Left ventricular aneurysm in a patient with Duchenne muscular dystrophy

Anévrysme du ventricule gauche chez un patient atteint de myopathie de Duchenne

Duchenne muscular dystrophy (DMD) is an X-linked recessive disorder that affects young patients. Heart failure is a classical complication in this disease. We report an asymptomatic infero-basal aneurysm of the left ventricle (LV) in a DMD patient.

Case report

A 38-year-old patient, on wheelchair, was admitted in hospital for routine explorations because of DMD. His medical history was pertinent for DMD, tracheotomy for full mechanical ventilation because of respiratory restrictive insufficiency, vertebral arthrodesis for scoliosis and subclinical cardiomyopathy. He took daily bisoprolol 1.25 mg. On admission, body temperature was 37.8°C; blood pressure 105/70 mmHg and heart rate 80 beats per minute (bpm). Cardiac and pulmonary auscultation was normal. Biological results were without any particularities. Electrocardiography disclosed sinus rhythm at 80 bpm, tall R waves at V1V2, normal axis and incomplete bundle right branch block. Transthoracic echocardiography (panel A) depicted normal valves, LV dilation (LV end diastolic diameter = 63 mm), alteration of the left ventricular ejection fraction (LV EF = 30%) with aneurysm located at the infero-basal of the LV (figure 1). A conservative treatment was decided and an angiotensin-converting enzyme (ACE) inhibitor was introduced in addition to a beta-blocker.

Discussion

DMD is an X-linked recessive disorder that affects 1 of 3500 male births [1]. This disease is caused by the absence of dystrophin, a protein located on the inner side of the skeletal and the cardiac muscle cells. This protein has a major structural role in muscle, as it links the internal cytoskeleton to the extracellular matrix. The lack of dystrophin leads to progressive fibre damage and membrane leakage. The consequence is a progressive muscle wasting and weakness of variable distribution and severity [1].

Cardiac involvement is present in about 90% of the patients and leads to heart failure. The posterior wall of the LV is often affected in patients with DMD and electrocardiograms disclosed usually tall R waves in V1 and deep Q waves in infero-lateral leads. Heart failure is caused by loss of cardiomyocytes because of dystrophin lack. Connective tissue and lipid infiltrates replace the loss of cardiomyocytes. This phenomenon leads to an increase of the LV wall stress and wall motion abnormalities [2]. LV aneurysm has been reported by others [3]. LV aneurysm is a consequence of the LV wall motion failure in the infero-basal segment. Medical management in patients with DMD and heart failure relies mainly on angiotensin-converting enzyme inhibitors and beta-blockers [1]. Cardiovascular surgery therapy is not indicated in asymptomatic DMD patients with LV aneurysm. Management of LV aneurysm in this disease relies mainly on medical therapy [3].

Conclusion

DMD is a genetic neuromuscular disorder, which affects heart leading to heart failure. LV aneurysm is rare in this disease and can be detected using echocardiography.

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References


Figure 1

Echocardiography (para sternal long axis view)

Left ventricular (LV) infero-basal aneurysm in patient with DMD dystrophy.

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