

A Case of Right Ventricular Diverticulum with Pectus Excavatum

Mayumi Takeoka*; Tatsuya Kawasaki; Michiyo Yamano; Kyoko Ego; Yumiko Iwami; Tomoko Sakaguchi; Masanobu Nishikawa; Eiko Konishi; Tadaaki Kamitani

*Mayumi Takeoka

Clinical Laboratory, Matsushita Memorial Hospital, Sotojima 5-55, Moriguchi, Osaka, 570-8540, Japan
Tel: +81-66992-1231, Fax: +81-66992-4845; Email: takeoka.mayumi@jp.panasonic.com

Abstract

A right ventricular diverticulum is an extremely rare condition, and is mostly diagnosed in childhood. We report an adult patient with a diverticulum in the right ventricle, in whom contrast echocardiography was useful in making the diagnosis and deciding treatment strategy. An asymptomatic 32-year-old woman with pectus excavatum was referred to our hospital because of an abnormal electrocardiogram in a health checkup. Echocardiography showed the right ventricle and atrium compressed by the sternum, findings consistent with pectus excavatum, but was otherwise considered normal. Cardiac magnetic resonance imaging (MRI) incidentally showed a cystic lesion (13 mm by 10 mm) close to the apex of the right ventricle. A right ventricular diverticulum was a likely diagnosis, but the connection between the abnormal structure and the right ventricular cavity was not confirmed on MRI due to motion artifacts. Contrast echocardiography with agitated saline showed a direct connection between the cystic lesion and the right ventricular cavity. The agitated saline bubbles in the abnormal structure were almost washed out within several heart beats, in accordance with the disappearance of the saline bubbles from the right ventricle, findings likely indicating preserved contractility of the cystic lesion. The patient has been doing well without medication or surgery for more than seven years.

Keywords

contrast echocardiography; pectus excavatum; right ventricle; ventricular diverticulum

Abbreviations

MRI: magnetic resonance imaging

Introduction

A ventricular diverticulum is a rare condition characterized by a localized protrusion of the free wall of the ventricles that contains three wall layers and displays normal contraction [1,2]. Right ventricular diverticulum, as compared with left ventricular diverticulum, is considered less frequent and most of the conditions are diagnosed in childhood; no more than 30 cases with right ventricular diverticulum have been reported to date [3-7]. We describe an adult patient with a diverticulum in the right ventricle, in whom contrast echocardiography was useful in making the diagnosis and deciding treatment strategy.

Case Report

An asymptomatic 32-year-old woman with pectus excavatum was evaluated at our hospital because of a possible diverticulum in the right ventricle. Approximately six years before the current evaluation, the patient was referred to our hospital because of an abnormal electrocardiogram in a health checkup. Echocardiography showed the right ventricle and atrium compressed by the sternum, findings consistent with pectus excavatum, but was otherwise considered normal. Cardiac magnetic resonance imaging (MRI) showed no evidence of pericardial defect, but incidentally revealed a cystic lesion (13 mm by 10 mm) close to the apex of the right ventricle (Figure 1). A right ventricular diverticulum was the most likely diagnosis, but the connection between the abnormal structure and the right ventricular cavity was not confirmed on MRI due to motion artifacts. Echocardiography was performed again, with no additional information. She had been lost to follow up for six years until this representation.

On examination, the height was 161 cm, the body weight was 48 kg, and the body mass index was 18.5 kg/m². The vital signs were normal. The patient had a sunken appearance of the anterior chest, a finding consistent with pectus excavatum and reportedly unchanged over a decade. Neither additional heart sound nor murmur was audible. The remainder of the examination was normal. An electrocardiogram showed incomplete right bundle branch block and left atrial overload. Chest radiographs showed pectus excavatum and scoliosis (Figure 2).

Echocardiography showed normal left ventricular size and function, and a compressed right ventricle and atrium without tricuspid valvular disease (Figure 3a). The cystic lesion close to the right ventricular apex which was shown on MRI performed six years previously was not detected on routine views. However, oblique four-chamber apical views demonstrated a sack-like structure near the right ventricular apex (Figure 3b). The abnormal structure was likely to be connected to the right ventricular cavity, but little signal was obtained on color Doppler (Figure 3c). Furthermore, it remained unclear whether the structure had contractile muscle because it was difficult to demonstrate the sack-like lesion during a complete cardiac cycle on MRI.

Contrast echocardiography with agitated saline showed a direct connection between the cystic lesion and the right ventricular cavity (Figure 4). The agitated saline bubbles in the abnormal structure were almost washed out within several heart beats, in accordance with the disappearance of the saline bubbles from the right ventricle, findings likely indicating preserved contractility of the cystic lesion in the right ventricular apex. A diagnosis of right ventricular diverticulum hence seemed reasonable. The patient has been doing well without medication or surgery for more than seven years after the initial presentation.

