The Egyptian Heart Journal (2016) 68, 205–208

HOSTED BY

ELSEVIER

Egyptian Society of Cardiology

The Egyptian Heart Journal

www.elsevier.com/locate/ehj
www.sciencedirect.com

CASE REPORT

Right partial pericardial agenesis: An unusual case discovered during a heart surgery

A.W. Terra *, **C. Dommerc, F. Levy, A. Eker**

Centre cardio-thoracique de Monaco, 11 bis avenue d'Ostende, BP 223, MC 98004 Monaco Cedex, Monaco[1]

Received 2 July 2015; accepted 5 September 2015
Available online 21 October 2015

KEYWORDS

Partial agenesis;
Right pericardium;
Unusual

Abstract The pericardial agenesis occurs from a premature atrophy of the left Cuvier vein during embryonic development (Larsen, 2003). It is a non-frequent disease, most of the time accidentally discovered and mainly found at the left side of the pericardium (Centola et al., 2009). We report a case of right partial pericardial agenesis discovered during a sternotomy.

1. Introduction

The pericardial agenesis occurs from a premature atrophy of the left Cuvier vein during embryonic development.[1] It is a non-frequent disease, most of the time accidentally discovered and mainly found at the left side of the pericardium.[2] We report a case of right partial pericardial agenesis discovered during a sternotomy.

2. Case report

A 71-year-old woman was symptomatic for dyspnea class III of the New-York Heart Association (NYHA). Clinical examination revealed an apexo-axillary systolic ejection murmur. A 12-lead rest electrocardiogram (ECG) (Fig. 1) showed an atrial fibrillation (AF) associated with a right bundle branch block. A cardiomegaly was found in chest X-ray (Fig. 2). Echocardiogram revealed a grade III to IV degenerative mitral regurgitation (MR) due to the mitral leaflet prolapse, a dilatation of the left and right atria. The right ventricle appeared to be unusual, double-bubbled in the right medioventricular area (peanut shape) (Fig. 3). Because of extensive mitral annular calcifications, a valve replacement was performed. During this intervention, the cardiac surgeon discovered an atrial septal defect (ASD) associated with a partial pericardial agenesis (PPA) of the right ventricle. The pericardial defect, circle shaped, was measured at 5 cm on the free wall of the right ventricle. The decision was to perform an ASD closure by direct suture and to leave the PPA.

3. Discussion

Pericardial agenesis was first described in 1559, but was fully documented 400 years later in a living subject.[3] Pericardial sac becomes separated from the transverse septum by the growth of pleuropericardial folds,[1] which are linked to the blood supply by the vitelline, umbilical, anterior and posterior veins, called ducts of Cuvier. It is thought that pericardial agenesis results from incomplete development of either the transverse septum or the pleuropericardial folds.[4]

Ellis set a classification for pericardial agenesis according to its degree and localization: total absence, complete or partial

* Corresponding author. Tel.: +377 92168000.
E-mail address: info@ccm.mc (A.W. Terra).
[1] www.ccm.mc.

Peer review under responsibility of Egyptian Society of Cardiology.

