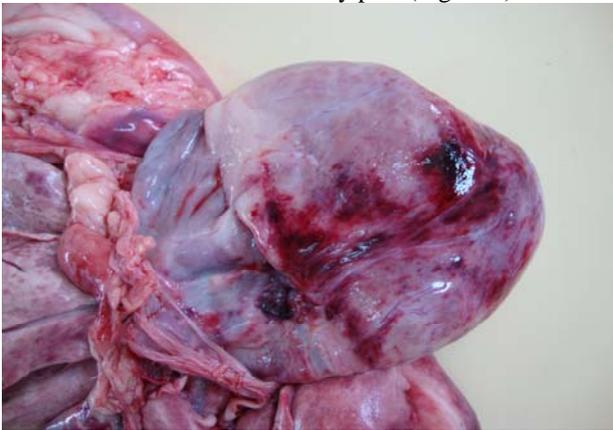


Figure 1. Heart, Dog 1. Dilated and pale heart with hemorrhages in the right and left ventricles. 13-year-old, female Boxer.

The liver was severely congested (Figure 2) and the lungs had edema, congestion and suffusions. Microscopically, approximately 65% of the RV epicardium and myocardium were expanded and replaced by adipose tissue (Figure 3). The LV was mildly affected, with less than 20% of fatty infiltration. Trichrome's staining of the RV myocardium revealed multifocal and mild proliferation of fibrous connective tissue (Figure 4). The lungs had congestion, edema and hemosiderin-laden macrophages (heart-failure cells). The liver had centrilobular congestion with moderate hepatocyte degeneration and necrosis.

Dog 2 had rounded heart with dilated LV and marked multifocal to coalescing pallor areas in the RV and LV. There was moderate pulmonary edema and centrilobular congestion of the liver. Microscopically, extending from the epicardium to the myocardium there was moderate amount of adipose tissue compressing and replacing approximately 50% of the RV muscle fibers (Figure 5 and 6). Fibrous tissue proliferation was not observed by Gomori's trichrome staining. There was not fatty or fibrofatty infiltration in LV muscle fibers in this animal. Vacuolar degeneration, congestion, fibrosis and hemosiderin-laden macrophages were in centrilobular

areas of the liver. Moderate congestion and mild edema were observed in the lungs, with no heart-failure cells.



Figure 2. Liver, Dog 1. Marked chronic hepatic congestion with round profile of the lobes.

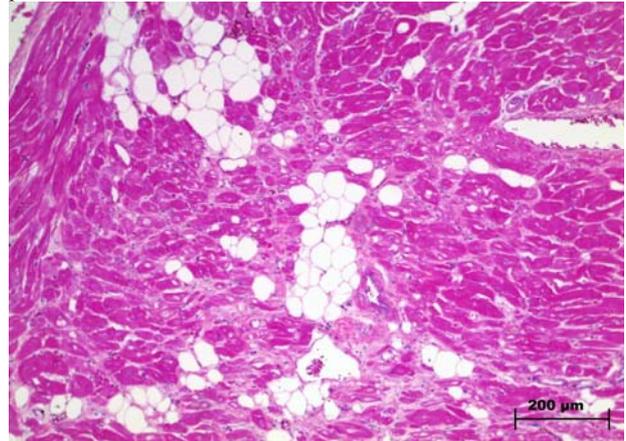


Figure 3. Heart, right ventricle, Dog 1. Adipose tissue infiltration and mild fibrous connective tissue proliferation. HE.

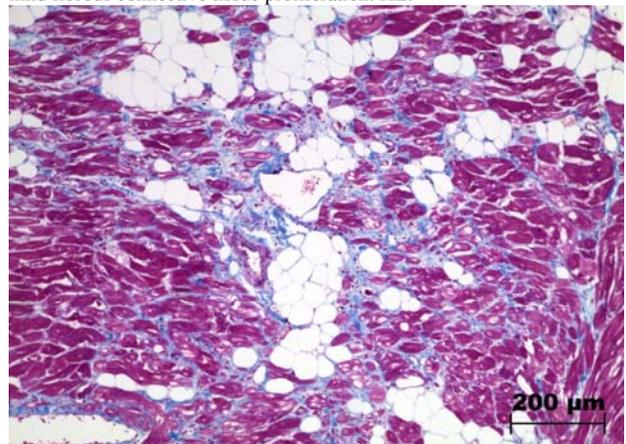


Figure 4. Heart, right ventricle, Dog 1. Same area from figure 3 evidencing fibrous connective tissue infiltration in blue. Gomori's trichrome.

