Double-chambered left ventricle, or bifid cardiac apex, is a rare cardiac anomaly in which the left ventricular chamber is divided in 2 by a muscular band or septum, and its true incidence is unknown. It is usually incidentally discovered, and its clinical significance is uncertain. It may be associated with other congenital anomalies. It is important to distinguish double-chambered left ventricles and diverticula from other left ventricular outpouchings, such as aneurysms or pseudoaneurysms, as they have important prognostic implications. Multimodality imaging has an important role in the accurate assessment of morphologic characteristics of left ventricular outpouchings. We present a case of a double-chambered left ventricle and demonstrate how multimodality imaging guided us to the proper diagnosis and management.

CASE PRESENTATION

A 35-year-old woman presented to the Emergency Department with a complaint of palpitations. The patient denied having a prior history of palpitations, arrhythmias, syncope, presyncope, chest pain, or shortness of breath. Her social history was negative for illicit substance use and positive for 1 cup of coffee per day.

In the Emergency Department, vital signs showed a regular heart rate of 90 to 100 bpm, blood pressure of 133/79 mm Hg, respiratory rate of 16, and SpO2 of 94% on room air. An electrocardiogram demonstrated normal sinus rhythm with first-degree atrioventricular block. Laboratory evaluation showed normal electrolytes, troponin, and D-dimer and an elevated N-terminal-pro hormone brain natriuretic peptide of 557 pg/mL. A chest x-ray did not show sinus rhythm/sinus tachycardia with first-degree atrioventricular block, heart rates ranging from 60 to 130 bpm, and no evidence of atrial or ventricular arrhythmias. Outpatient, two-dimensional transthoracic echocardiography demonstrated an accessory chamber of the left ventricle in the mid to apical ventricular septum (Figure 1). The accessory chamber had a synchronous contraction with the main left ventricular chamber. Left and right ventricular systolic function were normal with a left ventricular ejection fraction of 63% using Simpson’s biplane method of disks. There were no significant valvular abnormalities. Three-dimensional transthoracic echocardiography also demonstrated the contractility of the accessory chamber (Video 1).

Subsequently, coronary computed tomography angiography was ordered to evaluate for possible anomalous coronary artery circulation and other associated congenital anomalies. Coronary computed tomography angiography showed a normal origin of the coronary arteries without evidence of atherosclerotic plaque or stenosis or other congenital anomalies. The left ventricular accessory chamber measuring 4.8 × 2.5 cm was redemonstrated, extending from the mid to apical septum. It was separated from the main left ventricular chamber by a muscle bundle and had a wide neck. There was no evidence of calcifications (Figure 2).

Furthermore, cardiovascular magnetic resonance imaging (CMR) was performed to evaluate myocardial tissue. Left ventricular function was normal with a left ventricular ejection fraction calculated as 60%. Both the main and accessory left ventricular chambers had normal wall thickness and synchronous contractility. There was no evidence of delayed gadolinium enhancement, and therefore the scarring or fibrosis that is most often noted in aneurysm or pseudoaneurysm was excluded (Figure 3, Videos 2 and 3). Early long-TI (600) sequence was performed, and there was no evidence of intracardiac thrombus in the accessory chamber on CMR.

Given the presenting symptom of palpitations and concern for possible ventricular arrhythmias, the patient also underwent stress echocardiography. There was no evidence of exercise-induced arrhythmias or stress-induced dyskinesis of the accessory left ventricular chamber. The patient was reassured as multimodality imaging confirmed accessory left ventricular chamber and lack of exercise-induced arrhythmias.

DISCUSSION

Outpouchings of the left ventricle, such as aneurysm, pseudoaneurysm, diverticulum, or accessory left ventricular chamber, have variable prognosis and hence different management strategies. Therefore, when evaluating patients with left ventricular outpouchings, it is crucial for the clinician to determine whether the outpouching is a left ventricular aneurysm, pseudoaneurysm, diverticulum, or accessory left ventricular chamber.
Double-chambered left ventricle and muscular diverticulum contain all 3 cardiac layers (epicardium, myocardium, and endocardium) and have synchronous myocardial contraction in systole with the main left ventricular chamber; however, the accessory left ventricular chamber has a wide communication with the main left ventricular chamber. Left ventricular true aneurysms with a broader neck also contain all 3 myocardial layers but will only show dyssynchronous myocardial contraction. Pseudoaneurysms, on the other hand, have a narrow neck, thin wall, and dyssynchronous motion. Table 1 describes associated features of each of these accessory chambers.

