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Letter to the Editor

Unusual presentation of a patient with arrhythmogenic right ventricular dysplasia treated with a Glenn shunt

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Abstract

Clinically, arrhythmogenic right ventricular dysplasia (ARVD) usually presents with ventricular arrhythmias, and unusual presentations were reported as acute coronary syndrome, heart failure and electrical storm.

Taking all this different presentations and treatments in to account, we report a case of ARVD presenting with central cyanosis and clubbing simulating congenital heart disease. Besides this unusual presentation, the patient underwent also an unusual operation for this kind of abnormality, which cured the cyanosis completely.
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Keywords: Arrhythmogenic right ventricular dysplasia; Cyanosis; Glenn shunt

Arrhythmogenic right ventricular dysplasia (ARVD) is characterized by progressive fibrofatty replacement of the right ventricular myocardium [1]. Clinically, ARVD usually presents with ventricular arrhythmias [2], and this is also the underlying disease in a substantial number of sudden deaths among apparently healthy individuals [3]. There are also unusual presentations such as acute coronary syndrome, heart failure and electrical storm [4–6].

With regard to the treatment of ARVD, there are no precise guidelines to determine who are the patients who need to be treated and which is the best management approach. Usually, therapy is directed to prevent and/or treat malignant ventricular tachyarrhythmias with medications, implantable cardioverter defibrillator and radiofrequency ablation in selected cases [7]. In patients in whom ARVD has progressed to severe right ventricular or biventricular systolic dysfunction, treatment consists of current therapy for heart failure [8]. In case of refractory heart failure, the patients may become candidates for heart transplantation [8].

Taking all this different presentations and treatments in to account, we report a case of ARVD presenting with central cyanosis and clubbing simulating congenital heart disease. Besides the unusual presentation, the patient underwent also an unusual operation for this kind of abnormality, which cured the cyanosis completely.

A 16-year-old female with a history of cyanosis for 8 years was referred to our hospital for further evaluation. The patient has been living in the rural area and did not seek any medical attention until she applied for the first time to see a general practitioner due to gastric pain. After referred to several hospitals due to cyanosis, the patient finally sent to our hospital from a chest disease hospital, where she was investigated for pulmonary arterio-venous fistula and was found clear after a thorax magnetic resonance (MR) angiogram. Her main complaint was exercise dyspne, palpitation and fatigue since the day cyanosis had begun.

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She had a family history of a 30-year-old uncle who died suddenly with unknown cause.

The physical examination revealed central cyanosis, clubbing of digits, a raised jugular venous pressure and haepatomegaly. The presenting ECG showed T wave inversion in the precordial leads exploring the right ventricle (V1–V3). Right precordial QRS prolongation and epsilon waves were evident. The chest X-ray showed considerable right heart prominence. Pulmonary arteries were normal and lung fields were clear. Haemoglobin was 17.3 g/dl. Blood gases were pH 7.49, PCO_2 27 mm Hg, PO_2 53 mm Hg and O_2 saturation was 87%. Lung function tests (spirometry, lung volumes, flow volume loops and transfer factor) were all normal.

The transthoracic echocardiogram (TTE) showed a severely dilated right ventricle (RV). The right atrium (RA) was also dilated. The aneurysmal dilations of the diaphragmatic, apical and infundibular regions (so called "triangle of dysplasia") were conspicuous suggesting ARVD (Fig. 1a,b). RV ejection fraction (EF) was estimated as 10%. Although the tricuspid valve structure was normal, severe tricuspid regurgitation due to annular dilation existed.

