sinuses, nasal cavity, orbit, soft tissues, and respiratory system. Less common affected extranodal sites are the central nervous system, urogenital, and gastrointestinal system (3, 4). Cardiac involvement of RDD is seen in <1% and there were 19 reported cases in the literature, out of which, 3 were reported in children (5). In adults, cardiac involvement is most often seen in the right atrium (n=7), followed by left atrium (n=3), epicardium (n=4), left ventricle (n=3), and pulmonary artery (n=2). In childhood cases, interatrial septum (n=2) and tricuspid as well as pulmonary valve (n=1) are involved as the extranodal sites (6).

Etiology of RDD is still unknown with several possible mechanisms proposed such as disorder of immune regulation or viral infections (Herpes virus-6, Ebstein–Barr virus, cytomegalovirus) (4). Diagnosis of RDD is based on the pathological assessment. In immunohistochemical analysis, there is a positive reactivity of histiocytic cells with lymphocytes and macrophages due to emperipolesis, which is the abnormal phagocytosis of the autologous lymphocytes by histiocytes (1). In our case, the biopsy results verified the diagnosis of RDD in the parotid gland. We could not perform a biopsy of the heart mass because the patient refused surgery or any invasive treatment. According to the assessment of the MRI results, the mass in the heart had properties similar to the masses in the mediastinum and parotid gland.

Most of the patients with RDD have a stable disease; however, some of them have an aggressive pattern with the involvement of the extranodal sites. Generalised lymphadenopathy is a poor prognostic factor for RDD. Prognosis of RDD with cardiac involvement is largely unknown because of the limited cases. Treatment options are surgical excision, radiotherapy, corticosteroids, and chemotherapy. For patients with resectable lesions, surgical excision is a well-established treatment option with good survival. Besides this, O'Gallagher et al. (3) reported a patient with an intracardiac mass of RDD who was treated with corticosteroids, which led to little change in the size of the mass.

Conclusion

In conclusion, we presented RDD with right ventricle, parotid gland, and mediastinum involvement. At 1 year follow up, the patient was asymptomatic and did not undergo any treatment such as radiotherapy, corticosteroids, and chemotherapy. Although cardiac involvement of RDD is a rare condition, it should be considered in patients who have been diagnosed with cardiac mass. However, because of the limited number of cases in the literature, the prognosis and treatment modalities with cardiac involvement are largely unknown.

Informed consent: An informed consent was obtained from the patient.

Video 1. Right ventricle mass was seen in the apical 4-chamber view.

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Successful right anteroseptal manifest accessory pathway cryoablation in a six-month infant with dyssynchrony-induced dilated cardiomyopathy

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Introduction

Ventricular preexcitation via an accessory pathway (AP) results in an asynchronous spread of ventricular depolarization that may lead to an abnormal regional wall motion and systolic dysfunction in patients with Wolff–Parkinson–White (WPW) syndrome, even without documented supraventricular tachycardia (1-3). It is known that adults and children with preexcitation-associated cardiomyopathy can completely recover following resynchronization

therapy (3-5). However, in infants, a very small number of patients have been treated with resynchronization via catheter ablation and only one case with less than 6 months of radiofrequency ablation was found in the current literature (5, 6). We present our report of one of the youngest patients in the literature, who showed rapid and complete recovery of severe ventricular dyssynchrony and myocardial dysfunction after cryoablation of the right-sided manifest AP. Our case emphasizes that cryoablation can be preferred as a safe method for AP ablation in infants as well.

Case Report

A previously healthy 5-month-old girl was referred to our hospital for a heart murmur. She weighed 8 kg and the physical examination revealed a mild, grade 1-2/6 heart murmur, mild tachypnea, and hepatomegaly. There were typical ventricular preexcitation signs like short PR intervals, wide QRS complexes (130 ms), and delta waves on a 12-lead surface ECG, which were suggestive of an anteroseptal manifest AP (Fig. 1a). The echocardiographic examination showed left ventricular enlargement, dyskinetic movement of basilar septum characterized with rightward systolic bulging, and impaired heart function with a left ventricular end-diastolic diameter of 40 mm (z-score 4.4) and a left ventricular ejection fraction of 34% as calculated by Simpson's method (Fig. 1b). The Holter examination showed no dysrhythmic events. After all the results were thoroughly evaluated, we considered that the LV dyssynchrony caused by preexcitation due to the right anteroseptal AP may have lead to her LV dysfunction. In accordance with the further echocardiographic investigations, measurements for the interventricular mechanical delay (IVMD) and intraventricular septal-to-posterior wall motion delay (SPWMD) were 74 ms and 290 ms respectively, which were consistent with dyssynchrony (Fig. 1c). Due to the confirmed presence of symptomatic dyssynchrony-induced dilated cardiomyopathy, we perfomed an electrophysiologic study.

