A sCurveball on the Wards: Uncovering the Cause of Ecchymosis and Anemia

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Case Description

The patient is a 65 year old man with a history of tobacco use who reports progressive lower extremity ecchymoses and fatigue, and who is found to have acute anemia referred for workup of coagulopathy.

The patient presented with 1 week of progressive dyspnea that renders him unable to climb the stairs in his house. In the background, he reports three months of fatigue and gradually progressive generalized weakness, and one month of progressive swelling, extensive bruising of both lower extremities. He is a current smoker and denies drinking alcohol. Labs available at the time of presentation are notable for a hemoglobin of 6.6 (from baseline 16 one year prior), INR of 1.2 and PTT of 40. Fibrinogen is 450 and total bilirubin is 4.9.

He was initially being worked up as an outpatient for his fatigue and a progressive anemia, but the rapid drop in Hgb led to his admission to another hospital. A mixing study was conducted which was reported as “equivocal”, and he was transferred to our institution for further evaluation.

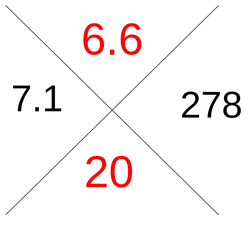
Social History

An initial social history addressed the issue of diet in a cursory fashion, and the patient reported enjoying a variety of food groups. Later, a more detailed social history reveals that the patient had been subsisting on a diet of boxed macaroni and cheese, frozen TV dinners, and bread for many years. He claimed to enjoy fruit-- orange specifically-- but in retrospect realized it had been years since he had had any.

Physical Exam

Revealed a pleasant, edentulous man with extensive ecchymoses that had become confluent and covered ~50% of the body surface area below the inguinal line bilaterally, with no involvement of the abdomen or upper extremities.

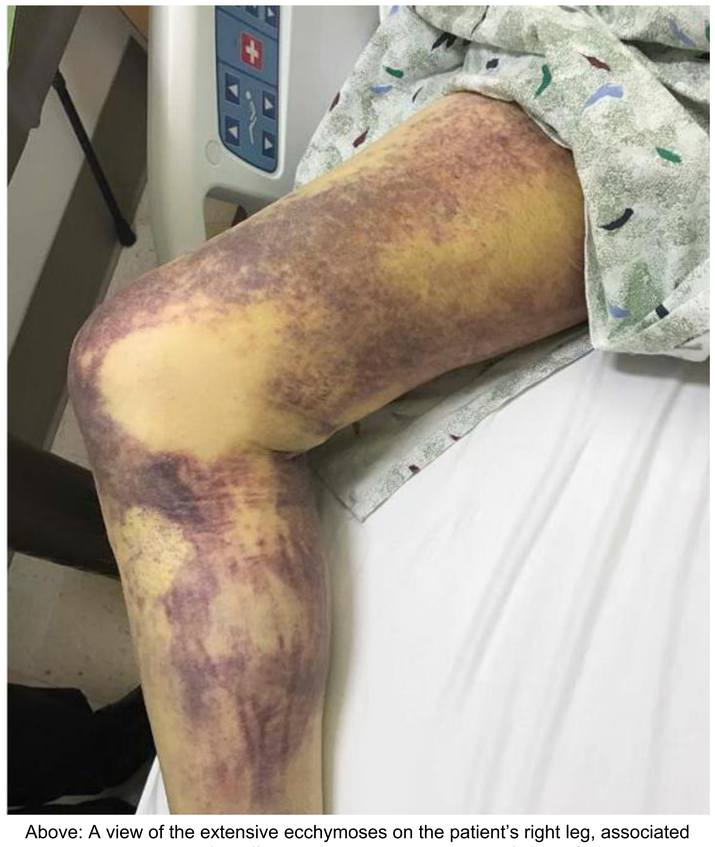
Kinky hairs, some erupting from small red papules were noted on the R shoulder.



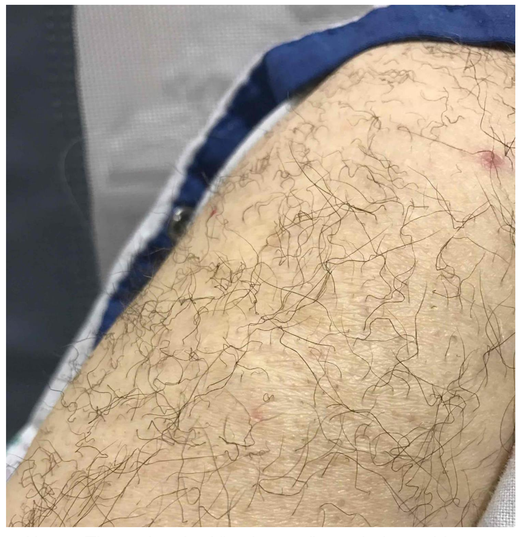
Labs

-Serum Cr: Normal
-INR: 1.2
-PTT: 40
-Fibrinogen: 319
-Bilirubin: 5
-Mixing study: aPTT decreased from 39-> 30 upon mixing

Normal: LDH, haptoglobin, Coombs, ANCAs, ANA, Factors VIII, IX, XI, XII



Above: A view of the extensive ecchymoses on the patient’s right leg, associated with painful swelling that made the patient unable to walk.



Above: The patient had “corkscrew” type hairs on his arms and shoulders, as well as scattered areas of folliculitis (upper right of photo).

Aove: The left leg. The bruising was extensive on both legs but-- curiously-- stopped abruptly at the level of the inguinal ligament with no areas of involvement on the trunk or upper extremities.

Hospital Course

- Differential diagnosis included acquired factor deficiency, vasculitis, and vitamin deficiency
- Repeated labs redemonstrated a slightly prolonged PTT, but workup was otherwise unremarkable for specific factor deficiencies (Factor VIII, IX activities preserved)
- Treated with 1g IV vitamin C daily
- Vitamin C level from admission resulted as undetectable
- Ecchymoses rapidly improved and the patient recovered his strength quickly
- Nutrition was consulted to provide the patient with recommendations on how to improve his diet at home, and was instructed to assist his brother in making changes as well.

Discussion:

Scurvy is a pathologic deficiency of vitamin C, a necessary cofactor for collagen synthesis. It is the only known acquired, non-autoimmune connective tissue disorder. In the developed world, it is rare given the presence of vitamin C in most common foods. As a result, it has become increasingly difficult to recognize. Nevertheless, scurvy is a potentially lethal but entirely treatable condition that can be diagnosed based on a careful history and physical exam, prior to a complex hematologic workup.

In its advanced stages, scurvy leads to anemia simply due to extravasation of blood due to breakdown of collagen protecting the intravascular compartments. Our patient experienced hemarthrosis, followed by exsanguination into soft tissues of the legs (a distribution reflected in other case reports). This resulted in a profound decrease of 10g/dL of Hemoglobin below his baseline with associated hyperbilirubinemia. The classic presentation of scurvy is with extensive bruising despite normal coagulation studies, so the extent of blood loss in this patient was felt to mimic an acquired factor deficiency.

Malnutrition-- and a predisposition to hypovitaminosis C-- can be suspected in patients with historical features including alcoholism, homelessness, recent hip fractures, and even retirement. The diagnosis of scurvy is more challenging in the absence of such features, and requires a detailed social history.

References:

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