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Case report

DOUBLE-CHAMBERED RIGHT VENTRICLE, MEMBRANOUS VENTRICULAR SEPTAL DEFECT, AND PULMONIC STENOSIS IN A BRITISH SHORTHAIR

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SUMMARY

A five-month-old British shorthair kitten previously diagnosed with hypertrophic cardiomyopathy and feline infectious peritonitis was presented for cardiology examination. The findings of right cardiomegaly on chest radiography and electrocardiogram did not support a diagnosis of hypertrophic cardiomyopathy. On echocardiogram, muscular bands dividing the right ventricular chamber into two sub- chambers, narrowing of the pulmonary valve annulus, and a left-to-right shunt through amembranous ventricular septal defect were seen. Therefore, a diagnosis of double-chambered right ventricle, membranous ventricular septal defect, and pulmonic stenosis was made. Despite medications, the kitten eventually developed Eisenmenger syndromeand hind-limb paralysis, and was euthanized. Post-mortem examination for the heart and liver confirmed the above diagnoses.

Keywords: congenital heart disease, double-chambered right ventricle, Eisenmenger syndrome, ventricular septal defect

INTRODUCTION

Double-chambered right ventricle (DCRV) is a rare congenital heart disease in which the anomalous fibromuscular bundles divide the right ventricle into a proximal high-pressure and a distal normal pressure chamber (Brockman *et al*., 2009). In cats, it has been reported by MacLean *et al*. (2002), Koffas *et al*. (2007), Brockman *et al*. (2009),and Mizuno *et al*. (2010). By contrast, literature on concomitant DCRV and ventricular septal defect in cats is scarce. Since there is only one report which reviewed the ante- and post-mortem findings in a cat with concurrent double caudal vena cava (Dirven *et al*., 2010), the aim of the present case report is to elucidate clinical presentation and disease progression, and post-mortem findings in a British shorthair kitten with concomitant DCRV, pulmonic stenosis, and ventricular septal defect.

CASE REPORT

A five-month-old British shorthair weighing 0.85 kg was presented due to an increased breathing effort. It had a history of ascites and was previously diagnosed with hypertrophic cardiomyopathy (HCM) via echocardiography and feline infectious peritonitis (FIP) by the preceding veterinarians. According to the owner, the diagnosis of FIP was based on the positive ImmunoComb® feline coronavirus antibody test (Biogal Galed Labs, Israel) although the Rivalta's test was negative and the plasma globulin level was normal. Since then, the cat was treated with clopidrogel (Plavix® 75 mg, 18.75 mg/cat, PO, SID), and benazepril (Fortekor™ 5 mg, 0.25 mg/kg, PO, SID) and diltiazem (Herbesser® 30 mg, 9.4 mg/kg, PO, SID). Besides, it had been receiving a course of antiviral treatment (GS-441524) for the treatment of FIP. From the above history, further diagnostic workups were aimed at investigating the cause of ascites and confirming the heart disease. The top differentials for the ascites in the kitten included FIP, right heart disease, and hypoproteinemia.

Upon physical examination, the cat was bright and alert with flaccid abdomen. There was no neurologic deficit or jaundice observed. Systolic left and right murmurs grade IV/VI were auscultated. Chest radiography revealed right cardiomegaly (vertebral heart score of 11), apex elevation, and dilated caudal vena cava (Figure 1a, 1b). Besides, electrocardiogram showed sinus rhythm with a heart rate of 188 bpm, in addition to right ventricular enlargement pattern indicated by deep S waves in leads I and II (Figure 1c). The above findings were untypical for HCM, and an echocardiogram was pursed under sedation with pethidine (CCM, 3 mg/kg, IM) due to the lack of patient cooperation. On right parasternal long-axis view (Figure 2a, 2b) of echocardiography, the left atrium (LA) was dilated [LA: aorta (Ao): 2.5; reference range < 1.6] whereas the interventricular septal (4.4-4.9 mm) and left ventricular (LV) free walls (4.6 mm) during diastole were mildly thickened. Besides, the right atrium (RA) was severely dilated and the right ventricular (RV) wall showed marked concentric hypertrophy (7.0 mm). Further, a hyperechoic band which extended from the RV apex to just distal to the right ventricular outflow tract (RVOT) was also seen. On color Doppler (Figure 2d), there was a left-to-right shunt through membranous ventricular defect, and presence of tricuspid regurgitation and turbulence at the RVOT. The velocity across the stenotic RVOT was 3.27 m/s, giving rise to a pressure gradient of 42.9 mmHg. Moreover, ultrasonography of the liver showed dilated hepatic veins.

