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A Case Report of Intraoperative Coagulopathy Secondary to Chronic Vitamin K Deficiency

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Abstract

Dietary Vitamin K is a well-known anti-hemorrhagic agent that plays an integral role in the coagulation pathway. Vitamin K is involved in synthesis of coagulation factors; II, VII, IX and factor X. Vitamin K deficiency leads to bleeding diathesis. Hemorrhages usually present in deep soft tissue, rather than mucosal or epithelial membranes, bleeding that is generally caused by disorders of platelets. Major causes of vitamin K deficiency include; medications and diseases involving fat metabolism with a resultant fat malabsorption. Warfarin and Cephalosporins are one of the commonly prescribed medications that lead to vitamin K deficiency. Disease affecting fat metabolism pathway, such as; diseases of the pancreas (cystic fibrosis), short gut syndrome and certain pathologies of the biliary tree. Vitamin K deficiency is more common in newborns. In adults it is uncommon because of its ubiquitous nature and the abundance of its sources. Hemorrhagic disorders in adults due to Vitamin K deficiency are not commonly encountered in practice. We are presenting a case of an adult who presented with a compartment syndrome secondary to a traumatic intramuscular bleeding. Our case highlights the importance of considering vitamin K deficiency in the differential diagnosis of unexplained hemorrhages resulting from a coagulopathy.

Keywords

Vitamin K deficiency; adult; coagulopathy

1. Introduction

Dietary Vitamin K is a well-known anti-hemorrhagic factor that plays an integral role in the coagulation pathway. Its major role in the coagulation pathway is as a coenzyme for gamma-

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glutamyl carboxylase, which is necessary for the activation of coagulation factors VII, IX, X, and prothrombin. Additionally, this enzyme is also required for the activation of the natural anticoagulants, proteins S and C. Hence, vitamin K deficiency thwarts the function of both the coagulation and anticoagulation mechanisms [1,2,3].

The adult daily requirement has been estimated about 90 mcg/day for women and 120 mcg/d in men [4,5], of which 80% is absorbed through the terminal ileum [4,6]. However, like the other fat-soluble vitamins (A, D and E), it requires the proteolytic action of pancreatic enzymes and bile salts to solubilize it for absorption through the ileum enterocytes, from which it is transported to the liver.

Major causes of vitamin K deficiency include; cystic fibrosis and hepatobiliary diseases such as primary biliary cholangitis, primary sclerosing cholangitis, biliary atresia and liver failure. Vitamin K deficiency could also result from the use of commonly prescribed medications such as; warfarin and cephalosporins which inhibit Vitamin K epoxide reductase, the enzyme needed to reduce the vitamin K after it has been oxidized in the carboxylation of glutamic acid residues of the coagulation factors [6,7].

This case aims to report a peculiar presentation of vitamin K deficiency in a man admitted to our surgical service for a decompressive fasciotomy for an intramuscular hematoma leading to compartment syndrome in the lower extremity.

2. Case Description

A 20-year-old man presented with a 2-day history of left thigh pain and swelling after lifting a pallet at work. Initially he felt a tearing sensation and pain which progressed to swelling over 48 hours. On physical exam, his left thigh was swollen, tender with restricted range of motion but palpable femoral, popliteal and dorsalis pedis pulses. Computed Tomography scan showed an intramuscular hematoma of the posterior compartment involving the adductor muscles. Due to the acute swelling and pain, the patient was taken to the operating room for decompression of the hematoma and treat the compartment syndrome.

While the patient was taken for an emergent limb-salvage surgery, decompressive fasciotomy, his coagulation work-up results were available. The patient's preoperative coagulation profile showed prothrombin time (PT) 100 sec (normal range 11–13.5 seconds), activated partial thromboplastin time (aPTT) of 96.5 seconds (normal range 30–40 seconds) and a fibrinogen level of 440 mg/dL (normal range 150–400 mg/dL).