The patient was intubated and the electrophysiologic study was performed under general anesthesia. The right and left femoral veins were catheterized and an esophageal catheter was also used. Three-dimensional (3-D) mapping (EnSite Velocity system; St. Jude Medical, St. Paul, MN, USA) and limited fluoroscopy were used together for delta mapping, and the earliest location was found within –38 milliseconds in the right anteroseptal/parahisian region. A 6 mm cryocatheter was used for ablation and at the 4th second of the first cryomapping, the AP disappeared (Fig. 2). Four complete cryo-lesions at –80 oC were given at this location/target. There was no complication during the procedure, except for right bundle branch block. After 72 hours post-ablation, the basilar septal hypokinesia and left ventricular function were markedly improved with a shortening fraction of 27%. At the 9-month follow-up, left ventricular functions and dyssynchrony measurements were found to have improved (LV ejection fraction 69%; IVMD=19 ms and SPWMD=5 ms; Fig. 3).

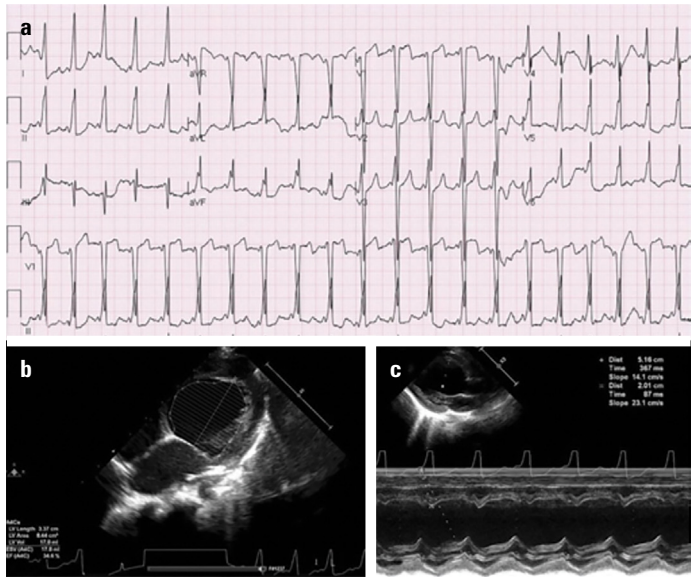

Figure 1. (a) 12-lead surface ECG recorded on admission showing marked delta waves consistent with an anteroseptal manifest AP (WPW preexcitation) (b) Decreased LV systolic function with an EF of 34.6% (measured with Simpson's method in an apical four chamber view) (c) M-Mode echocardiography image showing marked dyssynchrony caused by the ventricular preexcitation of the anteroseptal AP and dyssynchrony measurements

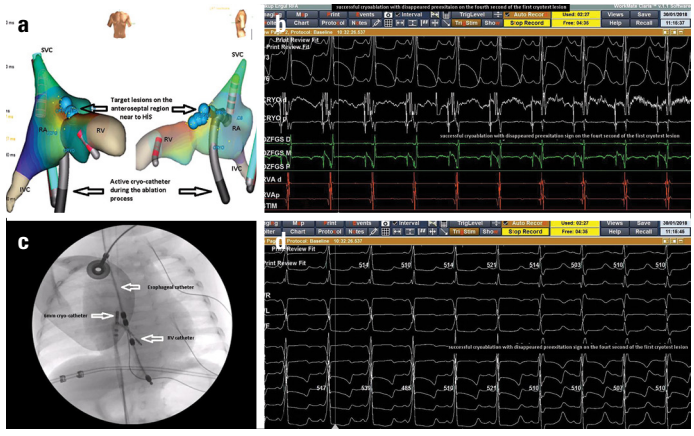

Figure 2. (a) Three-dimensional anatomy of the right atrium and ventricle, with blue dots showing targets on the anteroseptal region and diagnostic catheters in the high right atrium (HRA), esophagus (OZFGS), right ventricle (RVA), and the 6 mm cryocatheter active in the middle. (c) Fluoroscopic image in the left 300 oblique position, with the two diagnostic catheters in the esophagus and the RV and the 6 mm cryocatheter in the anteroseptal region. (b and d) Intracardiac and surface 12-lead electrograms recorded during successful cryoablation showing the disappearance of the preexcitation sign in the 4th second of the first cryotest lesion

