Systemic lupus erythematosus (SLE), a complex autoimmune disease, often has confusing manifestations. Our case is that of a 45 year old female with a four year history of SLE complicated by end stage renal disease on hemodialysis, hypertension, and history of lupus myocarditis. Presenting with abdominal pain, nausea/vomiting, weakness, and palpitations, she was found to have elevated troponins, elevated b-type natriuretic peptide, and heart block, for which a transcutaneous pacemaker was placed. Because of evidence of persistent myocardial inflammation, permanent pacemaker placement was delayed until the course of IVIG for presumptive lupus vs viral myocarditis could be completed. Despite the transcutaneous pacemaker, she developed intermittent high degree atrioventricular heart block and became progressively hypotensive. As the clinical team was preparing to initiate inotropic support, she had an asystolic episode and could not be revived. Given clinical question regarding the pacemaker’s lack of efficacy despite proper placement and function, an autopsy was requested.

The electronic health record and radiographic/cardiac studies were reviewed. The autopsy was restricted to chest only. The post mortem time was 6 hours. A section of the left ventricle was taken for electron microscopy (EM).

Four months prior to death, while being worked up for renal transplant, her cardiac studies showed decreased ejection fraction, increased wall thickness, and diastolic dysfunction. At autopsy, cardiomegaly (750g; expected: 148-296g) with left ventricular hypertrophy (free wall, 2.1cm; expected: 1.0-1.5cm; interventricular septum, 1.9cm; expected: 1.2-1.6cm) was present. There was no evidence of coronary artery disease, acute or remote myocardial infarction, ischemic changes, vasculitis, or myocarditis. Microscopic examination of all four chambers of the heart and interventricular septum showed extensive intracellular vacuolar change. EM showed highly abnormal myocytes with sarcolemmas disrupted by amorphous material and curvilinear bodies.

The vacuolar myopathy was consistent with hydroxychloroquine cardiotoxicity. While considered in the differential diagnosis, her cardiac findings/symptoms were thought secondary to SLE or long standing hypertension, not from medications. The hydroxychloroquine was discontinued approximately one week prior to death; however, given its long half-life, the effect was likely minimal. Clinicians need to be reminded that while uncommon, chloroquine/hydroxychloroquine use may be complicated by cardiac toxicity.