Case Report

Hemarthrosis as Initial Presentation of Scurvy

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ABSTRACT. Vitamin C deficiency or scurvy is a disease now rarely seen except for certain populations at risk. When it occurs, diagnosis can be difficult as it can mimic other disorders. Its manifestations are primarily due to an abnormality in collagen formation causing bleeding in the skin, joints, muscles, or gastrointestinal tract and dystrophic hair deformities. We describe a case of scurvy in a 43-year-old man who presented with new onset hemarthrosis with no history of bleeding disorder. He was found to have perifollicular hyperpigmentation and corkscrew hairs, highly suggestive of scurvy. He admitted to completely eliminating fruits and vegetables from his diet and his serum vitamin C level was markedly decreased. Treatment with daily vitamin C supplement led to relief of symptoms and resolution of skin changes. (J Rheumatol 2001;28:1923–5)

Key Indexing Terms:
SCURVY ASCORBIC ACID DEFICIENCY HEMARTHROSIS

In the 18th century, British physician James Lind was the first to recommend inclusion of fruits in the diet to prevent scurvy[1]. Since then, scurvy has been almost completely eradicated in developed countries except in certain populations at risk. Predisposing risk factors include alcoholism, diet fads, dialysis, poverty, malabsorption disorders, and cancers[2-4]. Two groups of individuals are especially susceptible — institutionalized elderly who may not have access to sources of vitamin C, and elderly, edentulous men who live alone and cook for themselves. Given the decreased incidence of this deficiency, it is often misdiagnosed unless these risk factors are identified.

Although the most common cause of hemarthrosis in both pediatric and adult patients is trauma, there are various other conditions that should be considered, including scurvy (Table 1). We describe a case of scurvy presenting as ankle pain and swelling due to hemarthrosis. The diagnosis was made based on the identification of specific skin lesions and low serum vitamin C level.

CASE REPORT

A 43-year-old Argentinian man presented with a 24 h history of sudden onset of bilateral ankle pain, swelling, and erythema. Pain was present at rest and worse with weight-bearing. He also developed low grade fever. There was no history of trauma or prior joint symptoms. He admitted to occasional alcohol and tobacco use but denied any illicit drug use.

Examination was significant for a temperature of 100.8°F, poor dentition, few expiratory wheezes bilaterally, and a 2/6 systolic ejection murmur over the right upper sternal border. Erythema, swelling, and tenderness were noted over the left lateral malleolus and right medial malleolus. Range of motion of both ankles was limited due to pain. Skin examination was significant for facial acne and maculopapular lesions, probably folliculitis, over both lower extremities.

Initial laboratory results included a slightly elevated erythrocyte sedimentation rate (ESR) at 35 mm/h (normal 0–17), low hematocrit at 36.2% (41–53) with a normal mean corpuscular volume, and platelets 114,000/mm^3 (150,000–350,000). White blood cell (WBC) count, prothrombin time, and partial thromboplastin time were normal. Bilateral foot and ankle films showed soft tissue swelling with no bony abnormalities. Fluoroscopy guided left ankle arthrocentesis was performed and 1.5 cc of bloody joint fluid was obtained. Joint fluid cell count had 255,000 red blood cells/mm^3 and 1750 WBC/mm^3 (80% neutrophils). Gram stain and crystal analysis were both negative.

Additional diagnostic investigations included rheumatoid factor, antinuclear antibodies, antineutrophil cytoplasmic antibodies, human immunodeficiency virus, hepatitis and syphilis serologies, PPD, tuberculin skin test, and blood cultures, which were all negative. Chest radiograph gave no evidence of hilar or mediastinal lymphadenopathy. Transthoracic and transesophageal echocardiograms revealed a bicuspid aortic valve with no valvular vegetations.

On closer examination, perifollicular hyperpigmentation and corkscrew hairs were noted, suggestive of scurvy (Figure 1). A vitamin C level was < 0.12 mg/dl (normal 0.2–1.90). On further questioning, the patient admitted to completely eliminating fruits and vegetables from his diet. He was started on 1 g/day vitamin C and within 2–3 days noted a significant decrease in his ankle pain and swelling. Two weeks later, he had no joint symptoms and had interval improvement of his skin lesions.

Table 1. Causes of hemarthrosis.

Trauma
Hemophilia and other blood dyscrasias
Pigmented villonodular synovitis
Hemangiomas
Osteochondral fractures
Intraarticular fractures
Intraarticular tumors
Anticoagulant medications
Neuropathic joints
Reflex sympathetic dystrophy
Scurvy

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Submitted July 17, 2000; revision accepted February 15, 2001.

