Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/jicc

Case Report

Restrictive ventricular septal defect in a case of tetralogy of Fallot with situs inversus and dextrocardia – A case report

Pranil Gangurde [a,*], *Abhay Tidake* [b], *Hetan Shah* [c], *Ajay Mahajan* [d]

[a] *Cardiology Registrar, Department of Cardiology, LTMMC and LTMGH, Sion, Mumbai, India*
[b] *Senior Resident, Department of Cardiology, LTMMC and LTMGH, Sion, Mumbai, India*
[c] *Associate Professor, Department of Cardiology, LTMMC and LTMGH, Sion, Mumbai, India*
[d] *Professor, Department of Cardiology, LTMMC and LTMGH, Sion, Mumbai, India*

ARTICLE INFO

Article history:
Received 22 December 2014
Accepted 31 March 2015
Available online 23 May 2015

Keywords:
Fallot
Dextrocardia
Restricted VSD

ABSTRACT

Tetralogy of Fallot is diagnosed in about 3.5% of patients who have congenital heart defects. In tetralogy of Fallot, ventricular septal defect is usually large because of malalignment of outlet septum. But the restrictive ventricular septal defect has been reported rarely & its association with dextrocardia and situs inversus is uncommon. We report all these findings in a 10-year-old boy and review the medical literature relevant to these combined conditions.

1. Introduction

Tetralogy of Fallot is diagnosed in about 3.5% of patients who have congenital heart defects. The condition consists of ventricular septal defect, overriding aorta, right ventricular outflow tract obstruction, and right ventricular hypertrophy.[1] Although tetralogy of Fallot is a common congenital anomaly, its association with dextrocardia with situs inversus with a restrictive ventricular septal defect is an uncommon variant. The most common cause of restriction is accessory tricuspid leaflet tissue.

2. Case report

A ten-year-old boy presented to the emergency department with complaints of effort intolerance with edema feet since last 6 months. He was also complaining of gradually progressive abdominal distension. He was apparently asymptomatic six months before, though he had experienced shortness of breath upon exertion. He had a history of cyanosis since early childhood. No history suggestive of cyanotic spell or squatting.

His body weight was 30 kg, and his height was 130 cm, which were in the 10th percentile.

* *Corresponding author*. Tel.: +91 9823871970.
E-mail address: pranil.bjmc@gmail.com (P. Gangurde).
http://dx.doi.org/10.1016/j.jicc.2015.03.025