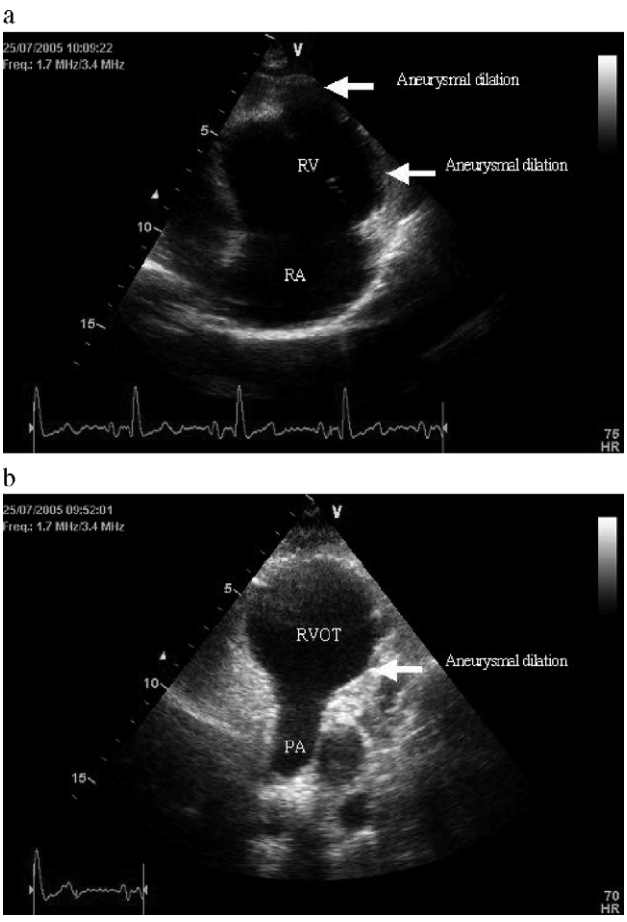

Fig. 1. (a) Arrows showing aneurysmal dilation of the right ventricular (RV) apex and RV inflow tract. RA: right atrium. (b) The aneurysmal dilation of the right ventricular outflow tract (RVOT) is the third component of the "Marcus triangle". PA: pulmonary artery.

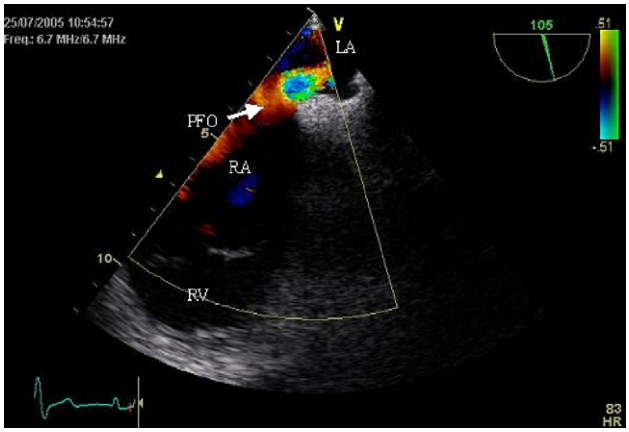

Fig. 2. Transesophageal echocardiography demonstrating a right to left shunt through a patent foramen ovale (PFO). LA: left atrium, RA: right atrium, RV: right ventricle.

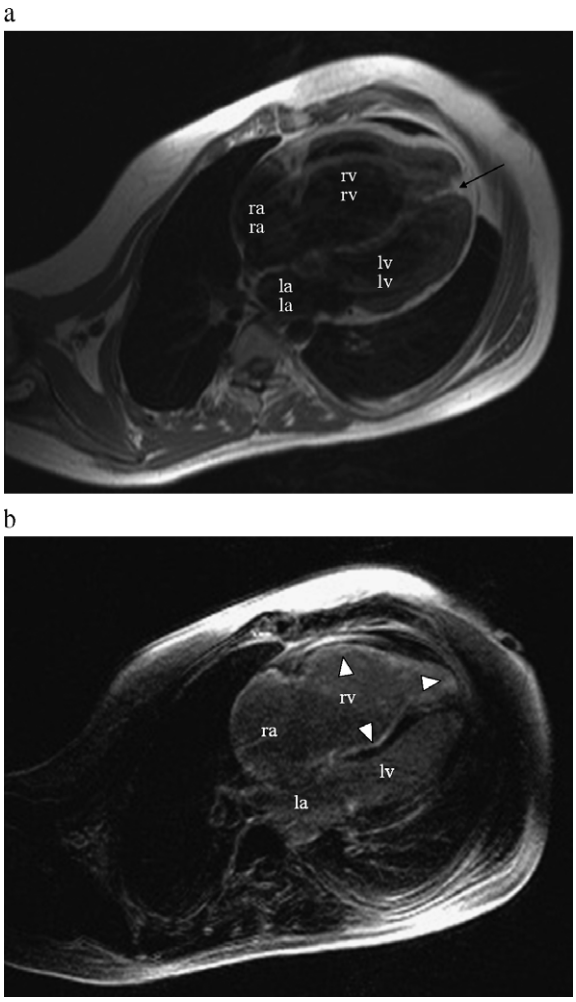

Fig. 3. (a) Cardiac magnetic resonance imaging (MRI) is showing fatty infiltration at the apex (black arrow). ra: right atrium, la: left atrium, rv: right ventricle, lv: left ventricle. (b) MRI obtained at the 15 min after contrast injection indicating fibrosis at the right ventricular lateral wall, apex and the right ventricular side of the septum (arrow). ra: right atrium, la: left atrium, rv: right ventricle, lv: left ventricle.

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