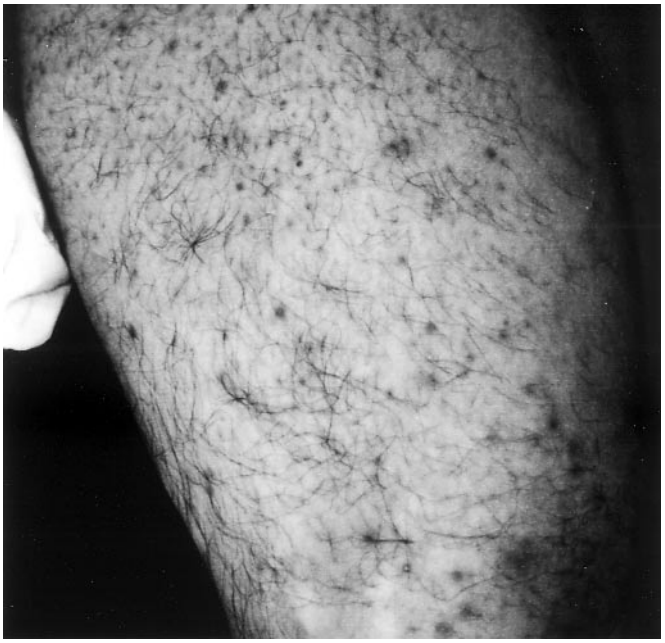

Figure 1. The patient's leg, showing skin changes of perifollicular hyperpigmentation 2 weeks after initiation of therapy.

DISCUSSION

The patient initially presented with ankle hemarthrosis, low grade fever, and a rash for which a rheumatology consult was called. Characteristic skin findings, diet history, and undetectable serum vitamin C level led to the diagnosis of scurvy. This case highlights that scurvy can still be seen in developed countries and should be considered when evaluating patients with identifiable risk factors for this deficiency who develop hemarthrosis.

The recommended daily allowance to maintain a normal total body pool of vitamin C is 60 mg/day. Absence of ascorbic acid in the diet results in symptoms within 90 days when the body pool falls below 300 mg or blood levels drop below 0.3 mg/dl[5]. The clinical manifestations of scurvy reflect the role vitamin C plays in certain physiologic processes. It is a cofactor in the hydroxylation of proline and lysine residues in the process of collagen formation[6]. Lack of this essential cofactor leads to defective collagen molecules, resulting in capillary fragility, dystrophic hair lesions, delayed wound healing, and bone fragility. Electron microscopic studies of the synovial membrane of a scorbutic patient have shown decreased number of mature collagen fibers in the interstitium and perivascular areas[7].

Scurvy also results in altered metabolism of iron and folate, contributing to the development of anemia. Vitamin C reduces iron from the ferric to the ferrous form, the state in which iron is taken up and released by ferritin[8]. Thus, low level of this vitamin leads to decreased cellular availability of iron. In addition, deficiency results in irreversible oxidation and depletion of metabolically active tetrahydrofolate[9].

The earliest clinical signs include petechiae, purpura, and ecchymoses, which may be falsely attributed to a primary bleeding disorder[5,10]. Nonspecific symptoms include low grade fever, easy fatigability, myalgias, and arthralgias[5,10]. Characteristic skin findings include enlarged, hyperkeratotic hair follicles, perifollicular hemorrhage, and fractured or coiled hair deformities. The frequently described gum swelling and bleeding are often most marked in those with preexisting gingivitis and are absent in edentulous patients[3,5]. Late stage complications include intramuscular hemorrhage, hemarthrosis, lower extremity edema, gastrointestinal bleeding, and poor wound healing[2,10]. Nonpalpable purpura is more commonly seen, but excessive cutaneous bleeding can result in palpable purpura mimicking vasculitis[2,10]. Aside from palpable purpura, myalgias, arthralgias, and hemarthrosis, patients can also present with sicca syndrome, the mechanism of which has not been completely elucidated[2,10,11]. These presenting symptoms often bring the patient to the attention of a rheumatologist, as in the case described.

Laboratory findings are nonspecific except for low serum vitamin C levels. The anemia seen in scurvy can be normocytic, microcytic, macrocytic, or megaloblastic[9,10,12]. Thrombocytopenia can also be observed. The deficiency of the metabolically active form of folate associated with scurvy leads to both ineffective megakaryopoiesis and erythropoiesis. When present, indicators of inflammation, such as an elevated ESR, can mislead clinicians into searching for infections or rheumatologic diseases[2,10,12]. Radiographic findings may include femoral head collapse due to ischemic necrosis, osteopenia, and cortical thinning with mild periosteal proliferation[13]. Studies showing decreased bone mineral density in scorbutic guinea pigs support the observation that scurvy can lead to osteoporosis[14].

The goal of treatment is to replenish and maintain vitamin C body stores. Although 10 mg/day of vitamin C can cure scurvy, a more rapid response is seen at higher doses[5]. Clinical improvement, such as defervescence, can be seen as early as 24 h after a dose of 1 g ascorbic acid, with resolution of other manifestations within 1–4 weeks[2,3,12].

The clinical features of scurvy are no longer well recognized and patients are often subjected to extensive, unnecessary evaluation and possibly toxic treatment for other disorders. Awareness of risk factors and characteristic signs and symptoms when evaluating patients can facilitate earlier diagnosis, especially when scurvy presents in the guise of a rheumatologic illness.

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