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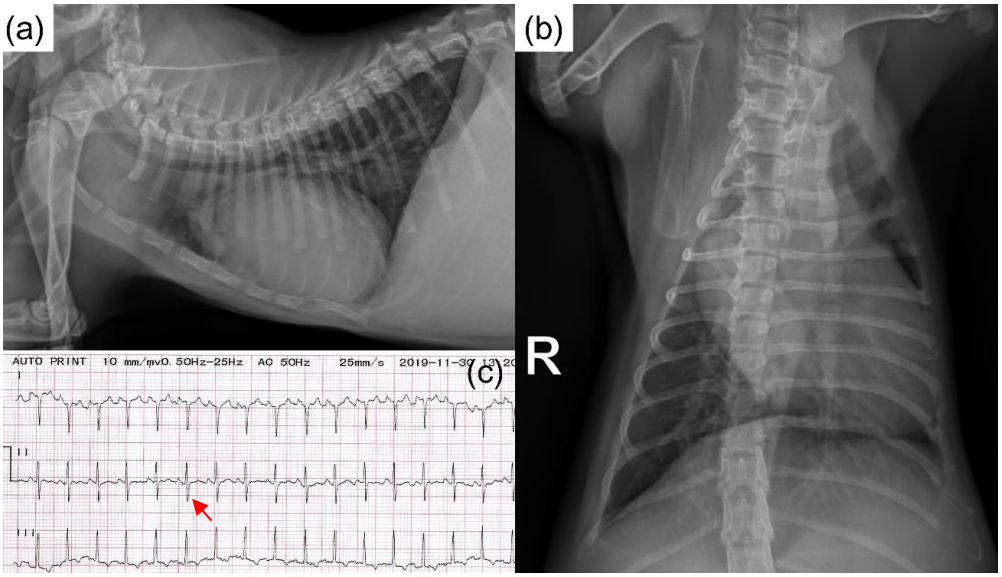

Figure 1. (a) Right lateral chest radiography showing severe cardiomegaly and dilated vena cava (asterisk). (b) The cardiac silhouette occupied almost the entire thoracic region on dorsoventral view. (c) Electrocardiogram showing sinus rhythm and deep S waves on leads I and II (red arrows).

Based on the above echocardiographic findings, a diagnosis of doubled-chambered RV, membranous ventricular septal defect, and pulmonic stenosis was made. These findings also led us to believe that the previous ascites was a manifestation of right congestive heart failure but not FIP, given the lack of consistent clinical signs such as jaundice, neurological deficits, and protein-laden effusion but the presence of hepatic venous congestion. Recommendations were given to replace diltiazem with atenolol (Ternolol 50 mg, 1 mg/kg, PO, SID) while maintaining the benazepril and clopidrogel medications. While the FIP treatment was not recommended, the owner decided to complete the course of treatment. Since then, the cat was more active and showed less breathing effort for about 2 weeks. Although the cat did not exhibit clinical signs of deterioration, the revisit 28 days later showed LV hypertrophy on the echocardiogram. The interventricular septal and LV free walls during diastole were 6.2 mm and 7.3 mm, respectively.