Two-dimensional transthoracic echocardiography is the initial, main modality for evaluation of cardiac structure and function. Two-dimensional echocardiography, especially with the aid of ultrasound-enhancing agent, can accurately evaluate left ventricular contractility and exclude left ventricular thrombus. Both aneurysm and pseudoaneurysm were excluded in this patient due to normal, synchronous contractility seen on the two-dimensional transthoracic echocardiogram (Videos 4 and 5). An association with congenital diverticulum and cardiac arrhythmias has been reported. One report found that the 2 most common presenting symptoms were syncope and palpitations. During clinical follow-up, sustained or nonsustained ventricular tachycardia was seen in 53% of the cohort.

Furthermore, coronary computed tomography angiography can evaluate for anomalous coronary origin, which can be a cause of ventricular arrhythmias or sudden cardiac death that can be associated with this entity. Coronary computed tomography angiography also evaluates for coronary artery disease, which can often be an important culprit of noncongenital left ventricular aneurysm or pseudoaneurysm. Coronary computed tomography angiography is also an excellent modality for evaluation of other congenital conditions that can coexist, and it can evaluate myocardial contractility if all cardiac phases are obtained, albeit at a cost of increased radiation to the patient.

Utilizing CMR in addition to the evaluation of cardiac function has an incremental role in myocardial tissue characterization. Cardiac magnetic resonance imaging demonstrates late gadolinium enhancement of the myocardium and pericardium in regions of fibrosis or infarct, which is often seen in cases of left ventricular aneurysm and pseudoaneurysm. In addition, CMR in pseudoaneurysm, which is a myocardial rupture contained locally by thrombi and pericardial adhesions without a myocardial layer, shows late gadolinium enhancement of the pericardium. Cardiac magnetic resonance imaging is also a more sensitive modality for the evaluation of left ventricular thrombi.

Cardiac diverticula are mostly found in the apical region. Apical diverticula are usually associated with midline thoracoabdominal defects and other heart malformations, whereas nonapical diverticula are isolated. Thrombosis, embolism, rupture, and ventricular arrhythmias can complicate these diverticula; however, the true incidence of these complications is not known.

Collectively, multimodality imaging can appropriately diagnose an accessory left ventricular chamber and differentiate it from other left ventricular outpouchings such as aneurysms or pseudoaneurysms.

**CONCLUSION**

The presence of an accessory left ventricular chamber is a rare condition with unknown clinical significance. The use of multimodality imaging is helpful for the differentiation of left ventricular outpouchings.
Figure 2  Cardiac computed tomography of the heart. (A) Axial plane. (B) Three-dimensional volume-rendered image. Arrows point to the left ventricular accessory chamber, which measured 4.8 × 2.5 cm. Ao, Aorta; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

Figure 3  Magnetic resonance imaging. (A) Cine segmented 4-chamber axial image. (B) Late gadolinium enhancement images with no evidence of gadolinium enhancement in the myocardium or pericardium. LA, Left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

Table 1  Characteristics of left ventricular outpouchings

| Architecture                              | Synchronous myocardial contraction | Communication/neck |
|-------------------------------------------|-----------------------------------|--------------------|
| Double-chambered left ventricle           | A division of the left ventricle involving the myocardium and endocardium creating a muscular septum | Yes                 | Wide               |
| Left ventricular diverticula              | Outpouching of the epicardium, myocardium, and endocardium | Yes                 | Narrow             |
| Left ventricular aneurysm                 | Outpouching of the epicardium, myocardium, and endocardium | No                  | Wide               |
| Left ventricular pseudoaneurysm           | Outpouching of the epicardium and pericardium only | No                  | Narrow             |
into aneurysm, pseudoaneurysm, or more benign forms such as diverticulum/double-chambered left ventricle. This case highlights how multimodality imaging was used to confirm the presence of a double-chambered left ventricle in this 5-chambered heart.

**SUPPLEMENTARY DATA**

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2022.06.004.

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