International Journal of Clinical Medicine, 2016, 7, 450-453
Published Online July 2016 in SciRes. http://www.scirp.org/journal/ijcm
http://dx.doi.org/10.4236/ijcm.2016.77048

Holmes Heart and HIV: A Rare Combination of Two "H"s in a 23-Year-Old Widow

Adikesava Naidu Otikunta, Praveen Kumar Alane, YV Subba Reddy, Syed Imamuddin

Department of Cardiology, Osmania General Hospital and Osmania Medical College, Hyderabad, India
Email: oadikesavanaidu@gmail.com

Received 3 May 2016; accepted 8 July 2016; published 11 July 2016

Abstract

A 23-year-old, lean, scoliotic female presented to our hospital with a history of shortness of breath and cyanosis on exertion. Her 2D echocardiography revealed single left ventricle with both atrio-ventricular valves opening in it. She had normally related great arteries, with severe pulmonary artery hypertension, without pulmonary stenosis. Her blood tests indicated that she was reactive to human immunodeficiency virus (HIV-1). The patients died within 2 months despite treatments with anti-retroviral therapy and decongestive therapy.

Keywords

Double-Inlet Left Ventricle, HIV, Holmes Heart, Left Ventricular Hypertrophy, Rudimentary Right Ventricle

1. Introduction

Single ventricle, an unusual cardiac malformation, is usually associated with transposed great arteries [1]. Holmes heart is a rare variety of single ventricle, in which the great arteries are normally related [2]. Despite being recognized for more than a century, only few cases of Holmes heart have been reported till date [2]-[6]. Herein, we report a case of a 23-year-old lean scoliotic HIV-positive female who was diagnosed with Holmes heart. To the best of our knowledge, no case with a combination of HIV and Holmes heart has been reported previously.

2. Case Report

A 23-year-old thin-built female presented with a history of shortness of breath and palpitations, which were mainly exertional. She was poorly nourished with aphthous ulcers in mouth. Family history revealed that she was a second child of healthy consanguineous parents; her other siblings being normal. At the age of 15 years,

How to cite this paper: Otikunta, A.N., Alane, P.K., Reddy, Y.S. and Imamuddin, S. (2016) Holmes Heart and HIV: A Rare Combination of Two "H"s in a 23-Year-Old Widow. *International Journal of Clinical Medicine*, **7**, 450-453.
http://dx.doi.org/10.4236/ijcm.2016.77048

she got married to a man who was a lorry driver by profession. We were informed that her husband had passed away due to some unknown illness a year after their marriage. Physical examination of the patient suggested that she had mild scoliosis of dorsal vertebrae, without any kyphosis. Her body was cyanotic. In addition, she had grade-II clubbing. One year ago, she was admitted for acute heart failure and was treated with decongestive therapy. No other significant past illness was noted.

Cardiovascular examination of the patient showed left ventricular apex with a huge parasternal shift. Upon auscultation, a single second heart sound and medium pitched ejection systolic murmur (grade 3/6) in the left parasternal area were heard. Other systems were found normal. Chest X-ray in the posterior-anterior view revealed cardiomegaly with increased pulmonary vascularity along with a prominent convexity on left margin of cardiac shadow, suggestive of rudimentary outlet chamber (**Figure 1**). Electrocardiogram showed a right axis deviation with a deep S wave in anterior precordial leads (**Figure 2**). The 2D echocardiography indicated a single left ventricle with both atrio-ventricular valves opening in it (*i.e.* double-inlet left ventricle; **Figure 3**). The functional morphological left ventricle was hypertrophied, with normal situs and with severe pulmonary arterial hypertension, without pulmonary stenosis. A rudimentary outlet chamber was seen anteriorly and a dilated pulmonary artery was arising from it to the left of the ventricle. Accordingly, Holmes heart was diagnosed. In view of history of heart failure, the patient was planned to be treated with decongestive therapy.

Additional laboratory investigations suggested neutrophilic leukocytosis. The total white-blood cell (WBC) count was 12,500 with neutrophils showing 78% and lymphocytes 20%. We could not perform a detailed infectious screening in view of financial constraints. The enzyme-linked immunosorbent assay (ELISA) was found to be positive for the detection of antibodies for human immunodeficiency virus (HIV-1). Her CD4 count was 29 cells per cubic millimeter of blood. We confirmed the diagnosis of acquired immunodeficiency syndrome (AIDS). Since patient's family refused further in-hospital treatment, she was discharged from the hospital with prescription of medical therapy comprising zidovudine 300 mg BD, lamivudine 150 mg BD, and efavirenz 400 mg OD in addition to decongestive therapy. Two months later, we were informed by her relatives that the patient had died at home.