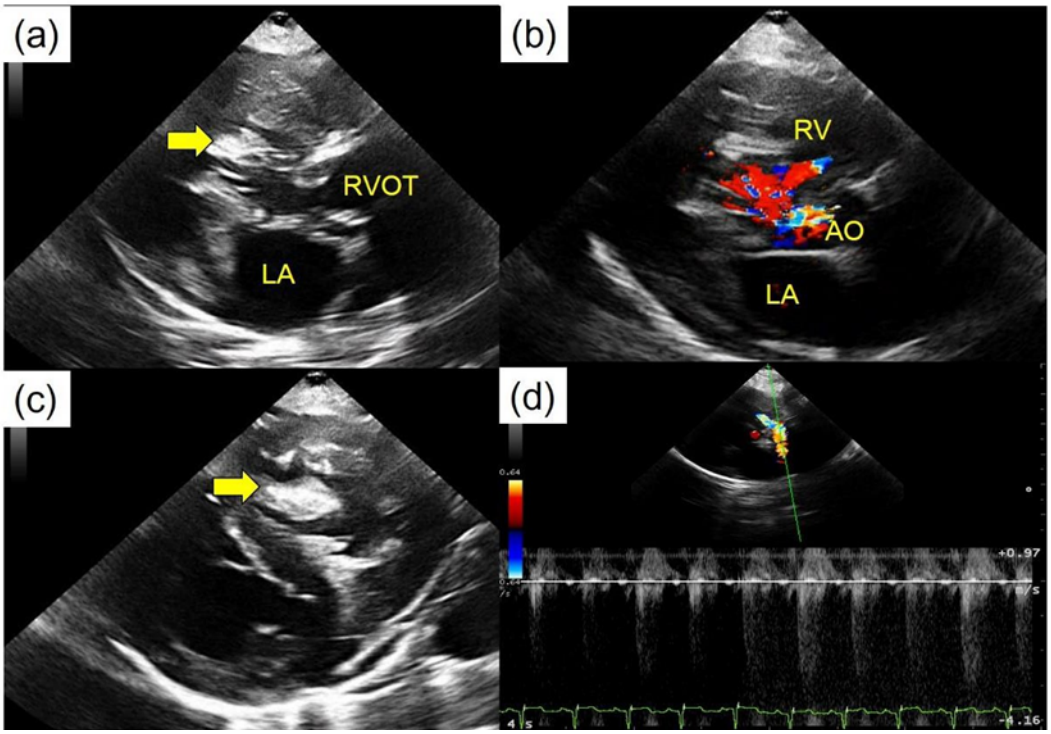

Figure 2. Echocardiogram showing (a) the presence of hyperechoic band in the right ventricle (RV) which inserts below the right ventricular outflow tract (RVOT). Note the severe RV hypertrophy. (b) The color Doppler showing communication between the left ventricle and RV through membranous ventricular septal defect. (c) The short axis view demonstrating the hyperechoic band and the RV hypertrophy. (d) On Doppler study, there was a high pressure gradient (3.27 m/s) across the stenotic RVOT, indicating pulmonic stenosis.

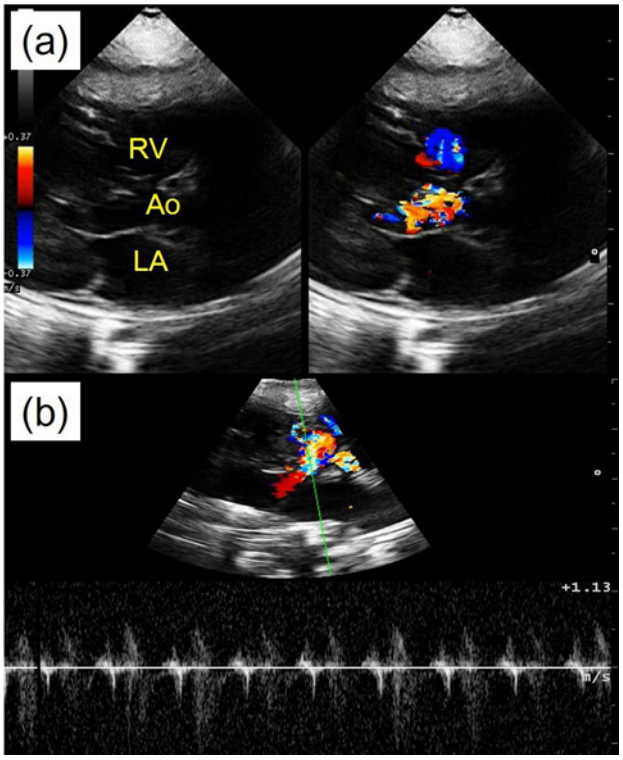

Figure 3. On day 28, the (a) B-mode and (b) Doppler studies showing (a) right-to-left shunt from right ventricle (RV) to left ventricle across the ventricular septum defect, indicating development of Eisenmenger syndrome. Ao: Aorta.