Discussion

Differential diagnoses of an abnormal lesion next to the ventricles include pericardial cyst, ventricular aneurysm, and ventricular diverticulum [2]. In the present case, a pericardial cyst could be ruled out because contrast echocardiography showed a direct connection to the right ventricular cavity. Furthermore, given the absence of agitated saline bubbles retained in the abnormal structure, a ventricular aneurysm was a less likely diagnosis since akinesis or dyskinesis is common in a ventricular aneurysm whereas a ventricular diverticulum has normal or near normal contractility [5,8-10]. A

ventricular diverticulum can also be distinguished from a ventricular aneurysm based on its wall components; a ventricular diverticulum includes endocardium, myocardium, and occasionally pericardium, some of which are lacking in a ventricular aneurysm [8-10], although histological findings have not been obtained in our case.

Left ventricular diverticula appear to be associated with midline thoracoabdominal abnormalities [11-13]. Among defects related to the thoracoabdominal midline, a congenital condition involving the abdominal wall, sternum, diaphragm, pericardium, and heart is known as Cantrell syndrome [14]. This syndrome has also been reported to be seen in patients with left ventricular diverticula [15-17], biventricular diverticula [18], and a right ventricular diverticulum [6]. Our case did not show the pentalogy of Cantrell, but had pectus excavatum, which might be considered as one of the midline thoracoabdominal defects. An isolated ventricular septal defect also seems to occur in patients with a right ventricular diverticulum [19,20], but to our knowledge, no case has been reported in which a right ventricular diverticulum is present with pectus excavatum.

It is reasonable that ventricular diverticula are considered congenital although this condition may occur secondary, such as after chest injury [21]. In our case, a congenital diverticulum is most likely, since the patient had no remarkable medical history except for pectus excavatum. Furthermore, the coexistence of a right ventricular diverticulum and pectus excavatum may support that the diverticulum is congenital. A ventricular diverticulum appears to develop at a weak portion of the ventricular wall during the first few weeks of embryonic life [21]. Proposed conditions related to developing ventricular diverticula include viral infection in the utero, muscle and connective tissue defect, and excessive primordial cell stimulation [21,22]. It should be, however, noted that these features of ventricular diverticula are mostly based on the investigation of left ventricular diverticula; available data are entirely lacking regarding a right ventricular diverticulum, as seen in the current case, due to the extreme rarity.

Although the natural history of right ventricular diverticula has not been systematically studied, it seems benign and the need for surgery is, therefore, not considered mandatory because most patients are asymptomatic without any complication associated with ventricular diverticula over lifetime [2,20,21]. Our patient was informed about any potential risk, including rupture, embolism, or ventricular arrhythmias [4], although the incidence is unknown, and determined not to undergo surgical resection of the right ventricular diverticulum. Furthermore, anticoagulant therapy was not administered because of not only the absence of thrombus but also the rapid washout of saline bubbles in the diverticulum. Further studies are needed to establish the management of a right ventricular diverticulum.

The right ventricular diverticulum in the present case was initially missed on conventional echocardiography, but was revealed by chance on MRI which was performed to assess the pectus excavatum. Although echocardiography is a first-line modality for assessing function and structure of the heart, the weakness is with apical lesions due to difficulty in visualizing. In such conditions, it is well known that MRI should be considered to assess the apex, e.g., apical hypertrophic cardiomyopathy. In this case, however, apical evaluation was still difficult even on MRI. Computed tomography of the heart after the administration of contrast material may be an alternative option, but our patient hesitated to undergo computed tomography because of concerns about exposure to radiation and contrast material. In this case, agitated saline bubble contrast echocardiography was useful not only in making the diagnosis

but also in deciding the treatment strategy.

Figures

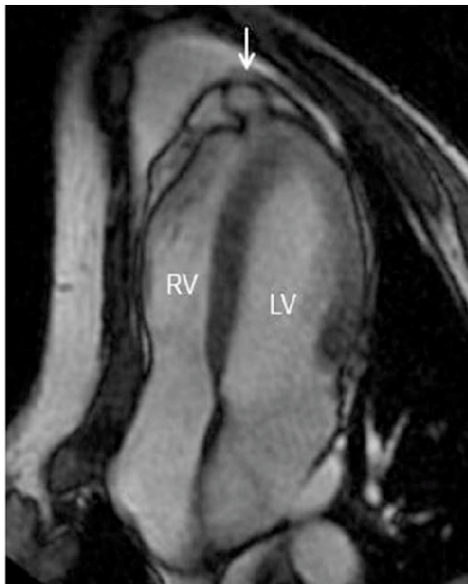


Figure 1: Magnetic resonance imaging four-chamber view shows a sack-like structure arising from the apex of the right ventricle (arrow), a finding suggestive of a right ventricular diverticulum. LV denotes left ventricle; RV, right ventricle.