3. Discussion

Double-inlet left-ventricle accounts for about 1% of all congenital heart malformations. Holmes heart, *i.e.* double-inlet left ventricle with normally related great arteries, is still rare. There is no precise information regarding the prevalence of Holmes heart, apart from a few case reports in the modern literature. The characteristic features of Holmes heart include absence of the sinus (body or inflow tract), double-inlet left-ventricle, and

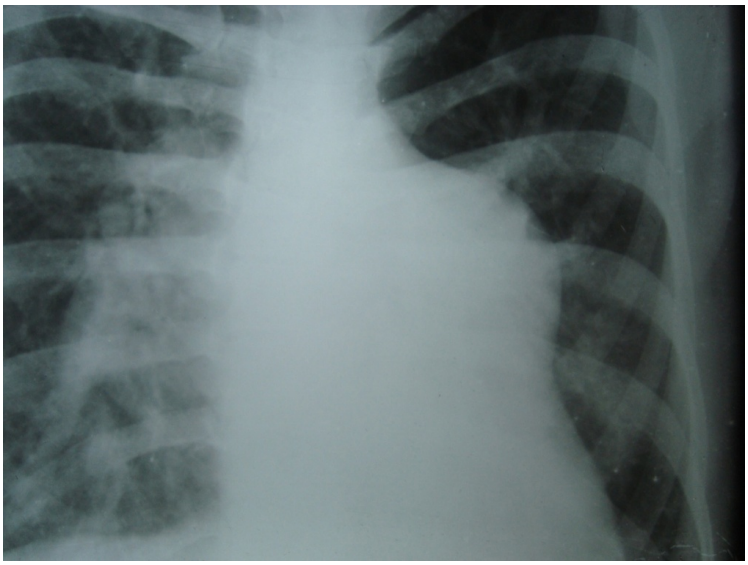

Figure 1. Chest X-ray showing cardiomegaly with increased pulmonary vascularity along with a prominent convexity on left margin of cardiac shadow, suggestive of rudimentary outlet chamber.

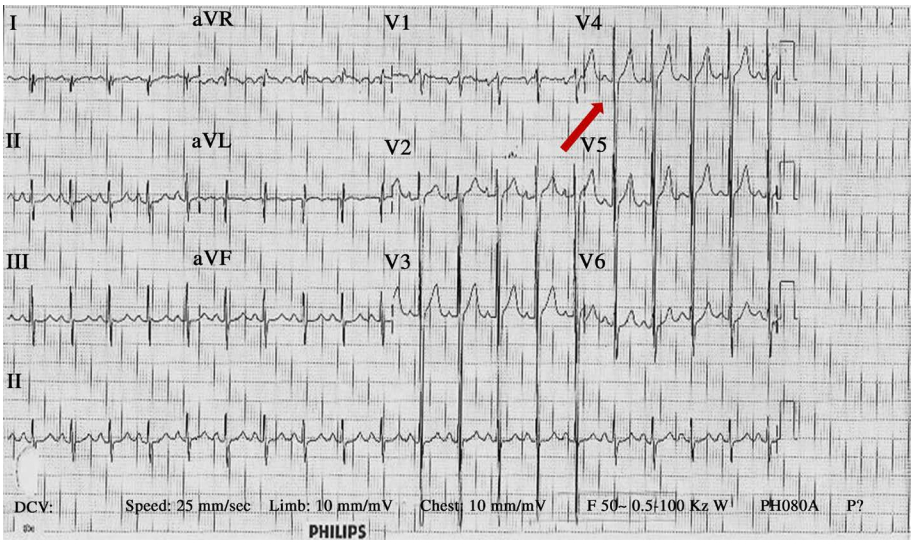

Figure 2. Electrocardiogram showing a right axis deviation with a deep S wave in anterior precordial leads.

