JACC: CASE REPORTS
PUBLISHED BY ELSEVIER ON BEHALF OF THE AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION.

VOL. 1, NO. 2, 2019

CASE REPORT

BEGINNER

CLINICAL CASE

Missing Vitamin C

A Case of Scorbutic Cardiac Tamponade

Saed Alnaimat, MD,[a] Abdullahi Oseni, MD,[b] Ying Yang, MD,[a] Vinesh Melvani, MD,[c] Adam Aronson, MD,[c] Katherine Harris, MD,[a] Sidakpal Panaich, MD[b]

ABSTRACT

Scurvy is a disorder of vitamin C deficiency which leads to vascular fragility and multisystemic complications. The paper presents a case of scurvy resulting in cardiac tamponade in addition to its classic manifestations. Life-threatening tamponade is an extreme presentation of scurvy and has not been reported in modern publications to the best of the present authors' knowledge. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2019;1:192–6) Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Scurvy has historically been known as a disease of voyagers since 1500 BC, but its clinical relevance in current practice is very limited. Scurvy is a disorder of vitamin C deficiency, which is essential for synthesis of collagen fibers. Substantial deficiency leads to disruption of connective tissue integrity and vascular fragility. Despite an excellent prognosis, the underlying cause of deficiency should be corrected to avoid recurrence.

HISTORY OF PRESENTATION

A 56-year-old male presented to a local hospital with exertional dyspnea for 2 weeks and spontaneous bruising of his left thigh. A computed tomography angiogram of his chest, which was obtained to exclude pulmonary embolism, demonstrated a large pericardial effusion (PE) compressing his right ventricle with flattening of right ventricular free wall, signifying cardiac tamponade (Figure 1).

On presentation, blood pressure was 97/77 mm Hg, heart rate was 101 beats/min, temperature was 99.1°F, respiratory rate was 24, and oxygen saturation was 98% on ambient air. He was malnourished and said that he had not eaten any fruit for many months. His chest was clear on auscultation. Heart sounds were normal with no murmurs. Jugular venous pressure was not elevated. Dental examination revealed loose teeth, extensive caries, receded gums, and severe periodontitis (Figure 2). He had nonblanchable petechiae, 2- to 3-mm perifollicular erythematous papules involving his trunk and lower extremities (Figure 3). There was an ecchymotic patch of the left medial thigh and scattered corkscrew hairs.

LEARNING OBJECTIVES

- To differentiate a diagnosis of pericardial effusion with a focus on laboratory workup and subtle clinical findings.
- To make clinicians aware of the uncommon but severe and potentially fatal complications of scurvy.

MEDICAL HISTORY

History included chronic alcoholism, gastroesophageal reflux disease, and esophageal stricture requiring multiple endoscopic dilations.

From the [a]Department of Internal Medicine, University of Iowa Hospitals and Clinics, Iowa City, Iowa; [b]Division of Cardiovascular Medicine, University of Iowa Hospitals and Clinics, Iowa City, Iowa; and the [c]Department of Dermatology, University of Iowa Hospitals and Clinics, Iowa City, Iowa. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

Manuscript received May 1, 2019; revised manuscript received June 23, 2019, accepted July 3, 2019.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2019.07.006

DIFFERENTIAL DIAGNOSIS

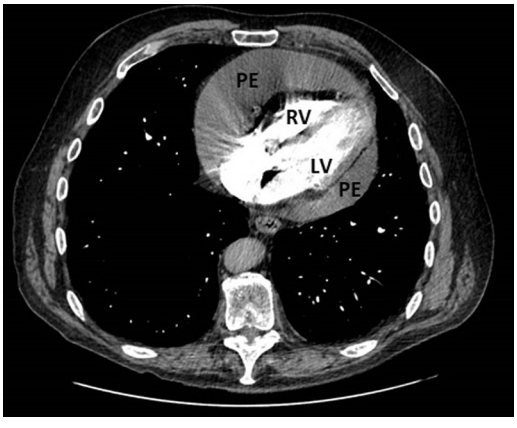
Differential diagnosis of PE is wide and includes idiopathic, infectious, malignant, autoimmune, uremic, postmyocardial infarction, and iatrogenic conditions.

INVESTIGATIONS

Transthoracic echocardiography confirmed the prior computed tomography finding of a large PE with right ventricular collapse during diastole (**Figure 4**, Video 1) and abnormal septal motion (**Figure 5**, Video 2). The patient underwent pericardiocentesis with drainage of 450 ml of bloody fluid.

