

Rumpel-Leede Sign: Consider Underlying Vitamin C Deficiency



To the Editor: I read with interest the article by Mohammad et al.¹ The authors report the case of a man in his 60s, in whom a petechial rash appeared on his arm after measuring his blood pressure early in his hospitalization. In the context of normal laboratory results, this finding was interpreted as an example of the Rumpel-Leede phenomenon (intracutaneous hemorrhage produced by prolonged proximal pressure, signifying increased capillary fragility). Given the specifics of this case (the patient had a history of intravenous drug use, retained needles in the forearm give a picture of self-neglect), consideration should be given to underlying vitamin C deficiency (scurvy), which can also be manifested in this manner.² More than a century ago, Rumpel and Leede described the “stasis test” in patients with scarlet fever. Shortly thereafter, Hess made a similar observation in scorbutic children (Hess test).³ As stated in this classic report, “The test is not specific for scurvy, but is a method of demonstrating a weakness of the vessel walls, whatsoever may be its cause.”^{2,3} The more generic term *tourniquet test* is also found in the medical literature, for instance, in the diagnosis of dengue fever.

This is not mere nitpicking about historical eponyms but has important practical consequences. Vitamin C deficiency is widely regarded as a disease of former times but has not vanished and is still found in certain groups at risk, including persons

with low socioeconomic status, such as individuals who use intravenous drugs.⁴ Whereas frank scurvy is rare in affluent societies, a study from the United Kingdom found a significant percentage of vitamin C deficiency in a materially deprived population. In this study, smoking and male sex were independent predictors of poor vitamin C status.⁵ Features of scurvy entail characteristic dermatologic manifestations (perifollicular purpura, corkscrew hair), gingivitis, and hemorrhage, which can be fatal. Capillary fragility due to impaired collagen synthesis is the underlying mechanism. Once diagnosed, it is easily cured by vitamin C substitution. Therefore, an early diagnosis of scurvy is of great importance. Could the authors elaborate if the patient was a smoker, if a nutritional history was obtained, and if other signs of vitamin C deficiency were actively looked for?

Physicians should remain vigilant for vitamin C deficiency and liberally obtain blood for ascorbic acid level when compatible skin findings are found in an appropriate context.

POTENTIAL COMPETING INTERESTS

The author reports no competing interests.

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In Reply—Rumpel- Leede Sign: Consider Underlying Vitamin C Deficiency



To the Editor: We thank Dr Martin Windpessl for his interest in our case of Rumpel-Leede sign. As was pointed out, the cutaneous manifestation of a petechial rash should also suggest vitamin C deficiency. On discovery of the rash, our team did include vitamin C deficiency as part of the differential, consulted with the dermatology service, and sent a vitamin C level that returned within normal limits. We regret not being able to elaborate on this work-up in our initial manuscript because of the word limit associated with submissions of this nature. Whereas consideration of vitamin C deficiency is certainly warranted, previous reports of cutaneous manifestations of vitamin C deficiency describe a diffuse, bilateral pattern of the rash.¹⁻³ Thus, in the setting of a normal vitamin C level and a sharply demarcated, unilateral, petechial rash in a patient with known risk factors for vascular fragility, we determined that this presentation was likely to be an occurrence of the Rumpel-Leede sign.

POTENTIAL COMPETING INTERESTS

The authors report no competing interests.

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NT-proBNP and High-Sensitivity Cardiac Troponin T Fail to Detect Cardiac Involvement in Erdheim-Chester Disease



To the Editor: Erdheim-Chester disease (ECD) is a rare non-Langerhans cell histiocytosis belonging to the L-group of the 2016 revised histiocytosis classification.¹ Although frequent and associated with clinical complications, cardiac involvement is underdiagnosed in ECD.² Cardiac magnetic resonance (CMR) imaging is the most robust way to detect cardiac involvement.³ However, access to this technique is limited. We evaluated the utility of B-type natriuretic peptides and troponin for the diagnosis of cardiac involvement in ECD.

