A 67-year-old woman with rheumatoid arthritis (RA; rheumatic factor and autoantibodies against cyclic citrullinated peptide positive) diagnosed 21 years ago was referred to our hospital because of increasing dyspnea on exertion, fatigue, and dizziness. She had been taking chloroquine (250 mg/d) for 20 years and indomethacin (25 mg/d) for treatment of RA and had no current signs of disease activity. Apart from medically controlled arterial hypertension and chronic renal failure (glomerular filtration rate 39 mL/min) with secondary hyperparathyroidism (intact parathormone 370 ng/L), she had been otherwise healthy.

The ECG on admission revealed sinus rhythm with a complete right bundle-branch block. Holter monitoring documented intermittent sinus arrest (up to 3.7 seconds) and frequent symptomatic episodes of junctional escape rhythm of approximately 40/min. Elevated levels of serum creatine kinase (608 U/L; normal value: <140 U/L), creatine kinase-MB (33.6 g/L; normal value: <0.03 g/L), troponin T (0.40 µg/L; normal value: <0.03 µg/L) seen at admission remained constant within this range during the subsequent hospital stay. Echocardiography revealed concentric left ventricular hypertrophy (septum thickness 18 mm), normal systolic function, and mildly impaired diastolic left ventricular function. Coronary angiography demonstrated only minor coronary atherosclerosis. At right heart catheterization, moderate pulmonary hypertension [42/12 mm Hg at rest, 70/27 mm Hg at 3 minutes exercise (25 W)] was diagnosed, accompanied by a slight dip-plateau phenomenon in the right ventricular pressure curve. Contrast-enhanced cardiovascular magnetic resonance imaging after 0.2 mmol/kg intravenous gadolinium administration using an inversion-recovery pulse sequence revealed left ventricular hypertrophy and an area of delayed contrast enhancement within the basal septum at the insertion point of the right ventricle (Figure 1). Right ventricular endomyocardial biopsies demonstrated severe cytoplasmatic vacuolization of cardiomyocytes by light microscopy (Figure 2A) without signs of vasculitis, rheumatoid cardiac activity, inflammatory myocardial disease, or amyloidosis. Upon transmission electron microscopy, electron-dense material in concentric lamellar bodies evident within lysosomes was shown to be a characteristic feature (Figure 2B). Thus, chloroquine-induced cardiomyopathy was diagnosed with characteristic “pseudo-myeloid bodies,”1 typical alterations of the cardiac conduction system, and pulmonary hypertension. Consequently, chloroquine was discontinued and a dual chamber pacemaker was implanted.

Chloroquine-induced cardiomyopathy is a rare iatrogenic disease associated with long-term intake of chloroquine, which is most often used for RA or malaria prophylaxis. The time period between the start of chloroquine therapy and disease manifestation may range from several months to more than 20 years. To date, only 22 cases have been reported.2 Here, we included ce-CMR findings in the diagnostic work-up, showing left ventricular hypertrophy and areas of delayed contrast enhancement in the basal septum at the insertion point of the right ventricle, suggesting altered gadolinium kinetics. Of note, a similar distribution pattern has been described previously in patients with hypertrophic cardiomyopathy and patients with pulmonary hypertension, and may also be detected in other forms of cardiomyopathy. This case demonstrates that ce-CMR may be a valuable adjunct in the work-up of symptomatic patients with a medical history of longstanding chloroquine intake.

Disclosures
None.

References
Figure 1. Short- (left) and long-axis views (right) on cardiac magnetic resonance imaging, showing marked concentric left ventricular hypertrophy and delayed signal hyperenhancement after gadolinium administration within the basal anterior interventricular septum (arrow).

Figure 2. A, Light microscopy of right ventricular biopsy specimen showing extensive cytoplasmic vacuolization of cardiomyocytes without signs of inflammatory cardiac disease (hematoxylin and eosin staining; original magnification ×250). B, Transmission electron microscopy of the right ventricular biopsy specimen. Within the cardiomyocytes are concentric lamellar structures of electron-dense material within lysosomes, the so-called “pseudomyeloid bodies,” which are characteristic of chloroquine-induced cardiomyopathy. Left, Original magnification ×23400; right, original magnification ×5000.
Contrast-Enhanced Magnetic Resonance Imaging of a Patient With Chloroquine-Induced Cardiomyopathy Confirmed by Endomyocardial Biopsy
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