BANTAO Journal 2015; 13(2): 84-86

BJ

BANTAO Journal

Case report

Left Ventricular Cleft Detected by Transthoracic Echocardiography in a Patient with Autosomal Dominant Polycystic Kidney Disease

Gulay Gok[1], Ibrahım Ilhan[1], Ibrahim Beydili[1], Tolga Sinan Guvenc[2], Adnan Kaya[1] and Kemal Magden[3]

[1]Mardin State Hospital, Department of Cardiovascular Disease, [2]Siyami Ersek Thorasic and Cardiovascular Research Center, Department of Cardiovascular Disease, [3]Mardin State Hospital, Department of Nephrology, Turkey

Abstract

Recently, the presence of left ventricular clefts has been identified frequently with the advancement of cardiac imaging modalities such as cardiac magnetic resonance imaging and computed tomography. Here we report a rare case of left ventricular cleft that was incidentally diagnosed with the ECG changes that imitated the presence of LMCA stenosis and diagnosed by transthoracic echocardiography in a patient with autosomal dominant polycytic kidney disease.

Key words: policystyc kidney disease, left ventricular clefts, ECG, echocardiography

Introduction

Left ventricular clefts are defined as slit or fissure-like protrusions through the >50% of compact myocardium and tending to occlude with the myocardial contractions. The presence of left ventricular clefts has a higher incidence in patients who has gene mutations related to hypertrophic cardiomyopathy. However, in our case the left ventricular cleft was incidentally detected in a patient who has end-stage autosomal dominant polycystic kidney (ADPKD) disease. Besides this, ADPKD has many cardiac manifestations; association with left ventricular cleft has not been reported previously to the best of our knowledge.

Case report

A 55-year-old woman without any known history of coronary artery disease or family history was referred to our Department due to ischemic ECG changes. She has had ADPKD for 5 years and she was planning to undergo hemodialysis due to end-stage ADPKD. Her blood pressure was 100/60mmhg and the heart rate was 73 bpm. ECG revealed ST elevation in lead aVRand ST depression predominantly in leads DII, DIII, aVF, V4-V6 (Figure 1).

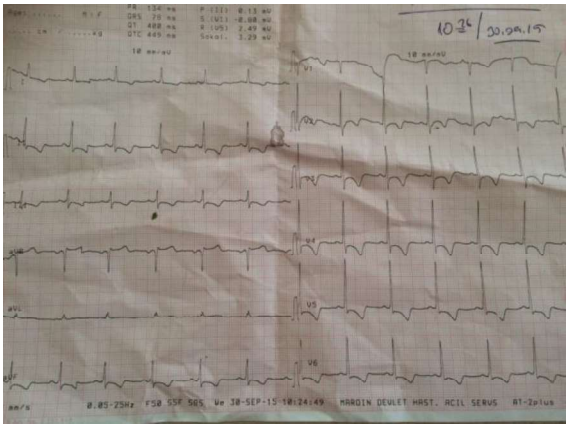

Fig. 1. ECG changes: ST elevation in lead aVRand. ST depression in leads DII, DIII, aVF, V4-V6

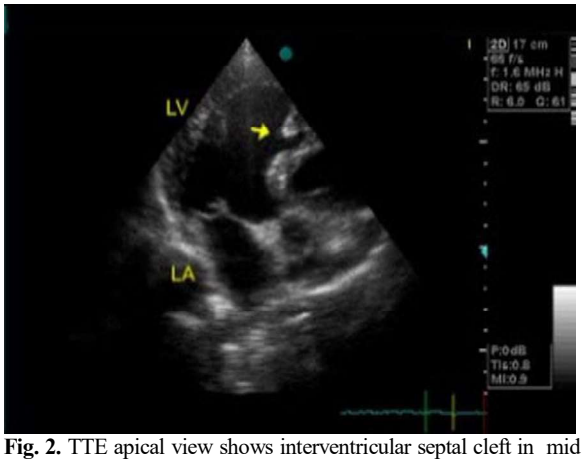

Fig. 2. TTE apical view shows interventricular septal cleft in mid diastolic frame. Arrow points the cleft in mid interventricular septum

Laboratory analyses showed that her mineral status and cardiac enzymes were in normal range. Subsequently, transthoracic echocardiography (TTE) was performed and it showed no wall motion abnormality in parasternal long or short-axis views. In apical 4-chamber view; a U-

Correspondence to: Gulay Gok, Nur street 20th Avenue Konbay Apartments Floor: 2 Artuklu/Mardin. Post office box: 47000; Phone: +90 53 64 902 907; Fax: 0482 212 42 23; E-mail: glygk84@gmail.com

shaped, slit-like appearance with an extension through the compact myocardium was noted in mid interventricular septum (Figures 2 and 3). This fissure-like abnormality was tending to narrow with myocardial contractions which was compatible with the diagnosis of left ventricular clefts. There was no evidence of flow though the myocardial defect with color doppler examination or no pressure gradient by pulse wave examination which helped to distinguish from ventricular septal defects by TTE (Figure 4). Although ECG changes theoretically showed the presence of LMCA stenosis, immediate coronary angiography showed no significant stenosis of the coronary arteries. The patient underwent dialysis program, soon after she was discharged with anticoagulation therapy.

