Noncompaction cardiomyopathy is frequently associated with myopathy (Left ventricular noncompaction with hypothyroidism and sensorineural hearing loss)

``Noncompaction`` kardiyomiyopati sıklıkla miyopati ile beraber görülür (Hipotiroidi ve sensörinöral işitme kaybı ile birlikte olan sol ventrikül kökenli "noncompactıon")

Dear Editor

With interest we read the article by Sahin et al. on a 19 years old woman with left ventricular hypertrabeculation/noncompaction (LVHT) in both ventricles associated with hypothyroidism and sensorineural hearing loss (1). The report evokes the following remarks.

Despite intensive research, the pathogenesis of LVHT is still unknown. In case of congenital LVHT the non-compaction theory is quite plausible (2), but how to explain LVHT, which definitively occurred during adulthood? In these cases the noncompaction theory is not applicable and LVHT is assumed to result from: a compensatory attempt of the impaired myocardium to eject physiologic stroke volumes by reduced contractility but enlarged surface; dissection of the endo- and myocardium due to mal-functioning gap junctions; penetration of persisting sinusoids into the left ventricular cavity and transformation into trabeculations; a frustrate attempt to hypertrophy an insufficiently contracting myocardium; an enlargement of the endocardial layer for improved oxygenation via the endocardium; an attempt of the impaired myocardium to resist against an impeding dilatation by tightening the myocardial structure. Which of these explanations holds true, however, is under debate.

The authors correctly mention that LVHT is often associated with multi-system disease. One of the most frequently involved systems is the peripheral nervous system. This is why we repeatedly claimed that each LVHT patient should also be seen by a neurologist. It would be interesting to know if the presented patient underwent a neurological investigation and if there were any indications for a neuromuscular disorder. Of particular interest is if there were signs of double vision, ptosis, limb weakness, exercise-induced muscle soreness, easy fatigability, muscle cramps, or sensory disturbances. So far, LVHT has been described together with dystrophinopathies, dystrobevinopathy, laminopathy, zaspopathy, centronuclear myopathy, myotonic dystrophy, myoadenylate-deaminase-deficiency, mitochondriopathy, Pompe's disease, Barth syndrome, Friedreich ataxia, and hereditary neuropathy. In a study on 62 LVHT-patients 82% had a specific or non-specific neuromuscular disorder (3). The authors should also provide data if there was affection of the endocrinological system, eyes, kidneys, gastrointestinal tract, or bone marrow. The combination of thyroid dysfunction, impaired hearing, and LVHT suggests hereditary disease like amyloidosis, mitochondrial disorder or Down syndrome.

Though the authors mention that LVHT occurs familiarly they don't provide any information about the parents and other relatives of their patient. Were the parents consanguineous? Was either of them affected by a neurological disease or did either of the two suffer from a multisystem disease? Were any of the relatives investigated for heart disease, particularly LVHT? Familial LVHT has been described in patients with myotonic dystrophy and metabolic myopathy (4).

Though frequently described as a risk of LVHT, systemic embolism does not appear to be a major complication of LVHT. In a recent study on 62 LVHT-patients thromboembolic events were found in only 10% of them as compared to 15% in age-sex, and left ventricular-function-matched controls (5).

LVHT-patients not only require extensive cardiac investigations but also comprehensive neurological investigations including regular follow-ups given the fact that a high number of these patients suffer from a neuromuscular disorder. In reverse, patients with a neuromuscular disorder require a comprehensive cardiological investigation.

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Author's Reply

Dear Editor

Our patient was seen by neurologist and was found to have only sensorineural hearing loss. Our patient's parents were examined with echocardiography and no sings of noncompaction of their hearts were revealed.

Thanks to our colleagues for contributions.

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