IVC - inferior vena cava, RA - right atrium, RV - right ventricle, SVC - superior vena cava

Discussion

The possibility of a causal relationship between ventricular preexcitation and cardiomyopathy without tachycardia in patients with WPW syndrome was first described in a 67-year-old male patient with a diagnosis of WPW in 1998, who showed sig-

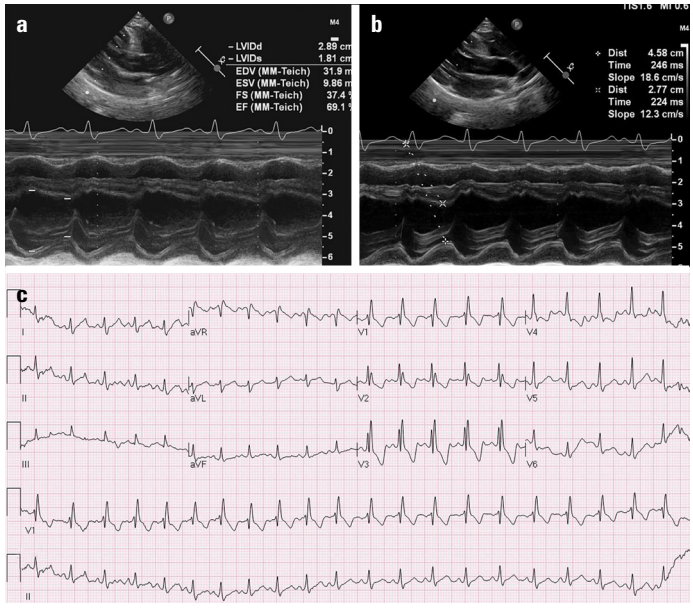

Figure 3. (a) M-mode echocardiography image in parasternal long-axis view showing improved left ventricular systolic function with an EF of 69%. (b) M-mode echocardiography image in parasternal long-axis view showing improved dyssynchrony measures. (c) 12-lead surface ECG, showing no ventricular preexcitation sign and right bundle branch block after successful ablation of the AP

nificant improvement in myocardial function after ablation (7). In 2004, Emmel et al. (8) reported the first such cases in children, where four patients aged 12–156 months of age range were diagnosed with WPW and were considered as having non-tachycardia-induced cardiomyopathy with a left ventricular shortening fraction of 13%–25%. In these cases, a significant improvement in ventricular function followed by a loss of preexcitation with spontaneous or radiofrequency catheter ablation suggests that ventricular preexcitation plays a role in cardiomyopathy without tachycardia (8). Further, the AP located in the right septal region with a longer QRS duration has been reported to be strongly associated with abnormal wall motion and septal dyskinesia both in children and adults (3, 9). In the present case, similar to previous reports, the location of the AP was in the right anteroseptal region and the QRS duration was 130 ms, which was consistent with left bundle branch block (LBBB) morphology.

Abadir et al. (4) reported a significant improvement in dyssynchrony and left ventricular systolic function after catheter ablation of the AP in 16 children aged 14.2±3.7 years. However, in infants, successful radiofrequency ablation has been reported only in two cases, one patient aged 4.5 months and another aged 16 months (5, 6). Furthermore, spontaneous termination of accessory route transmission and pharmacological suppression was reported in a few WPW-diagnosed infants with significant dyssynchrony and cardiomyopathy in the literature (8, 10). Inadequate catheter ablation experience and the increased risk of complications due to low body weight have led to the pharmacological suppression of the AP in infants. Nevertheless, experienced clinicians prefer catheter ablation instead of amiodaron due to the knowledge of drug-resistant cases, frequent relapses of the symptoms of preexcitation, dysfunction following discontinuation of the drug, and long-term drug use causing severe side effects (5, 6). Kwon et al. (6) reported a successful radiofrequency catheter ablation in their 4.5-month-old patient, whose left ventricular function progressively deteriorated and was unresponsive to amiodarone despite the gradually increased dose. This was the first patient aged less than 6 months who was treated for cardiomyopathy caused by WPW-related dyssynchrony in the literature (6). Five years after this, Wu et al. (5) reported that improvement in cardiac functions after radiofrequency ablation began in the second year and full recovery was achieved after 3.5 years in a 16-month-old infant with WPW. Similarly, we preferred catheter ablation to suppress the right anteroseptal AP in our mildly symptomatic 6 month-old patient. In addition to the increased risk of AV block due to radiofrequency ablation in infants and young children, cryoablation was preferred instead of radiofrequency considering the parahisian localization of the AP and preexcitation was successfully suppressed. Unlike Wu et al. (5), we observed improvement in cardiac functions and decrease in dyssynchrony on the third day following ablation.