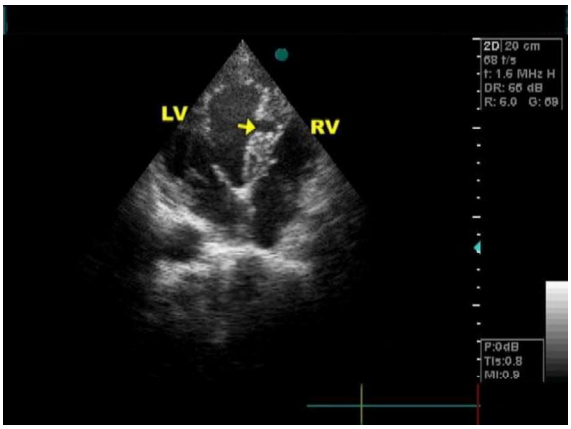

Fig. 3. TTE apical 4C view shows the U-shaped, slit-like appearance of the cleft in mid interventricular septum

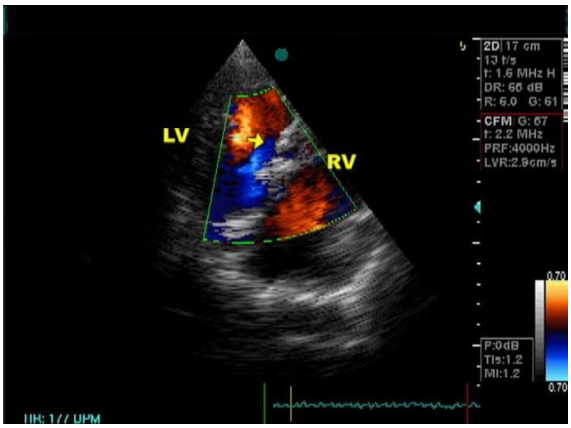

Fig. 4. TTE with color doppler interrogation showing no flow through the cleft
(Abbreviations: TTE: Transthoracic Echocardiograhy. LV: left Ventricle, RV: right Ventricle, LA: Left Atrium)

Discussion

The presence of left ventricular clefts has been identified by CTA and MRI imaging modalities over the past decades. It has been described as V- shaped, slit-like appearance, extending >50 of myocardium and tending to narrow in systole and with a higher prevalence in patients with hypertrophic cardiomyopathy (HCM) gene mutation [1,2]. Petryka *et al*. found higher percentage of left ventricular clefts in patients who were referred for cardiovascular MRI with HCM, myocarditis, and hypertension [3]. The cause of clefts is not known but it is thought to be disarray of myocardium in patients with HCM [4]. Left ventricular clefts should be differentiated from left ventricular diverticulum, which are defined as saccular protrusions beyond the myocardial borders and have a high incidence of thromboembolism. The treatment of left ventricular cleft is done with anticoagulation therapy to prevent tromboembolism. Surgery is usually not needed.
In our case the shape of the fissure-like appearance was compatible with the definition of the left ventricular cleft, but it was in a patient with ADPKD which is a rare coexistence. The detected cardiac manifestations of ADPKD are: left ventricular hypertrophy, heart valve abnormalities, coronary artery dilatation, atrial septal aneurysms, interrupted aortic arch, aortic dissection, dilatation and coarctation [5-7]. But, the presence of left ventricular cleft in patients with ADPKD has not been previously reported. Boutter *et al*. have found that pkd1 and pkd2 gene mutations are associated with ventricular and atrial septation in mice and that could be a possible explanation of coexistence of left ventricular clefts and ADPKD [8,9].

Conclusion

Patients who have ADPKD and with pkd1 and pkd2 mutations may have cardiac septation anomalies such as atrial septal defects, interatrial septal aneurysm, etc. Left ventricular cleft, which is thought to be congenital defect of myocardium, may be detected by MRI or TTE in patients with ADPKD as it was the case with our patient.

Conflict of interest statement. None declared.

References

1. Johansson B, Maceira AM, Babu-Narayan SV, *et al*. Clefts can be seen in the basal inferior wall of the left ventricle and the interventricular septum in healthy volunteers as well as patients by cardiovascular magnetic resonance. *J Am Coll Cardiol* 2007; 50: 1294-1295.
2. Germans T, Wilde AA, Dijkmans PA, *et al*. Structural abnormalities of the inferoseptal left ventricular wall detected by cardiac magnetic resonance imaging in carriers of hypertrophic cardiomyopathy mutations. *J Am Coll Cardiol* 2006; 48: 2518-2523.
3. Kuribayashi T, Roberts WC. Myocardial disarray at junction of ventricular septum and left and right ventricular free walls in hypertrophic cardiomyopathy. *Am J Cardiol* 1992; 70(15): 1333-1340.
4. Petryka J, Baksi AJ, Prasad SK, *et al*. Prevalence of inferobasal myocardial crypts among patients referred for cardiovascular magnetic resonance. *Circ Cardiovasc Imaging* 2014; 7(2): 259-256.
5. Helal I, Reed B, Mettler P, *et al*. Prevalence of cardiovascular events in patients with autosomal dominant polycystic kidney disease. *Am J Nephrol* 2012; 36(4): 362-370.
6. Hadimeri H, Caidahl K, Bech-Hanssen O, Nyberg G. Echocardiographic findings in kidney transplant patients with

autosomal dominant polycystic kidney disease. *Scand J Urol Nephrol* 2009; 43(5): 416-419.
7. Catapano F, Pancaldi S, Pace Napoleone C, *et al.* An unusual cardiac manifestation in autosomal dominant polycystic kidney disease. *Case Rep Nephrol* 2012; 2012: 978170.
8. Boulter C, Mulroy S, Webb S, *et al*. Cardiovascular, skeletal, and renal defects in mice with a targeted disruption of the Pkd1 gene. *Proceed Nat Acad SciUSA* 2001; 98(21): 12174-12179.
9. Wu G, Markowitz GS, Li L, *et al*. Cardiac defects and renal failure in mice with targeted mutations in Pkd2. *Nature Genetics* 2000; 24(1): 75-78.