A child with obstructive oral cavity hemorrhagic bullae and refusal to walk

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Abstract

Background: This case represents a dramatic presentation with obstructive oral hemorrhagic bullae. Scurvy is a preventable disease that is uncommon in developed countries and particularly in developmentally normal children. Scurvy may be encountered by physicians of multiple medical disciplines due to its variable presenting symptoms.

Case presentation: A 2-year-old American born female with no significant past medical history presented to the emergency department with oral cavity bleeding. She was seen by a dentist who referred her to the hospital. Upon presentation, she complained of bleeding gums while brushing her teeth. On further questioning, parents reported patient was a picky eater who refused fruits and vegetables. Vitamin C level was <5 µmol/L, consistent with severe vitamin C deficiency. Four weeks after starting ascorbic acid, patient had resolution of gingival manifestations and was ambulating normally.

Conclusion: Scurvy is a rare disease in developed countries. Pediatric risk factors include developmental delay, selective or restrictive diets, and intestinal malabsorption. Characteristic features include tissue bleeding, gingivitis, non-palpable purpura, and bone disease and dramatic improvement with vitamin C supplementation. Unless it is recognized early, patients may experience increased morbidity, unnecessary laboratory tests, imaging, and procedures. The diagnosis of scurvy is based on presentation and history and requires a high index of clinical suspicion.

Background

Scurvy or severe vitamin C deficiency is a rare disease in developed countries. It was first discovered by Sir James Lind in 1753. Dr. Lind served as a physician with the Royal Navy in the 1700's, during which he conducted a study comparing the diets of sailors with symptoms of "putrid gums…with spots [and] weakness in their knees" [1]. We separated the men into groups and changed their diets. The group that subsisted largely on citrus fruits had a resolution in their symptoms within days. Based on these findings, future sailors were provided rations of lemon juice, thus almost completely eradicating the illness within days. Based on the 2003-2004 cross sectional study by the National Health and Nutritional Examination Survey, the prevalence of vitamin C deficiency in individuals 6 years and older in the United States is about 7.1%, though the majority are clinically asymptomatic [2]. In the pediatric population, at risk groups include children with mental or physical disabilities, autism, eating disorders, patients on restrictive diets, end-stage renal disease patient on dialysis, patients who have undergone bypass surgery or have gastrointestinal malabsorption, and patients with enteral feedings as sole source of nutrition [3-5]. Golriz et al. [4] retrospectively identified 32 patients diagnosed with severe vitamin C deficiency and notably all patients identified had an underlying medical condition, primarily neurologic disorders or sickle cell anemia [6]. Children with a neuro disability should always be subject to a meticulous dietary history in the setting of acute illness [4].

In this case report, we describe an otherwise healthy 2-year-old female who presented with clinical symptoms of scurvy. Untreated scurvy will continue to progress and potentially be fatal. In a young child, progressing oral lesions may become obstructive and with associated gingival bleeding pose airway concerns.

Case presentation

A 2-year-old female with no significant past medical history presented to emergency department with oral cavity bleeding. She was seen by a dentist who referred her to the hospital. Upon presentation, she complained of bleeding gums while brushing her teeth. On further questioning, parents reported patient was a picky eater who refused fruits and vegetables. Vitamin C level was <5 µmol/L, consistent with severe vitamin C deficiency. Four weeks after starting ascorbic acid, patient had resolution of gingival manifestations and was ambulating normally.

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Surgery, Rheumatology, and Nutrition. Surgical pathology revealed nonkeratinizing squamous mucosa with focal ulceration, marked hemorrhage, and granulation tissue.

Review of systems was unyielding and nutritional history disclosed she was a picky eater and ate almost exclusively starches. She was previously healthy, was fully immunized, and took no medications at home.

A Maxillofacial CT was completed to rule out a vascular anomaly and was unremarkable. Secondary to worsening of lower extremity swelling postoperatively an MRI was completed and revealed bilateral lower extremity edema with myositis of the left thigh and calf with no evidence of osteomyelitis. Laboratory results are depicted on table 1. The only identified abnormalities were limited to an Iron level that was 30% below normal, a Prealbumin level 64% below normal, and a Sed Rate 3.5% above normal. With findings suggestive of nutritional deficiency, a more extensive dietary history was obtained. Mother reported that she ate no vegetables or fruit, favored starches, and when she ate pizza she would remove the red sauce and the cheese. The findings of gingival ulceration combined with refusal to walk and lower extremity myositis with edema was suggestive of vitamin C deficiency; vitamin C level was noted to be <5 µmol/L, consistent with severe vitamin C deficiency.

With the diagnosis of Scurvy, the family was given dietary orientation and specific instruction with literature on how to deal with a child who is a picky eater. The patient was placed on a high dose Ascorbic Acid supplementation and a multivitamin with Iron. She was discharged home to be followed by her PCP, ENT and a Nutritionist.

Four weeks after starting Ascorbic Acid supplementation, the patient had complete resolution of gingival manifestations and was ambulating normally.

**Discussion and conclusion**

A deficiency of vitamin C from the diet leads to disruption of collagen synthesis leading to malfunction and absence of mature collagen. Signs of scurvy typically develop after 1 to 3 months of inadequate dietary vitamin C intake [7]. Early symptoms such as fatigue, irritability, myalgias and arthralgias are nonspecific. The classic initial symptoms are vague and include irritability, extremity pain, and abdominal pain [8]. There may be dermatological findings such as petechiae, bruising, hyperkeratosis, and corkscrew hairs [9]. Due to vitamin C’s role in collagen synthesis, the capillaries of these patients are fragile and tend to ooze or hemorrhage in friable tissues such as the gums [9]. Anemia is a common finding, as patients may be iron deficient in combination with losses through bleeding [9]. Eighty percent of patients present with musculoskeletal findings such as arthralgia, myalgia, hemorrhaxis, muscular hemorrhage, and subperiosteal hemorrhage [10]. Otolaryngologic manifestations include periodontal changes, hyperkeratosis, hair loss, keratoconjunctivitis, and xerostomia [11]. The typical defining manifestations such as tissue bleeding, gingivitis, nonpalpable purpura, and bone disease associated with subperiosteal hemorrhages, fractures, often with refusal to ambulate, occur later [12]. There has only been one reported case of an airway concern in a patient with scurvy in which after removal of an LMA (Laryngeal Mask Airway), a 2-year-old patient had copious oral bleeding resulting in an oxygen desaturation and subsequent reintubation [13]. In this author’s review of the literature, there has
not been a case of obstructive hemorrhagic oral bullae in a patient presenting with scurvy.

Vitamin C deficiency is encountered by physicians of multiple medical disciplines. A high index of clinical suspicion in high risk patients is necessary to allow for accurate and timely diagnosis and prevent patient morbidity, unnecessary laboratory tests, imaging and procedures.

References