Further blood shunt from the RV to LV across the VSD was observed during systole, indicating the occurrence of Eisenmenger syndrome (Figure 3). The atenolol dosage was adjusted to the increased body weight (1.4 kg). On day 52, the cat showed high sleeping respiratory rates and distended abdomen. Serum biochemistry showed hypoglycemia [2.65 mmol/L; Reference range (RR): 4.11-8.84 mmol/L], mildly decreased creatinine (68 µmol/L; RR: 71-212 µmol/L), and markedly elevated alanine aminotransferase (>1000 U/L; RR: 12-130 U/L). The increase in alanine aminotransferase level was likely caused by hepatic congestion and hypoxia due to the right heart failure. Further, the normal globulin (47 g/L; RR: 28-51 g/L) and albumin (27 g/L; RR: 22-40 g/L) levels also indicated that the FIP and hypoproteinemia were less likely to cause the ascites. To control the ascites, furosemide (Lasix 40 mg, 1 mg/kg, PO, BID) was prescribed, which resolved the ascites for about 50 days. On day 148, it developed bilateral hind-limb paralysis and the owner opted for euthanasia.

Post-mortem limited to the heart and liver was performed with the owner's consent. Grossly, the RA and RV were markedly enlarged. Two muscular bands extended from the RV to about 5 mm distal the pulmonary valves were found (Figure 4a). On cross-section (Figure 4b), there was severe hypertrophy of the interventricular septum (10 mm), and LV (14 mm) and RV (11 mm) walls. The RV wall cross-section had multifocal whitish areas indicating myocardial infarction. A communicating hole between LV and RV was also identified (Figure 4c).

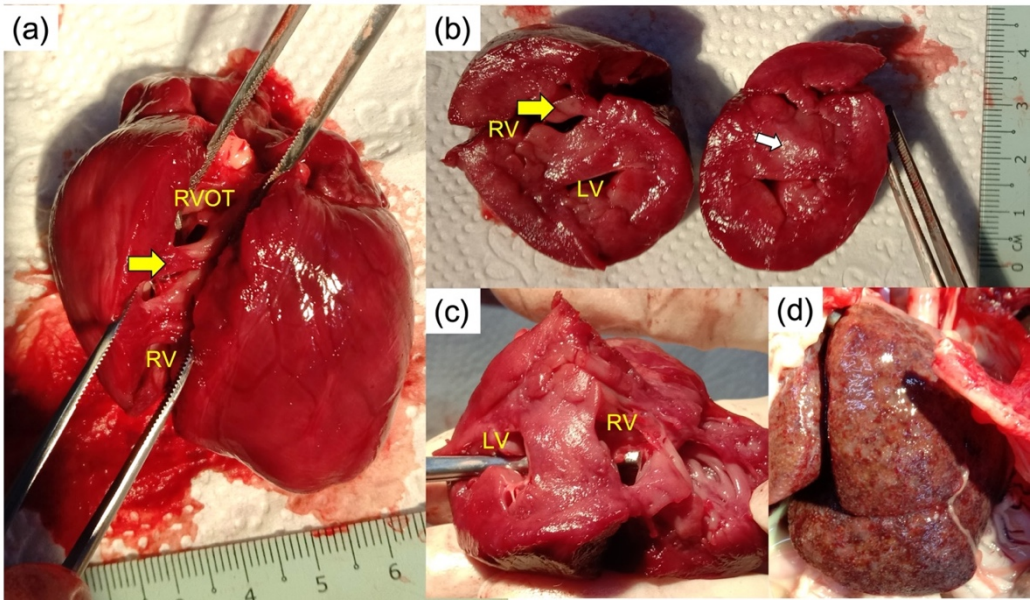

Figure 4. Post-mortem examination showing (a) presence of two muscular bands which extended from the right ventricular (RV) apex to distal to the right ventricular outflow tract (RVOT). (b) On cross-section of the heart, there was hypertrophy of the LV and RV. The muscular band (yellow arrow) and multiple whitish areas (white arrow) suggesting infarction were also observed. (c) A connecting hole from the LV to RV was documented. (d) The liver showed cirrhosis and nodular gene ration.