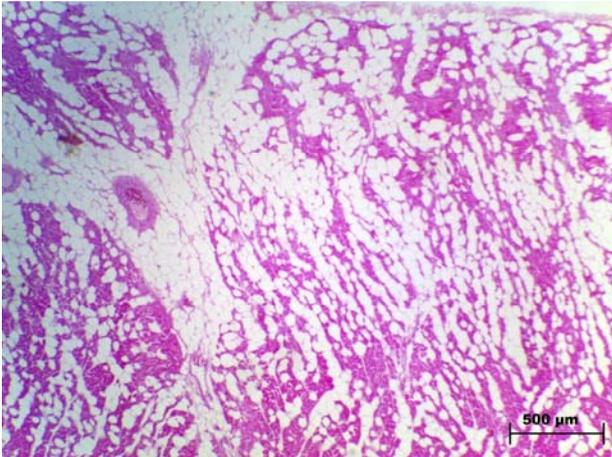


Figure 5. Heart, right ventricle, Dog 2. Severe adipose tissue infiltration disrupting epicardium and myocardium. HE.

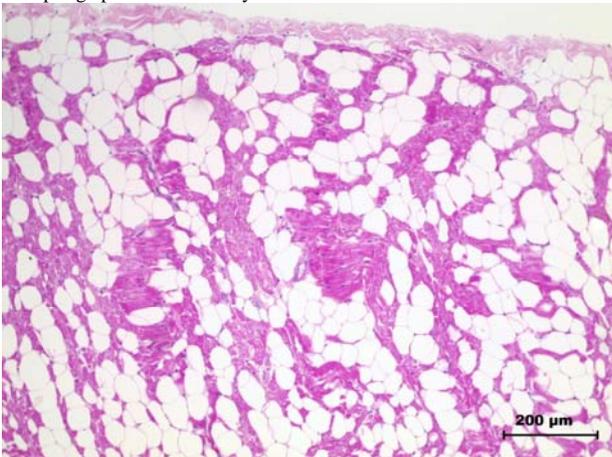


Figure 6. Heart, right ventricle, Dog 2. Higher magnification of Figure 5. HE

Discussion

The gross and histopathological changes from both dogs described in this report are typical of ARVC, which is an uncommonly described inherited disease mostly reported in Boxer dogs (1, 2, 3). It has also been described in Labrador Retriever and Dachshund. In the present study, affected dogs were a Boxer dog and a Labrador Retriever mixed breed dog. Therefore, one may infer that the ARVC presented in both dogs from this report is inherited. The most common clinical signs of ARVC are ventricular arrhythmias, syncope, heart failure or sudden death (2, 3). Dog 1, presented both clinical history and gross and microscopic changes of congestive heart-failure, with pleural effusion, congestion, edema and heart-failure cells in the lungs, and chronic passive congestion in the liver. Therefore, these findings are consistent with a chronic and progressive ARVC leading to bilateral (right and left) heart failure. Although, dog 2 had poor clinical history, since it was abandoned at the Center for Zoonosis Control (CZC), gross and microscopic changes were typical of ARVC.

The ARVC findings of adipose tissue infiltration separating and compressing RV cardiomyocytes, from the epicardium to the myocardium, as presented here in both dogs, have been previously reported (2). Additionally, dog 1 showed infiltration by fibrofatty tissue in the RV, similar to other descriptions (2, 3). The mild fatty infiltration of the LV in this same animal has been previously described in Boxer dogs (3, 5), and since it consisted of mild infiltration, left-sided heart failure is most likely a consequence of the severe RV lesion. Both forms of ARVC were observed in this report, with the fibrofatty form in dog 1 and the fatty form in dog 2. It is not known if there is any difference between the incidences of these two forms of ARVC in dogs.

Differential diagnosis was toxic degeneration or necrosis, infectious myocarditis (e.g. caused by canine parvovirus) and lymphosarcoma. The gross and histopathological findings associated with the clinical signs were crucial to support the definitive diagnosis of ARVC.

Acknowledgements

The authors are thankful to the Center for Zoonosis Control (CZC) for submitting dog 2 for necropsy.

References

1. AFIP 2009-2010. Case II. 5th Wednesday Slide Conference, Conference, Armed Forces Institute of Pathology. Available in http://www.askjpc.org/wsc/wsc_showcase2.php?id=231
2. BASSO C., FOX PR., MEURS KM., JEFFREY AT., SPIER AW., CALABRESE F., MARON BJ., THIENE G. Arrhythmogenic right ventricular cardiomyopathy causing sudden cardiac death in Boxer dog: a new animal model of human disease. *Circulation*, 2004, 109, 1180-5.
3. BOUJON CE., AMBERGER CN. Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) in Boxer. *J. Vet. Cardiol.*, 2003, 5, 35-41.
4. HARPSTER NK. Boxer cardiomyopathy: a review of the long-term benefits of antiarrhythmic therapy. *Vet. Clin. North Am. Small Anim. Pract.*, 1991, 21, 989-1004.
5. MEURS KM. Boxer dog cardiomyopathy: an update. *Vet. Clin. North Am. Small Anim. Pract.*, 2004, 34, 1235-44.
6. O'GRADY MR., O'SULLIVAN ML. *Clinical cardiology concepts for the dog and cat*. Available in <http://www.vetgo.com>