

## Wolff-Parkinson-White and left ventricular noncompaction in a Fabry patient: A case report

### Fabry'li bir hastada Wolf-Parkinson-White ve sol ventrikül nonkompaksiyonu: Bir olgu sunumu

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**Summary**– Fabry disease is the second most common inherited (X-linked recessive) lysosomal storage disease associated with multiple organ involvement. Cardiac involvement of Fabry disease varies. Successful radiofrequency ablation of a Fabry disease patient with Wolff-Parkinson-White syndrome and left ventricular noncompaction is described in the present report.

**Özet**– Fabry hastalığı en sık ikinci kalıtsal (X'e bağlı resesif) lizozomal depo hastalığı olup birçok organ tutulumu ile ilişkilidir. Fabry hastalığının kalp tutulumu değişiklik göstermektedir. Bu yazıda, başarılı radyofrekans ablasyon yapılan Wolf-Parkinson-White sendromu ve sol ventrikül nonkompaksiyonu olan bir Fabry hastalıklı olgu sunuldu.

Fabry disease is the second most common inherited (X-linked recessive) lysosomal storage disease associated with accumulation of globotriaosylceramide (Gb3) in the vascular endothelium, myocardium, nervous system, kidney, and smooth muscle cells, due to deficiency and/or decrease of lysosomal enzyme  $\alpha$ -galactosidase A. Glycosphingolipid storage in the lysosomes leads to cellular dysfunction, ischemia, fibrosis, and organ failure. The kidney, heart, peripheral nerves, and skin are the organs most affected.<sup>[1]</sup>

Estimated incidence of the disease is 1:5000 male births. However, due to subclinical and/or late-variant phenotypes, incidence is likely to be much higher, even as high as 1:3100 male births.<sup>[2]</sup>

A Fabry disease patient with Wolff-Parkinson-White (WPW) syndrome and left ventricular (LV) noncompaction is described in the present report.

#### CASE REPORT

A 28-year-old woman with prior diagnosis of Fabry was referred due to ongoing palpitations and a syncope attack. The patient presented to the dermatology

clinic of a university hospital with multiple papules (angiokeratomas) on knees, lower back, buttocks, hips, and thighs (Figure 1). Meanwhile, the patient was suffering from acroparesthesia, and made several visits to the neurology department. The patient was diagnosed as a Fabry disease carrier with low levels of serum  $\alpha$ -galactosidase A, and was followed without treatment after a complete evaluation. She was referred to our tertiary cardiovascular hospital due to palpitations and a syncope attack. Rest-electrocardiogram (ECG) showed a short PR interval (<120 ms) and broad QRS complexes with a slurred upstroke to the complexes (the delta wave), which was diagnostic of WPW syndrome. Transthoracic echocardiography showed an ejection fraction of 65% with suspicion of LV noncompaction and grade 1 diastolic dysfunction. Cardiac magnetic resonance imaging (MRI) confirmed LV lateral noncompaction with a ratio of noncompacted to compacted of higher than 2.3 (Figure 2).

#### Abbreviations:

ECG	Electrocardiogram
LV	Left ventricular
MRI	Magnetic resonance imaging
WPW	Wolff-Parkinson-White

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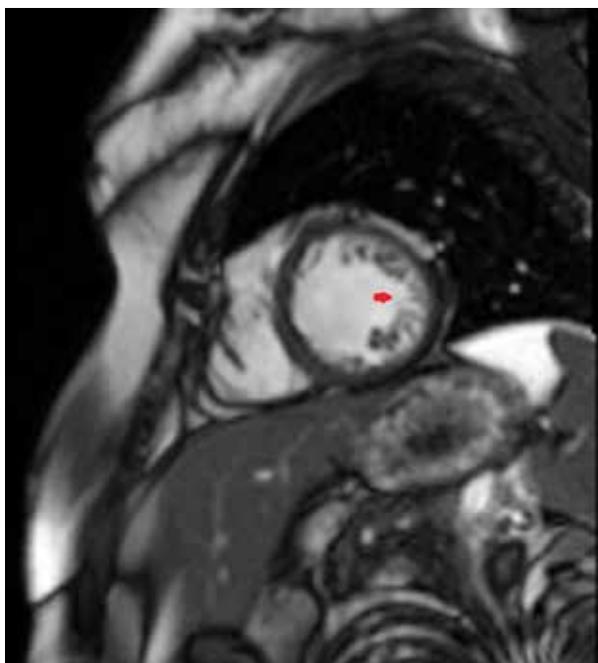
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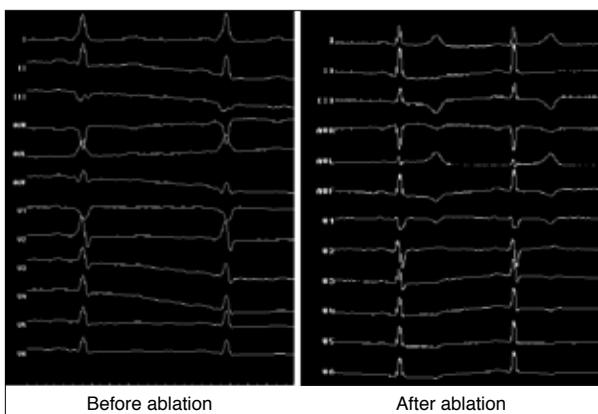




**Figure 1.** Multiple papules (angiokeratomas) were present on knees, hips.



**Figure 2.** Cardiac MRI was consistent with LV noncompaction.



**Figure 3.** Surface ECG performed just before ablation showed shortening of PR interval and delta waves. Surface ECG performed just after ablation showed recovering PR interval and delta waves.