Workup of pericardial fluid showed 2,000,000 red blood cells/mm^3; negative gram stain; negative bacterial, fungal and acid-fast bacillus cultures; negative cytology; and normal amylase. Prothrombin time was 1.3 with normal activated thromboplastin time. An esophagram revealed no esophageopericardial fistula. Autoimmune workup was negative for antinuclear antibody, rheumatoid factor, anticitrullinated protein antibody, SS-A, SS-B, P-ANCA, and C-ANCA antibodies, with normal C3 and C4. A punch biopsy of the left lower extremity demonstrated follicular hyperkeratosis, follicular plugging, twisted hair follicles, and perifollicular hemorrhage (**Figure 6**), consistent with scurvy. Plasma vitamin C concentration was <5 μmol/l, confirming the diagnosis of scurvy.

FIGURE 1 Large Pericardial Effusion Compressing the RV, With Signs of Cardiac Tamponade



LV = left ventricle; PE = pericardial effusion; RV = right ventricle.

MANAGEMENT

The patient's dyspnea largely resolved after pericardiocentesis. The patient was started on oral ascorbic acid supplementation (1,000 mg once daily) with substantial improvement of his petechiae and papules within a few days.

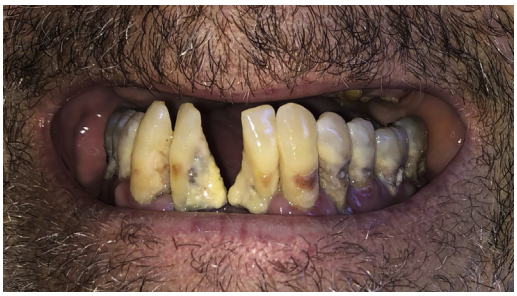
DISCUSSION

Scurvy has historically been described in the era of great maritime expeditions, as well as ancient Egyptian, Greek, and Roman literature. In 1536, an autopsy examination was performed by the French explorer Jacques Cartier, whose men were severely afflicted with scurvy. In the "Third and Last Volume of the Voyages" (p. 226, London, 1600), Richard Hakluyt included Cartier's vivid description of the gross anatomic examination: "That day Philip Rougemont, borne in Amboise, died, being 22 yeeres olde. He was found to have his heart white, but rotten, and more than quart of red water about it" (1).

In 1753, James Lind, a Scottish surgeon from the Royal Navy, published a hypothesis of treating and preventing scurvy during voyages by maintaining a diet rich in citrus fruits. After introduction of daily citrus consumption during voyages in the late 1700s, prevalence of scurvy decreased significantly in mariners.

Vitamin C allows hydroxylation of procollagen, which is crucial in collagen synthesis. Deficiency of vitamin C leads to impaired connective tissue integrity and fragility of blood vessels. Tissues with abundant collagen, such as mucous membranes and skin, are the most common sites of scurvy manifestations.

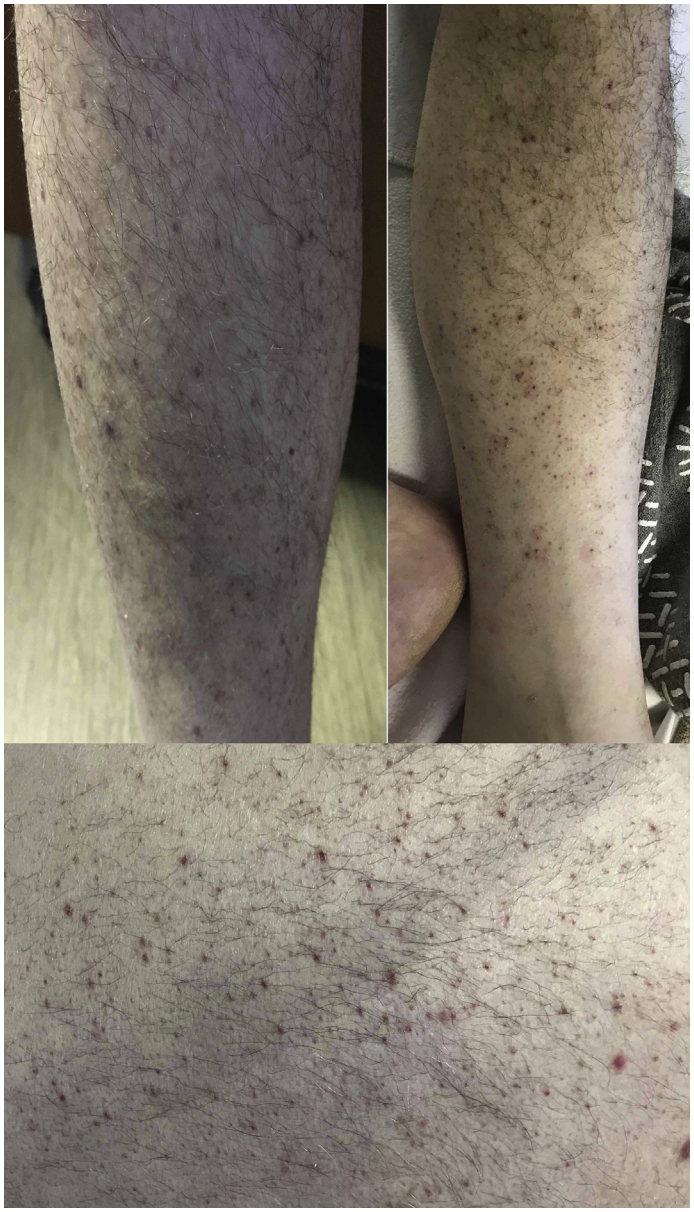
FIGURE 2 Severe Periodontitis With Receded Gums and Extensive Dental Caries