The liver also showed mottled appearance and cirrhosis with nodular degeneration (Figure 4d) suggestive of chronic passive congestion, and there was severe purulent ascites. The post-mortem results confirmed that the echocardiographic diagnosis of right congestive heart failure and Eisenmenger syndrome were due to DCRV and VSD. However, without performing a full-body post-mortem examination, histopathology, and immunochemistry, the FIP was not ruled out.

DISCUSSION

The present case represents Malaysia's first reported case of double-chambered right ventricle (DCRV), membranous ventricular septal defect (VSD), and pulmonic stenosis in a cat and provides insightful information about ante- and post-mortem findings of the rare feline congenital heart disease which subsequently developed Eisenmenger syndrome and hind-limb paralysis. While Koffas *et al*. (2007) discovered membranous VSD incidentally in a cat from nine cats diagnosed with DCRV, their study focused more on the clinical characteristics of DCRV so to that of the concurrent DCRV and VSD remains obscure. On the other hand, Koie *et al*. (2000) reported the concurrent DCRV and VSD in a female pug which died due to encephalitis.

In humans, the DCRV is often associated with other cardiac anomalies such as VSD, pulmonic stenosis, and tetralogy of Fallot (Chang *et al*., 1996; Fellows *et al*., 1977). Of these, VSD is present in up to 90% of cases and occurs as a result of improper expansion of the bulboventricular junction and incomplete fusion of the bulbar and endocardial cushion elements that normally close the superior portion of the ventricular septum (Galiuto *et al*., 1996). Further, the anomalous muscle bundles of the DCRV proposed to be due to localized aberrant overgrowth of the trabeculated myocardium (Hindle *et al*., 1968).

In cats, dilation of the obstructed RVOT with a balloon catheter (MacLean *et al*., 2002) and an arthroscopic grasping instrument (Mizuno *et al*., 2010) were attempted with disappointing outcomes. Indeed, the thick muscular band in the present case did not appear to be easily mended via an interventional therapy. On the other hand, Koffas *et al*. (2007) and Brockman et al. (2008) reported successful treatment of the DCRV via partial ventriculotomy and incised patch graft technique. However, there is no report on the treatment of the concurrent DCRV and VSD in cats. Indeed, the incised patch graft technique may relieve the high pressure within the RVOT, but the cat weighed only 1.4 kg, which was a little too small to allow an open-chest surgery. Furthermore, repairing the VSD under total venous inflow occlusion was deemed impossible without cardiopulmonary bypass and cardiopledgia.

The cardiogenic cause of ascites in the kitten was supported by central venous congestion, indicated by dilated caudal vena cava on the chest radiographs, right atriomegaly on echocardiography, dilated hepatic veins on liver ultrasounds, and mottled appearance of the liver on necropsy. By contrast, ascites due to feline infectious peritonitis (FIP) was questionable given the lack of consistent clinical signs and laboratory results. Although the cat showed positive antibody titer for feline coronavirus, as demonstrated by the preceding veterinarian, hyperglobulinemia and positive Rivalta's test were not observed, making the diagnosis of FIP highly unlikely. Further, the cause of hind-limb paralysis remained unclear, although we speculated that it could be caused by reduced tissue perfusion and hypoxia due to Eisenmenger syndrome as well as arterial thromboembolism given the severe bi-atrial enlargement.

CONCLUSION

While feline infectious peritonitis represents the top differential in kittens or young cats which show cavitary effusion, congenital heart defects such as double-chambered right ventricle (DCRV) with concurrent ventricular septal defect and pulmonic stenosis should also be suspected. In the context of making the diagnosis, the skill to evaluate the clinical presentation and interpret the laboratory and diagnostic imaging results is indispensable. Unfortunately, treatment outcome for DCRV is often disappointing.

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