Electrophysiological study was performed due to syncope attack and diagnosis of WPW on ECG. Pre-excitation was present at the beginning of the study. Atrioventricular reciprocating tachycardia was induced by stimulation from right ventricular apex. Refractory period of antegrade accessory pathway conduction was 270 ms from coronary sinus stimulations. The accessory pathway was located in the para-Hisian region after mapping around tricuspid valve. Successful radiofrequency ablation was performed (averages of 57°C heat and 30 Watts, 4 times, approximately 4 min total duration) on the accessory pathway. No recurrence was observed. Surface ECG showed disappearance of delta wave and PR shortening (Figure 3). The patient has been symptom-free for approximately 2 years.

## DISCUSSION

Concomitant WPW syndrome and Fabry disease have been reported in 1 male and 2 female patients.<sup>[3]</sup> The present is believed to be the first report to describe successful radiofrequency ablation of a Fabry disease patient with WPW syndrome.

Short PR (<0.12 ms) interval has been reported as high as 21%–40%.<sup>[4]</sup> In spite of the use of the term ‘pre-excitation’ to describe the ECG finding of Fabry disease, no electrophysiological studies have described a bypass tract from atrium to ventricle in a Fabry patient.<sup>[4–6]</sup> In the present case, a real accessory pathway of pre-excitation of WPW syndrome was located in the para-Hisian region. Following successful radiofrequency ablation of accessory pathway, prolongation of PR (>120 ms) and evanescence of delta wave were observed on surface ECG.

LV noncompaction is thought to arise from the arrest of normal myocardial compaction during embryogenesis.<sup>[7]</sup> Diagnosis is based on echocardiography criteria of Jenni<sup>[8]</sup> and MRI finding of noncompacted layer to compacted layer ratio. However, the present case did not fulfill echocardiograph parameters of LV noncompaction, and diagnosis was made based on noncompacted to compacted ratio of higher than 2.3. Azevedo et al. described a case of Fabry disease with LV noncompaction, and concluded that LV noncompaction could be a morphologic expression of different diseases rather than a true, distinct cardiomyopathy.<sup>[9]</sup>

To our knowledge, the present is the first report to describe successful radiofrequency ablation in a patient with Fabry disease, WPW syndrome, and LV noncompaction. Fabry disease and WPW syndrome were considered to be distinct diseases in the patient, and LV noncompaction was considered to be a morphologic expression of Fabry cardiomyopathy.

**Conflict-of-interest issues regarding the authorship or article: None declared.**

## REFERENCES

- Zarate YA, Hopkin RJ. Fabry's disease. Lancet 2008;372:1427–35. [CrossRef](#)
- Spada M, Pagliardini S, Yasuda M, Tukul T, Thiagarajan G, Sakuraba H, et al. High incidence of later-onset fabry disease revealed by newborn screening. Am J Hum Genet 2006;79:31–40. [CrossRef](#)
- San Román-Monserrat I, Moreno-Flores V, López-Cuenca D, Rodríguez-González-Herrero E, Guillén-Navarro E, Rodríguez-González-Herrero B, et al. Comprehensive clinical evaluation of a large Spanish family with Anderson-Fabry disease, novel GLA mutation and severe cardiac phenotype. Med Clin (Barc) 2014;142:497–504. [CrossRef](#)
- Jastrzebski M, Bacior B, Dimitrow PP, Kawecka-Jaszcz K. Electrophysiological study in a patient with Fabry disease and a short PQ interval. Europace 2006;8:1045–7. [CrossRef](#)
- Shah JS, Hughes DA, Sachdev B, Tome M, Ward D, Lee P, et al. Prevalence and clinical significance of cardiac arrhythmia in Anderson-Fabry disease. Am J Cardiol 2005;96:842–6.
- Pochis WT, Litzow JT, King BG, Kenny D. Electrophysiologic findings in Fabry's disease with a short PR interval. Am J Cardiol 1994;74:203–4. [CrossRef](#)
- Sen-Chowdhry S, McKenna WJ. Left ventricular noncompaction and cardiomyopathy: cause, contributor, or epiphenomenon? Curr Opin Cardiol 2008;23:171–5. [CrossRef](#)
- Jenni R, Oechslin E, Schneider J, Attenhofer Jost C, Kaufmann PA. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. Heart 2001;86:666–71. [CrossRef](#)
- Azevedo O, Gaspar P, Sá Miranda C, Cunha D, Medeiros R, Lourenço A. Left ventricular noncompaction in a patient with Fabry disease: overdiagnosis, morphological manifestation of Fabry disease or two unrelated rare conditions in the same patient? Cardiology 2011;119:155–9. [CrossRef](#)

**Keywords:** Catheter ablation; Fabry disease; left ventricle noncompaction; Wolf-Parkinson-White syndrome.

**Anahtar sözcükler:** Katater ablasyonu; Fabry hastalığı; sol ventrikül nonkompaksiyonu; Wolf-Parkinson-White sendromu.