ABBREVIATIONS AND ACRONYMS

Ao = aorta
CT = computed tomography
GERD = gastroesophageal reflux disease
JVP = jugular venous pressure
LA = left atrium
LV = left Ventricle
PE = pericardial effusion
RA = right atrium
RV = right ventricle
TTE = transthoracic echocardiogram

FIGURE 3 Diffuse Petechiae and Erythematous Papules Over the Torso and Bilateral Lower Extremities



Note the presence of corkscrew hairs on the torso.

Humans lack enzymes to synthesize ascorbic acid and rely solely on exogenous sources. Because ascorbic acid is water soluble, it is not stored in the body. An average adult requires 75 to 90 mg of daily vitamin C (2). Risk factors for vitamin C deficiency include alcoholism, low socioeconomic status, senile age, restrictive eating habits, poor dentition, gastrointestinal diseases, and psychiatric disorders (3,4). Alcoholics represent one of the largest groups at risk for scurvy because of the associated malnutrition and because alcohol decreases the absorption of vitamin C (3). In a small study, alcohol consumption (0.58 g/kg) produced a 47% increase in urinary ascorbic acid excretion 4 h after ingestion. Another study of healthy men pretreated with high doses of vitamin C (2 g/day for 2 weeks) before alcohol consumption (0.8 g/kg) demonstrated a significant increase in plasma alcohol clearance compared with that in the nonpretreated group. These poorly understood interactions may increase the risk of scurvy among individuals who abuse alcohol (5).

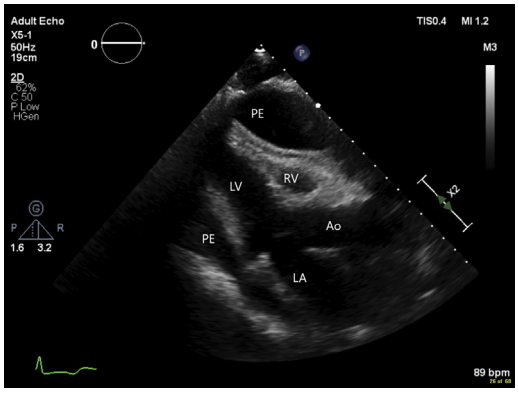
Clinical symptoms of scurvy often manifest after 8 to 12 weeks of vitamin C deficiency and generally occur when the plasma concentration of ascorbic acid is <11 mmol/l. Early stages may present with constitutional symptoms such as malaise and anorexia, followed by mucocutaneous manifestations including gingival bleeding, periodontitis, and dental caries. Skin manifestations of scurvy may resemble a systemic vasculitis (6). Findings include follicular hyperkeratosis, corkscrew hairs, petechiae, and impaired wound healing. Iron deficiency anemia may directly result from chronic hemorrhage or impaired iron absorption (7). Scurvy in children may mimic neurological disorders owing to lethargy and inability to ambulate because of bone pain (4). Most cases of scurvy present with mucocutaneous manifestations, but there are a few case reports of gastrointestinal hemorrhage (8) or PE (9).

Diagnosis of scurvy is based mainly on history, physical examination, and therapeutic response to vitamin C supplementation. Plasma ascorbic acid level of <11 μmol/l supports the diagnosis, although recent intake may elevate the plasma level resulting in a false negative test result. Measurement of ascorbic acid in leukocytes is a better representation of body stores as it is less affected by immediate enteral repletion (10). Unfortunately, this test is not readily available.

Treatment of scurvy consists of daily supplementation with ascorbic acid, 1 to 2 g/day for 3 days followed by 500 mg/day for 1 week, followed by 100 mg/day for 1 to 3 months (5). An intravenous preparation of vitamin C is also available. Correcting the underlying cause of deficiency, with alcohol cessation and dietary counseling, is important. Recovery from scurvy has an excellent prognosis and often improves drastically with repletion of vitamin C.