We retrospectively included patients with a biopsy-proven diagnosis of ECD who were referred to the internal medicine department of a French tertiary care center or the hematology department of an Israeli tertiary care center and had undergone both CMR and assessment of B-type natriuretic peptide and troponin levels, at least once, between 2007 and 2019.

Cardiac magnetic resonance imaging was performed on a 1.5T scanner (Siemens Aera). All images were re-read in a blind fashion by an experienced radiologist (M.B.).

Cardiac involvement was defined as abnormal epicardial or pericardial enhancement or infiltration. Cardiac biomarker levels were determined at the physician's discretion, in the routine clinical care setting. Natriuretic peptide test assessment was performed using N-terminal pro-brain natriuretic peptide (NT-proBNP) at the French center (normal, <300 pg/mL) and brain natriuretic peptide (BNP) at the Israeli center (normal, <100 pg/mL). Troponin assessment was performed using high-sensitivity cardiac troponin (hs-cTnT) at the French center (normal, <14 µg/L) and troponin I at the Israeli center (normal, <0.04 µg/L). Only NT-proBNP and hs-cTnT were considered in quantitative analysis.

The study was approved by the appropriate ethics committee (*Comité de Protection des Personnes d'Ile de France III* [#2011-A00447-34]) and was conducted in accordance with the Declaration of Helsinki.

We included 122 patients in total (118 in France, 4 in Israel), with a mean age of 58.7 years (± 13.9 years). Most of the patients (70%) were male. *BRAF*^{V600E} mutations were detected in 75 patients (61.5%). Biologic assessment was performed a median of 12 months (1 to 33 months) after cardiac imaging. Coronary artery disease was present in 28 patients (23.1%). Mean left ventricular ejection fraction was 57% ($\pm 7\%$). Erdheim-Chester disease-related cardiac involvement was present in 57 patients (46.7%).

Median NT-proBNP concentration was 173 (73 to 470) pg/mL (normal, <300 pg/mL). Median NT-proBNP levels were similar between patients with and without right atrioventricular sulcus infiltration (195 vs 154 pg/mL; $P=.3$; **Figure A**). Area under the curve

was 0.68 for cardiac involvement detection using NT-proBNP (**Figure C**). With an optimal threshold of 114 pg/mL, NT-proBNP diagnostic performances were as follows: sensitivity, 0.79; specificity, 0.58; accuracy, 0.68; negative predictive value, 0.76; and positive predictive value, 0.62. The NT-proBNP/BNP levels were high in 38 (32.8%) patients. Cardiac involvement was detected on imaging in 22 (57.9%) of the patients with high NT-proBNP/BNP levels and in 30 (38.5%) of those with normal NT-proBNP/BNP levels ($P=.08$). High levels of NT-proBNP/BNP were associated with the presence of coronary artery disease ($P=.001$), atrial fibrillation ($P=.001$), older age ($P=.001$), and higher serum creatinine concentration ($P=.02$; **Table**).

Median hs-cTnT concentration was 9 (6 to 16) µg/L (normal, <14 µg/L). Median hs-cTnT levels were similar between patients with and without right atrioventricular sulcus infiltration (10.3 vs 9 µg/L; $P=.3$; **Figure B**). Area under the curve was 0.64 for cardiac involvement detection using hs-cTnT (**Figure D**). With an optimal threshold of 4.8 µg/L, hs-cTnT diagnostic performances were as follows: sensitivity, 0.96; specificity, 0.25; accuracy, 0.59; negative predictive value, 0.88; and positive predictive value, 0.54. Troponin levels were high in 39 patients (32%). Cardiac involvement was detected on imaging in 20 patients with high troponin levels (51.3%) and 37 (45.1%) patients with normal troponin levels ($P=.7$). High troponin levels were associated with the presence of atrial fibrillation ($P=.001$), older age ($P=.001$), and higher serum creatinine concentration ($P=.001$; **Table**).

In other infiltrative diseases, such as amyloidosis and Fabry disease, cardiac biomarkers have been