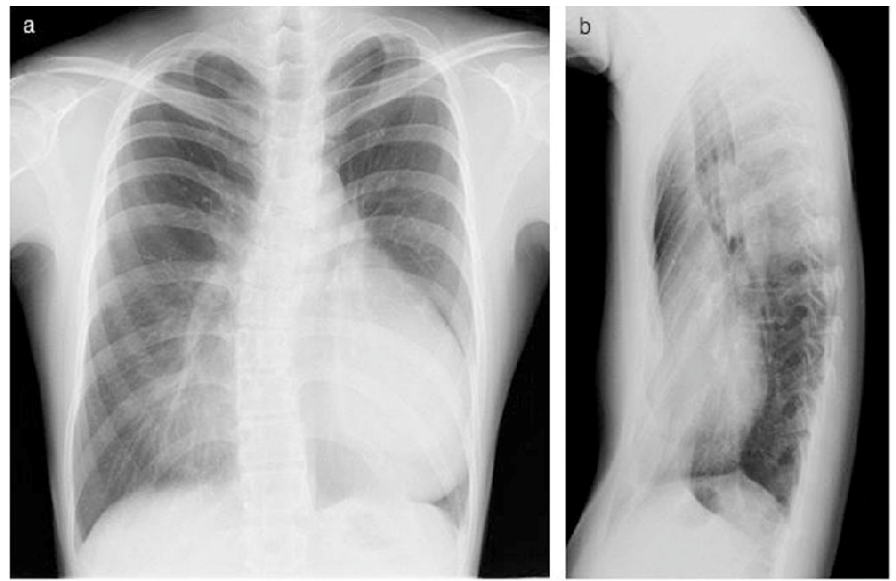


Figure 2: Chest radiograph posterior-anterior view shows scoliosis and cardiomegaly with a shift of the heart shadow to the left chest (a). A left lateral view shows a short distance between the anterior and posterior chest walls (b), as compared with the width of the chest, a feature consistent with pectus excavatum.