Conclusion

In conclusion, ventricular preexcitation due to APs, especially those located in the right septal and paraseptal area, may cause ventricular dyssynchrony and cardiomyopathy in infants younger than 6 months of age. Therefore, WPW syndrome should be considered as a reason for preexcitation in infants whose cardiomyopathy is investigated without tachycardia, especially in the presence of prolonged QRS and LBBB morphology. Additionally, with cryoablation performed by the experienced clinicians, there is a chance of successful and safe ablation of the AP and complete restoration of cardiac function in infants younger than 6 months of age.

Informed consent: Written informed consent was obtained from the parents.

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Importance of endomyocardial biopsy in patients with myocarditis: A case report

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Introduction

Endomyocardial biopsy (EMB) is an invasive diagnostic tool to clarify specific etiologies of dilated cardiomyopathy with unknown etiology and suspected infiltrative cardiomyopathy. Recent case series indicate that EMB is a unique method for the diagnosis of myocarditis in >30% of unexplained cardiomyopathy cases (1). Differentiating specific types of myocarditis and infiltrative disease with EMB can lead to decide the appropriate therapy earlier and to improve the poor diagnosis of these diseases by this way. Based on the 2016 heart failure guidelines, EMB should be considered in individuals with rapidly progressive heart failure (HF) despite standard therapy, when there is a probability of a specific diagnosis which can be confirmed only in myocardial samples and specific treatment is available and effective (Class IIa and Level C).

Case Report

A 28-year-old male patient presented to our emergency room with exertional dyspnea. His complaint existed 3 months ago and increased in time. There was no abnormal finding on his medical history and electrocardiogram. He had Class II HF symptoms and signs according to the New York Heart Association (NYHA). First, bolus intravenous loop diuretics and bronchodilator treatment via respiratory mask were applied. His transthoracic echocardiogram (TTE) examination revealed global hypokinetic left ventricular function. His left ventricular ejection fraction (LVEF), left ventricular global longitudinal strain (LV-GLS), and tricuspid annular plane systolic excursion were calculated as 26%, –6.8 cm, and 1.4, respectively (Video 1). He was hospitalized, and routine HF medication was given to the patient according to his monitorization and physical examination results. Coronary angiography demonstrated normal coronary artery flow. His peak pulmonary artery pressure was 60 mm Hg on TTE, and it was confirmed by right heart catheterization later. A 24-hour rhythm Holter examination showed short-term non-sustained ventricular tachycardia, and implantable cardioverter defibrillator implantation was planned. Despite ramipril 5 mg once a day, metoprolol 25 mg twice a day, spironolactone 50 mg once a day oral treatment, and furosemide 40 mg four times a day intravenous treatment, his HF symptoms were progressive. TTE and fluoroscopy-guided EMB were performed to diagnose the etiology of dilated cardiomyopathy. Nine cardiac biopsy specimens were obtained and sent to the Institut Kardiale Diagnostik und Therapie center. Extensive inflammation without acute myocarditis was found. CD4 T cells, LFA-1 cells, and CD45R0 were calculated as 14.43 mm^2 (>10 cells/mm^2), 28.5 mm^2 (>14 cells/mm^2), and 71.95 mm^2 (>40 cells/mm^2), respectively. Before starting the immunosuppressive treatment, atrial fibrillation was revealed. After intravenous amiodarone 1200 mg treatment, oral medication was administered as 200 mg twice a day. After 10 days, his transaminase levels increased to 10 times due to amiodarone toxicity. A combination of prednisone 1 mg/kg/day tapered biweekly 10 mg and azathioprine 100 mg/day was started when transaminases decreased to normal range after amiodarone cessation. TTE was repeated on week 2 of treatment before discharge, and there was no remarkable