Scurvy in a patient with Home Parenteral Nutrition (PN)

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Background
- Vitamin C is essential for the maintenance of intercellular connective tissues, osteoid, dentine and collagen. (1)
- Vitamin C deficiency is rare nowadays. Clinical manifestations ranges from non-specific symptoms to mucosal and cutaneous symptoms (gingival swelling, ecchymosis, bleeding), musculoskeletal symptoms (bone, joint pain and swelling), poor wound healing, cancer and death in advanced cases.
- The recommended daily allowance (RDA) of vitamin C is 15–45 mg for age 1–13 years and 65–75 mg for age 14–18 years. (1)
- Preterm and term infants up to 12 months of age on PN should receive 15–25 mg/kg/day, and older children 80mg/day of vitamin C. No recommended routine monitoring schedule (2)
- Scurvy has been reported in children with restrictive diet due to developmental disorders, Anorexia nervosa, Coeliac disease, inflammatory bowel disease, Cancers, haemodialysis, severe allergies and enteral feeding (3)

Case presentation
- A 16-year-old boy with intestinal failure due to ultra short bowel syndrome on home PN for 14 years. He had restricted dietary intake due to intestinal rapid transit.
- Presented with 4 weeks history of non-pruritic rash on his legs and 2 weeks of bruising with ankle and knee pain. There were numerous peri-follicular petechiae distributed symmetrically over his lower legs with a few scattered lesions on the arms and trunk. (see photos I and II)
- Initial investigations did not reveal blood dyscrasia, infection, or autoimmune disorder. Vitamin A, D, E, B1, B12, folate, Zinc, Copper, Selenium, and Manganese were within normal limits.
- Rheumatology and dermatology opinions were sought; diagnosis of Scurvy was suspected.
- Skin Biopsy showed perifolliculitis with erythrocyte extravasation suggestive of Scurvy.
- Four days after starting vitamin C supplement, symptoms completely resolved. Subsequently, plasma vitamin C level came back low at 1.6 μmol/L (<11 μmol/L indicates deficiency).
- An enquiry with the compounding company revealed inadvertent use of a uni-layer bag for his home PN, allowing accelerated degradation of Vitamin C despite addition of the appropriate amount at manufacture.

Discussion
- PN is a lifesaving treatment for patients who cannot be adequately nourished by other feeding routes.
- Vitamin C is not routinely measured.
- Analysis of ascorbic acid in the uni-layer bag showed degradation by 50% every 24 hours due to oxidation and by 14 days virtually none was detected. Our patient used bags stored for up to 14 days. In a multi-layer bag, 50% degradation occurs over 14 days, but the amount remaining in the bag was above the recommended daily allowance of 100mg/day. Rate of vitamin C decay depends on the constituents of PN, storage environment and the bag containing it. (1, 4)

I- Perifollicular Petechiae
II- Bruising and Petechiae

Take home messages
- Increased survival is now seen in children with ultra short bowel. 6
- Vitamin C content is markedly reduced if PN given near expiry date.
- Stock control is advised so that patients with ultra short bowel benefit from optimum vitamin C content of home PN.

References:
2. ESPGHAN/IFSPP/CISPIN guidelines on pediatrics parenteral nutrition: Vitamin. 2018.

Disclosures:
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Patient permission obtained to publish the photos