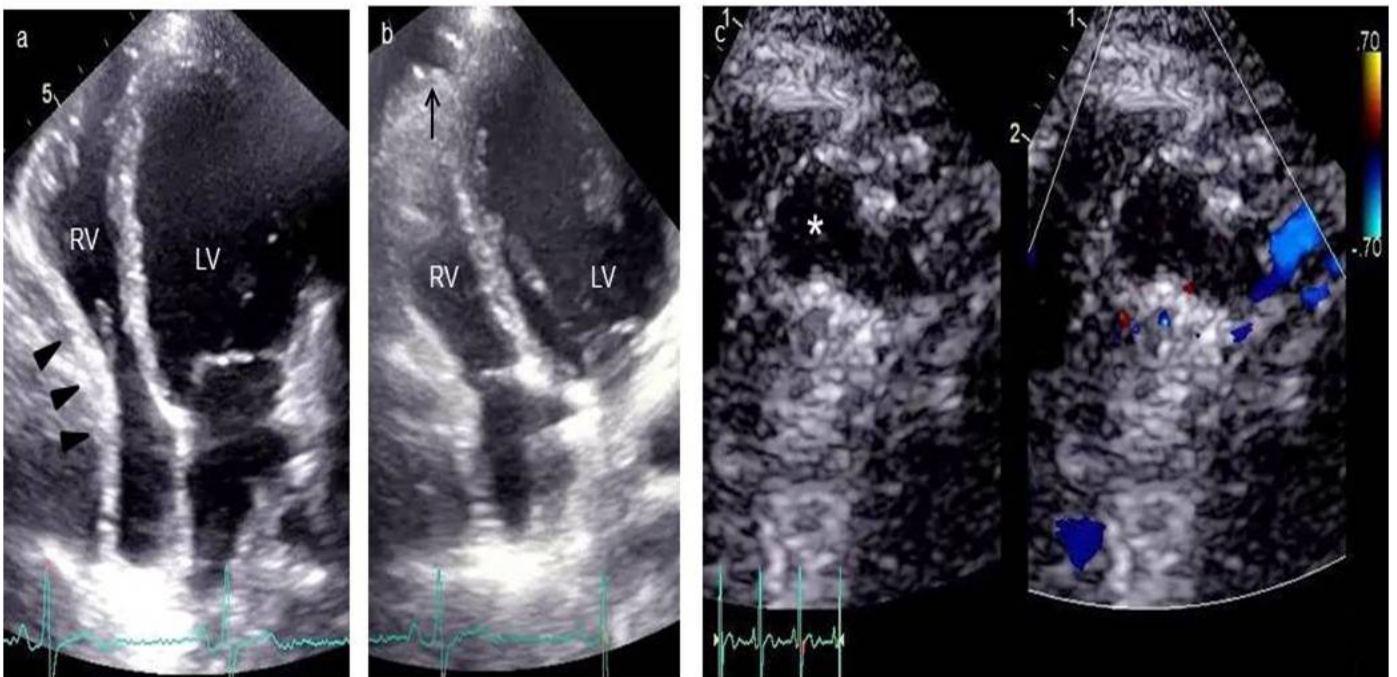


Figure 3: Transthoracic echocardiographic four-chamber view shows a compressed right ventricle and atrium (a, arrowheads). The oblique view of the four-chamber image reveals, with extreme difficulty, a cyst-like structure close to the ventricular apex (b, arrow). The abnormal lesion seems to be connected to the right ventricular cavity on B-mode (c, left panel, asterisk), but little signal was detected on color Doppler (c, right panel).

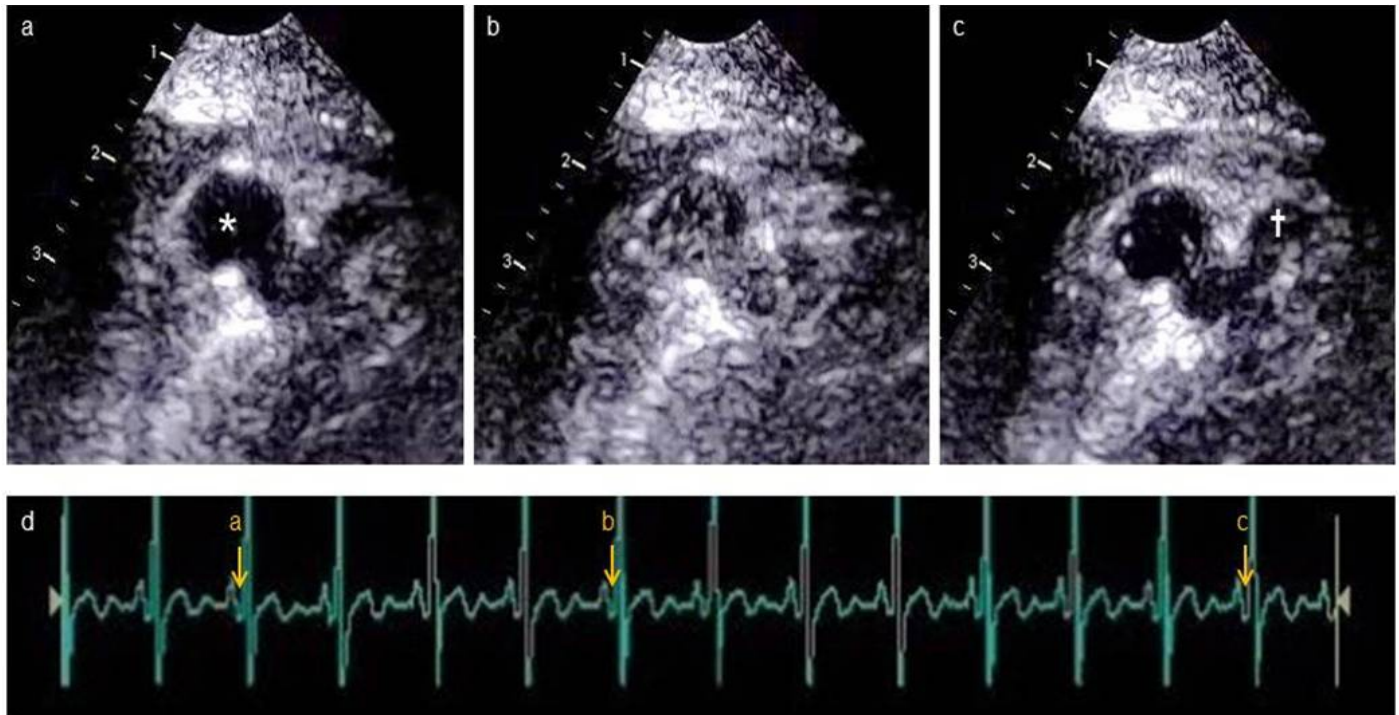


Figure 4: Contrast echocardiography shows that a cystic lesion close to the right ventricular apex (a, asterisk) is filled with agitated saline bubbles (b), which disappeared in several beats (c). Note the disappearance of agitated saline bubbles from the right ventricle (c, dagger). Panel a, b, and c were obtained on the timing of each arrow in Panel d.

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Authors Information: Mayumi Takeoka^{1*}; Tatsuya Kawasaki²; Michiyo Yamano²; Kyoko Ego¹; Yumiko Iwami¹; Tomoko Sakaguchi¹; Masanobu Nishikawa¹; Eiko Konishi¹; Tadaaki Kamitani²

¹Clinical Laboratory, Matsushita Memorial Hospital, Osaka, Japan

²Department of Cardiology, Matsushita Memorial Hospital, Osaka, Japan

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