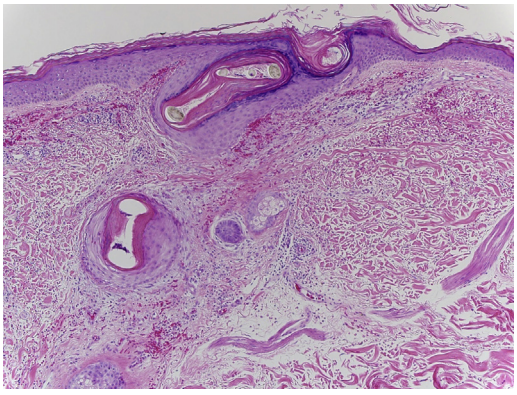
Although scorbutic hemopericardium has been described in published reports, the present report is the first case of scurvy leading to life-threatening cardiac tamponade in the modern era, to the best of

FIGURE 4 Large Pericardial Effusion With RV Collapse in Diastole



Transthoracic echocardiogram in the parasternal long-axis view shows a large pericardial effusion with RV collapse in diastole (a highly specific sign for tamponade). See Video 1.

FIGURE 6 Biopsy Results Showing Follicular Hyperkeratosis, Follicular Plugging, Twisted Hair Follicles, and Perifollicular Hemorrhage



Histopathology of skin biopsy shows follicular hyperkeratosis, follicular plugging, twisted hair follicles, and perifollicular hemorrhage. There is mild superficial perivascular chronic inflammation with extravasated erythrocytes.

the authors' knowledge. This patient had 2 risk factors for vitamin C deficiency: he was a heavy alcohol user, and his diet had been limited to pasta, pudding, and cottage cheese for several years.

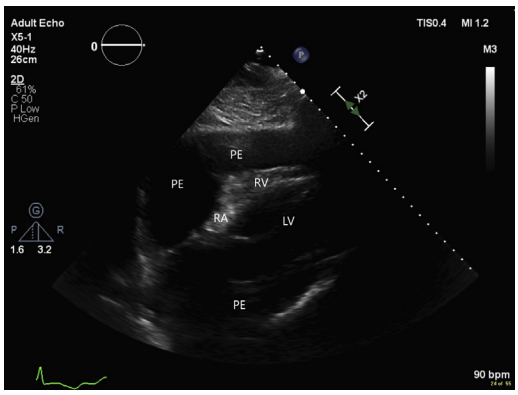
FOLLOW-UP

Transthoracic echocardiography repeated 5 days later showed resolution of PE (**Figure 7**, Video 3). His hospital course was uneventful. The patient received outpatient follow-up with his cardiologist and had serial unremarkable echocardiograms.

CONCLUSIONS

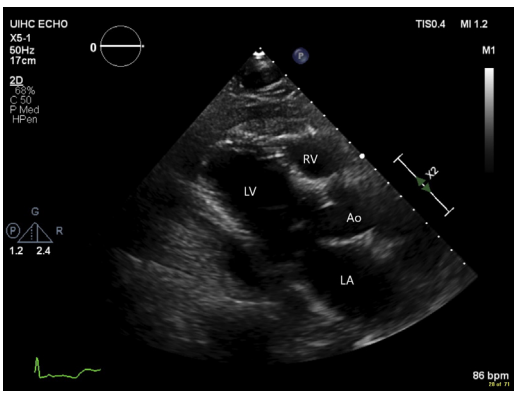
Although not commonly seen in industrialized countries, scurvy is re-emerging in the malnourished

FIGURE 5 Large Pericardial Effusion With Complete Collapse of the RA and RV In Diastole



Transthoracic echocardiogram in a subcostal view shows a large pericardial effusion with complete collapse of the RA and RV in diastole, as well as abnormal septal motion. See Video 2. RA = right atrium; RV = right ventricle.

FIGURE 7 Postpericardiocentesis With Normal Chamber Sizes in Diastole



See Video 3. Ao = aorta; LA = left atrium; LV = left ventricle; RV = right ventricle.

JACC: CASE REPORTS, VOL. 1, NO. 2, 2019
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alcoholic patient population. Clinicians should have a high index of suspicion in order to promptly diagnose the disease, especially among populations at risk.

ADDRESS FOR CORRESPONDENCE: Dr. Saed Alnaimat, Department of Internal Medicine, University of Iowa, 200 Hawkins Drive, SE 612 GH, Iowa City, Iowa 52242. E-mail: saed-alnaimat@uiowa.edu.

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KEY WORDS alcoholism, malnutrition, scurvy, tamponade, vitamin C

APPENDIX For supplemental videos, please see the online version of this paper.