

Histopathological study of the cardiac conduction system in systemic lupus erythematosus

Systemic lupus erythematosus (SLE) is a chronic multisystem inflammatory connective disease characterized by the production of auto-antibodies and immuno-complexes. SLE can affects all organs including heart.

Overall, the prevalence of cardiac involvement is estimated to affect more than 50% of SLE cases. All portions of the heart can be involved: pericardium, myocardium, cardiac conduction system, as well as coronary arteries. Pericarditis is the most common finding, while endocarditis is characterized by small nonbacterial vegetations along the valve leaflets known as Libman Sacks endocarditis. The involvement of the cardiac conduction system in SLE has been less commonly described but should always be taken into account.

SLE affects particularly young women and the passive acquisition of maternal IgG antibodies during pregnancy cause neonatal lupus, which is often related to congenital heart block.

Pre- or perinatal death from heart block due to severe autoimmune lesions of the atrioventricular junction has been reported with emphasis to the possible lethal association between maternal auto-antibodies and QT-prolongation. [1] Recently, we reported a case of sudden unexpected intrauterine death of a term fetus in a anti-cardiolipin positive mother. [2] The findings of the postmortem examination including the study of the cardiac conduction system and brainstem on serial sections ruled out the clinically suspected atrio-ventricular block due to the anti-cardiolipin antibodies, and disclosed severe bilateral hypoplasia of the arcuate nucleus which is an important chemoreceptor center for the control of breathing activity, located on the medullary ventral surface. [3,4]

As the volume of data on new morphological and functional alterations of the cardiac conduction system increases, it becomes worldwide essential that victims of SLE, especially in cases of sudden deaths in young age, be submitted to an indepth necropsy examination, focusing particularly on the study of the cardiac conduction system on serial sections. [3,5] To examine the cardiac conduction system, two blocks of heart tissue should be obtained, for paraffin embedding. The first block contains the junction of superior vena cava and right atrium encompassing the entire area of the sino-atrial node. This sinoatrial block should be cut serially sectioned in a plane parallel to the crista terminalis. The second block contains the atrioventricular node (AV), His bundle down to bifurcation and bundle branches, with two centimeters of attached septum above and below. This AV junctional block is serially sectioned in a plane parallel to the two atrioventricular valve rings. All sections are to be cut serially at intervals of 40-mm (levels) and stained alternately with hematoxylin-eosin and trichromic Heidenhain. Should such an investigation not be feasible in the local facility, it is advisable that the hearts be preserved in buffered formalin and transported to a referral specialist center experienced in the study of cardiac conduction system.

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