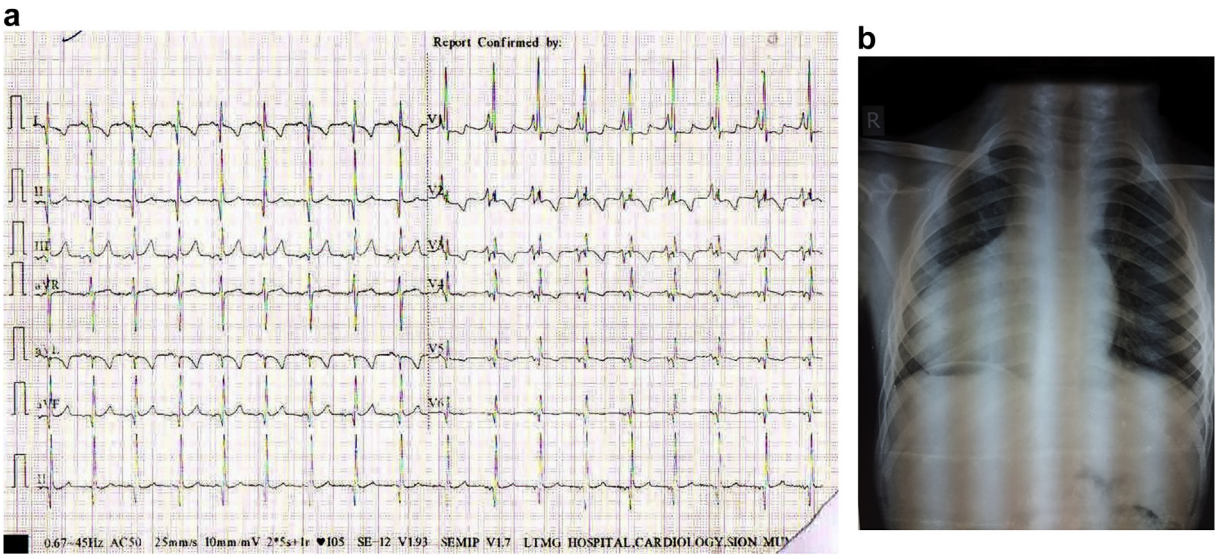

Fig. 1 – a – A 12-lead electrocardiogram was consistent with dextrocardia showing sinus tachycardia with right axis deviation with right atrial enlargement with right ventricular hypertrophy with strain pattern. b – Chest radiographs showed dextrocardia with total situs inversus, cardiomegaly with right ventricular hypertrophy with right atrial enlargement.

On examination, his heart rate was 110 beats/min, blood pressure of 90/60 mmHg. He had uniform central cyanosis, scleral hyperemia, grade 3 pandigital clubbing, prominent 'a' wave in Jugular venous pulse, RV type of apex, grade 3 parasternal heave on right side. Cardiac auscultation revealed a 3/6 pansystolic murmur at left lower sternal border & grade 3/6 systolic ejection murmur, best heard at the right upper sternal border. His systemic oxygen saturation was 85% with a PCV of 57%. Routine blood investigations showed derranged liver function tests (raised serum bilirubin with total 3.5 mg/dL, direct – 1.2 mg/dL, raised liver enzymes) and derranged renal function tests (serum creatinine – 2.4 mg/dl, blood urea – 73 mg/dl).

A 12-lead electrocardiogram was consistent with dextrocardia showing sinus tachycardia with right axis deviation with right atrial enlargement with right ventricular hypertrophy with strain pattern (Fig. 1a).

Chest radiographs showed dextrocardia with total situs inversus, cardiomegaly with right ventricular hypertrophy with right atrial enlargement (Fig. 1a).

Transthoracic echocardiograms revealed the anatomy of dextrocardia with situs inversus and tetralogy Of Fallot. There was dilated morphological right atrium and right ventricle, left sided aortic arch with overriding of aorta (Fig. 2a). A large subaortic ventricular septal defect noted with near complete closure by septal tricuspid leaflet (Fig. 2b). Restrictive VSD with pressure gradient across (Fig. 3). There was a gradient of 110 mmHg across the RVOT (Fig. 4) with prominent subinfundibular hypertrophy of muscle bundle in right ventricular outflow tract (Video 1). Pulmonary arteries were normal and confluent. There was severe morphological tricuspid valve regurgitation. (Video 2)

Supplementary data related to this article can be found online at http://dx.doi.org/10.1016/j.jicc.2015.03.025.

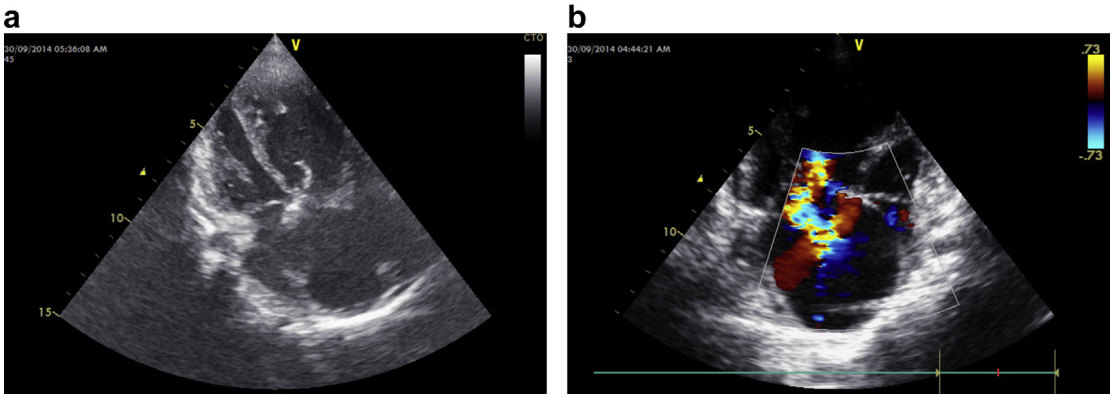

Fig. 2 – a – There was dilated morphological right atrium and right ventricle, left sided aortic arch with overriding of aorta. b – A large subaortic ventricular septal defect noted with near complete closure by septal tricuspid leaflet.