While the hematoma evacuation was taking place, patient was given Fresh Frozen Plasma (FFP) that normalized his coagulation parameters with a PT of 25 seconds and an aPTT of 45.5 seconds. Coagulation profile repeated the day after showed again a PT >100 seconds and an aPTT of 90.2 seconds, mixing studies were obtained for both and the PT, aPTT numbers corrected to normal range, supporting the diagnosis of a factor deficiency. Factor V and X assays after the FFP were normal, 60% and 123%, respectively.

Further history taking indicated that the patient had an abdominal surgery at 3-days of age due to neonatal jaundice with a prolonged course in the neonatal ICU. The patient had an

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"Exploratory-Laparotomy, Jejuno-Ileostomy and Kasai procedure (Hepatoportoenterostomy)" secondary to colonic perforation and ruptured common bile duct, due to Choledochal Cyst, type IVc, and Biliary atresia.

Subsequently, later in childhood the patient was referred to a pediatric Gastroenterologist for Steatorrhea, poor weight-gain and short-stature. He underwent an extensive workup in childhood and was diagnosed with fat-soluble vitamins deficiency, celiac disease and other similar illnesses had been ruled out. Upon the diagnosis vitamins levels were; Vitamin K <0.13 ng/mL (Normal range 0.2–3.2 ng/mL), Vitamin E 0.2 mg/dL (Normal range 3–18 mg/ dL), Vitamin A 20 mcg/dL (Normal range 20–60 mcg/dL), and Vitamin D <3.4 ng/mL (Normal range 30–60 ng/mL). Also, vitamin K depended factors were low, elaborated by factor assays; Facto VII 16.6% (Normal range 70–150%), Factor IX 45% (Normal range 70–120%), Factor X 13% (Normal range 70–150%) and Factor II assay was 36% (Normal range 70–120%), thought Factor V, a non-vitamin K dependent, showed a normal assay of 98% (Normal range 70–150%). Hence, diagnosed with Fat soluble vitamin Deficiency and was prescribed life-long oral vitamin K, of which he was not compliant.

3. Discussion

Vitamin K deficiency is common in the newborns and can be treated by administration of vitamin K in early hours of life [8,9]. Infant is at risk for vitamin K deficient bleeding (VKDB), may present with bleeding from mucosal surfaces, gastrointestinal hemorrhage, bleeding of the umbilical stump or circumcision site, and/or intracranial hemorrhage [10,11,12,13].

Vitamin K deficiency in adults is uncommon because of its ubiquitous nature and the wide abundance of its sources, including; vegetables and other food products. Other factors that makes Vitamin K deficiency in adults less commonly encountered are; the body's ability to recycle Vitamin K, and adequate gut flora production of Vitamin K. In adults, iatrogenic Vitamin K deficiency could result from medications or surgeries that alter fat metabolism leading to Vitamin K deficiency; resection of the terminal ileum, hepatobiliary surgeries that increases enterohepatic circulation or pancreatectomy [14,15,16].

The coagulation abnormalities caused by vitamin K deficiency are manifested as prolonged prothrombin time (PT) and International Normalized Ratio (INR). When the deficiency is mild, only the PT may be prolonged, due to a predominant effect on factor VII. In severe vitamin K deficiency, both the PT and activated partial thromboplastin time (aPTT) may be affected.

Reported cases of Vitamin K deficiency in adults are scarce and rarely results into an extracranial bleeding. The unique aspects of this presentation are age, the severity and the site of the bleeding leading to compartment syndrome. Most of published reports indicate bleeding in newborns, with unique and unusual presentations occurring mostly later in infancy [12,13,17,18,19,20].

Clinicians should be aware of the possibility of vitamin K deficiency in cases of unexplained hemorrhages resulting from a coagulopathy. In order to prevent such potentially life-

threatening complications, patients and parents should be educated well about Vitamin K and drugs that might interact with it leading to a deficiency. Introducing bracelet alerts would also be a viable option.

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