http://dx.doi.org/10.1016/j.ehj.2015.09.001

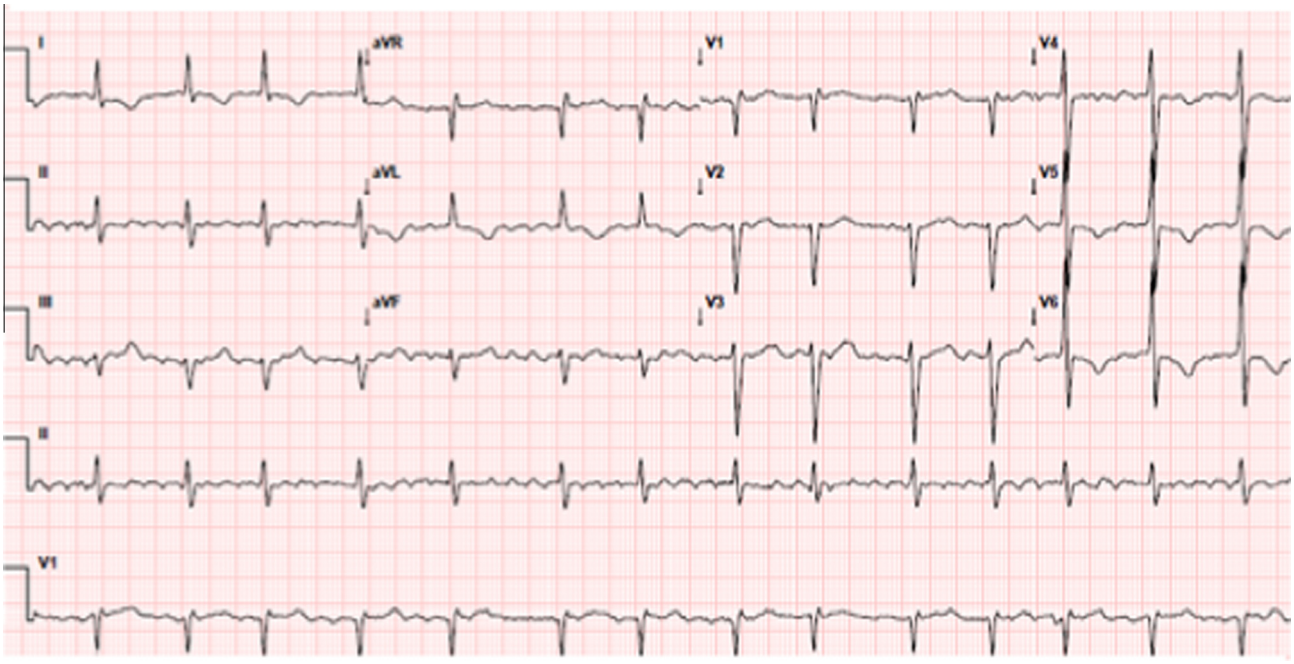

Figure 1 ECG right bundle block branch and atrial fibrillation.

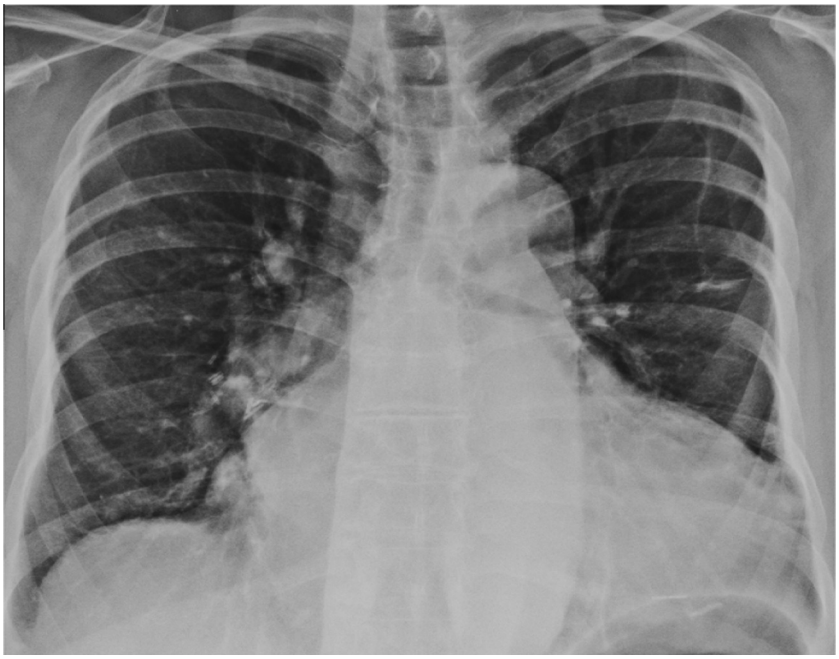

Figure 2 Cardiomegaly on chest X-ray.

left-sided defect, complete or partial right-sided defects, and diaphragmatic pericardium defects.[5]

In 70% of identified cases, there were left-sided pericardial defects due to the premature atrophy of the left duct of Cuvier. Right-sided or complete defects are very uncommon.[2]

Congenital absence of pericardium is a rare malformation, with a male preponderance (3 males per 1 female). Almost 30% of these patients present other congenital cardiovascular or pulmonary anomalies.[6] They may be associated with other syndromes such as VATER syndrome (vertebral defects, anal atresia, trachea esophageal fistula, renal and radial dysplasia), Marfan's syndrome and Pallister-Killian syndrome.[7] In 30% of these cases, agenesis is combined with a persistent arterial duct or ASD,[1] as described in our case.

Van Son et al. reported that 93.3% of patients were asymptomatic and that the defect was discovered incidentally.[8] In the remaining cases, the clinical presentation is nonspecific; the symptoms most commonly encountered are as follows: chest