Extensive aortic aneurysm associated with neonatal lupus erythematosus

Sir,

A preterm male neonate, weighing 1,350 g was delivered at our center to a 25-year-old *primigravida* at the 32nd gestational week. The mother did not have any history of connective tissue disorder, smoking, alcohol abuse, or substance abuse. On initial physical examination, the neonate had bradycardia (50–55 beats/min), grunting, subcostal retraction, and cyanosis and, therefore, was intubated and mechanical ventilation and surfactant therapy were initiated. Cardiac examination revealed grade III/VI systolic murmur and electrocardiogram showed complete heart block (CHB). On the second postnatal day, echocardiography showed *patent ductus arteriosus* (PDA), and dilatation of the ascending aorta and the aortic arch [Figures 1 and 2] without any structural abnormalities or effusions. The patient received ibuprofen and isoproterenol for treatment of the PDA and bradycardia, respectively. The mother underwent rheumatologic investigations that showed positive serum anti-double stranded deoxyribonucleic acid (anti-dsDNA) and anti-Ro/Sjögren's-syndrome-related antigen A (anti-Ro/SSA) antibodies, thus, the diagnosis of neonatal lupus erythematosus (NLE) was made. Isoproterenol was discontinued gradually, while the heart rate remained at 60–65 beats/min. The patient was followed with serial echocardiographic studies that showed the progression of aortic dilatation over time and persistent bradycardia. Therefore, after 1 month, the infant was referred for inserting a dual-chamber pacemaker. After normalizing the cardiac rhythm on follow-up echocardiographic studies, the progression of aortic dilatation disappeared and interestingly, the aortic diameters gradually regressed.

Aortic dilatation is extremely rare in the neonatal period and was only reported in Marfan syndrome, Ehlers-Danlos syndrome, and congenital cardiac malformations.[1] For the first time, Radbill *et al.* in 2008 and later Davey *et al.* (2011) and Altit *et al.* (2014) in retrospective studies showed that more than 50% of children with congenital CHB had dilatation of the ascending aorta on the initial echocardiogram that was associated with maternal autoantibody seropositivity.[2-4] Aneurysm formation in neonates with NLE is proposed to be through the inflammation in the aortic adventitia induced by transplacental passage of maternal autoantibodies.[2-4] This inflammatory process compromises the integrity of the aortic wall, making it fragile and susceptible to aneurysm formation.[2-4] Because all reported neonates had congenital CHB, it might be concluded that the same inflammatory processes underly alterations in the conduction tissue and fetal aortic adventitia.[2,3]

The progression of aortic dilatation, as seen in our case, could be due to the slow heart rate, necessitating a larger stroke volume, thus increasing the expansile stress on the fragile aortic wall.[2,3] Normalization of the heart rate in these neonates seems to be a possibly useful strategy. In our case, after pacemaker insertion and normalization of cardiac rhythm, the aortic dilatation started to regress; similarly, in the study of Altit *et al.*, of 17 patients with CHB, aortic dimensions regressed after pacemaker

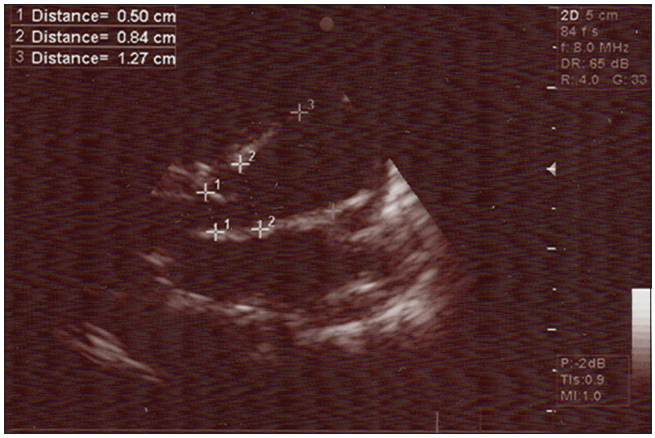

Figure 1: Long-axis echocardiography view showing the 1. Aortic annulus (Z-score: -1.05), 2. Aortic sinuses (Z-score: 0.53), and 3. Dilated sinotubular junction (Z-score: 5.36) (Normal diameters based on body surface area: Aortic Annulus: 0.47–0.65, Sinuses: 0.66–0.95, Sinotubular junction: 0.50–0.77)

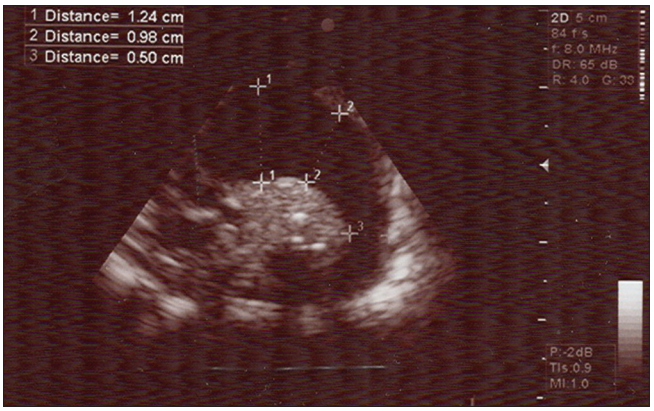

Figure 2: Suprasternal echocardiography view showing the 1. Dilated ascending aorta (Z-score: 5.18), 2. Dilated transverse arch (Z-score: 3.01), and 3. Isthmus (Z-score: 0.67) (Normal diameters based on body surface area: Ascending aorta: 0.50–0.77, Transverse aortic arch: 0.48–0.80, Isthmus: 0.34–0.59)

insertion.[4] Surgery or endovascular treatments are rarely required in the management of these cases and only indicated when the aneurysm presents with signs of dissection or compresses other thoracic structures. Close follow-up with serial echocardiographic studies with special attention to aortic diameters and early normalization of heart rate are the recommended approaches in these cases.[2-4]

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Conflicts of interest

There are no conflicts of interest.

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