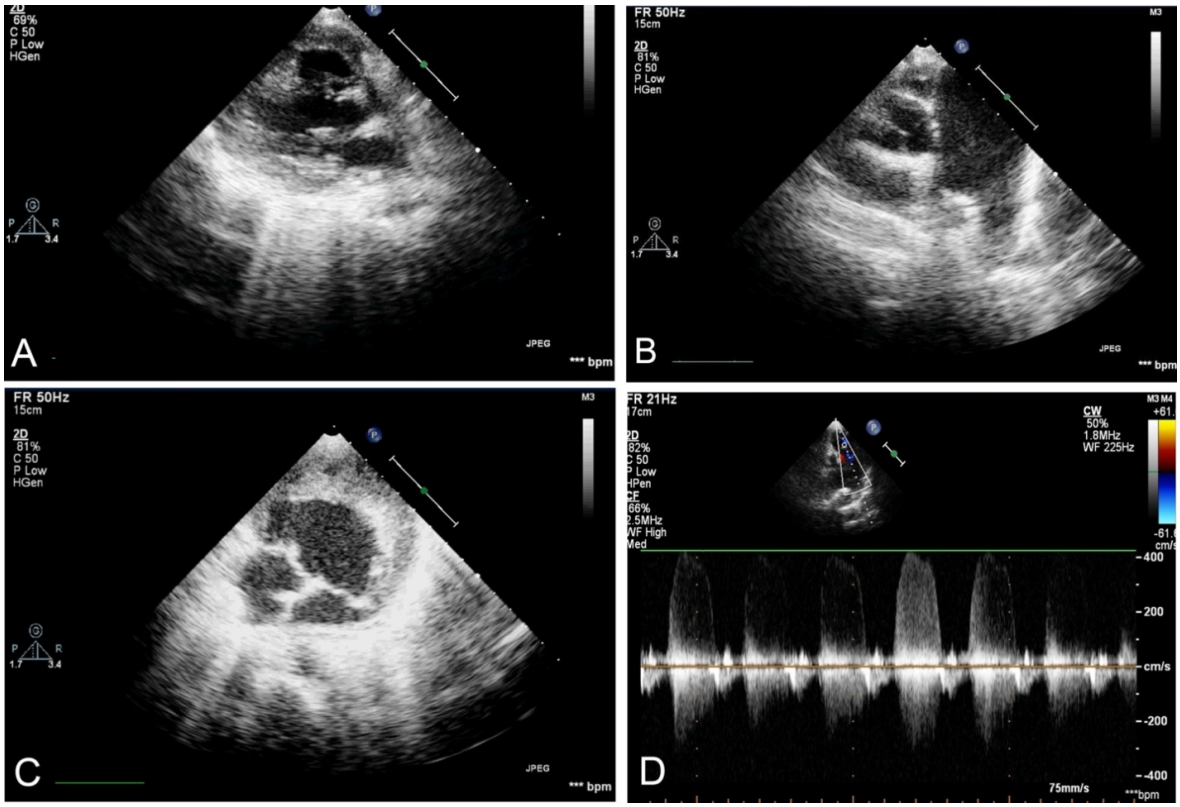

Figure 3. 2D echocardiography showing hypertrophy of left ventricle (A) Morphological left ventricle with rudimentary right ventricle (B) Normally related great vessels (C) Opening of both atrioventricular valves in single ventricle (D) Dilated pulmonary artery with moderate-to-severe pulmonary regurgitation.

normally related great arteries, with the pulmonary artery arising from the infundibular outlet chamber and the aorta arising from the single left ventricle [7]. Such patients with single ventricle usually die from heart failure, arrhythmia, or sudden death during infancy or childhood; however, only some patients reach adulthood [6].

In recent years, Sethi *et al*. have reported a case of a 10-year-old boy with effort dyspnea and cyanosis who was diagnosed to have Holmes heart type of univentricular heart with parachute mitral valve. They demonstrated

the usefulness of 2-dimensional and 3-dimensional echocardiography and multidetector computed tomography in the diagnosis of Holmes heart [8]. In another recent report, Weichert *et al*. described the echocardiographic findings of three fetuses with Holmes heart and addressed the pre- and perinatal management as well as additional abnormalities [9]. Conversely, Coasts *et al*. reported a case of double-inlet left-ventricle with concordant ventriculoarterial connections and a straddling tricuspid valve adherent to the malaligned ventricular septum in a 59-year-old woman [10]. However, reports of Holmes heart in adult patients are very rare.

Klaus *et al*. reported a case of single ventricle with normally related great arteries with sub-pulmonary stenosis in a 26-year-old adult patient [5]. In contrast, the 23-year-old patient in the present case did not exhibit pulmonary stenosis. Similar to our patient, a review of 14 cases of single ventricle with normal great arteries, without pulmonary stenosis, had reported that the oldest individual was 17 years in clinical group and 23 years in necropsy group [1]. Survival to even sixth decade has been reported by Gabbarini in a patient with Holmes heart without pulmonary stenosis [6]. Rahimtoola *et al*. explained the probable reasons for longer survival in Holmes Heart patients without pulmonary stenosis stating that only 16% of such patients displayed complete mixing, while more than half of the patients had their oxygenated blood selectively streamed to systemic circuit and unoxygenated blood selectively streamed to the lungs (favorable streaming) [11].

Apart from long survival, presence of scoliosis and diagnosis of AIDS along with the Holmes heart are distinctive features in the present case. Although the source of HIV infection or duration of suffering from HIV could not be identified, we believed that it could have been transferred from her deceased husband. Unfortunately, the patient died at her home within 2 months from the presentation at our hospital. The information related to her death was retrieved verbally through her relatives with no access to information related to her postmortem. We are of opinion that precise details about HIV infection or postmortem report could have offered more valuable insights regarding the patient discussed in the present case.

Overall, the distinctive features of the present case include long survival with Holmes heart (*i.e.* up to 23 years) and co-presentation of HIV and scoliosis with Holmes heart. Our case also provides valuable diagnostic information of Holmes heart by 2D echocardiography.

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