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Case Report

An Unusual Case of Spontaneous Bilateral Orbital Hematoma

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Abstract

Scurvy is preventable disease caused by deficiency of vitamin C. It can present with non specific signs and symptoms. Therefore it can be easily missed. Severe features of vitamin C deficiency are rare now-a-days. The most common risk factors for scurvy in children include poor nutrition, restricted diet secondary to developmental disorders or psychiatric illness coupled with low socioeconomic status. This case report demonstrates the importance of high index of clinical suspicion in diagnosing scurvy in children with long standing neurological illnesses on highly deficient diets especially in those who are unable to feed themselves.

Background

Scurvy is a spectrum of disorder characterized by severe prolonged vitamin C deficiency. It is important for synthesis of collagen, catecholamines, and carnitine and for cholesterol metabolism [1]. Incidence of vitamin C deficiency is documented to be as high as 73.9% in developing countries like India [2]. It is characterized by hemorrhagic gingivitis, subperiosteal hemorrhages, perifollicular haemorrhages, petichiae, epiphyseal separation, bone pain, swelling and anemia. Infants and children on highly restrictive diets devoid of fruits and vegetables with chronic illness like developmental disorders and/or physical disabilities are at significant risk. Orbital hemorrhage most commonly occurs due to trauma or post surgery [3]. Scurvy is a historically important cause of orbital hemorrhage, rarely seen in recent times. Therefore cases of scurvy get unnoticed for long periods, mandating need of high clinical suspicion in such cases. Only few cases of scurvy leading to orbital hematoma are reported in literature. We report a child with bilateral orbital hematoma due to vitamin C deficiency.

Case Presentation

A 4 year old boy diagnosed as a case of West syndrome with global developmental delay was referred from orthopaedics department with left leg above knee slab placed in view of distal end femur fracture since 2 weeks with history of bilateral knee swelling with bony tenderness since three months. He also had complaints of gradually progressive swelling of bilateral eyelids with proptosis of right eye since 10 days. There was no history of fever, active eye discharge or any other signs of sepsis. There was no known history of trauma or abuse. No history suggestive of insect bite, preceding upper respiratory tract infection or foul smelling nasal discharge or signs of infection on the danger area of face was found. Further probing into the history revealed that the child had frequent episodes of gum bleeding and was predominantly on milk based diet with no history of fruit intake. There was no history suggestive of bleeding diasthesis. Child was bedridden on Ryle's tube feeding with a GMFCS score of level 5. For West syndrome child was on steroids and multiple antiepileptics. On examination child was lethargic with bilateral eyelid swelling along with proptosis of right eye measuring 30 mm. It was non reducible, non pulsatile and associated with limitation of eye movements in all directions however pupillary response was normal (Figure 1). There was significant periorbital bluish discolouration with chemosis. Fundus and cup: disc ratio was normal. Child had significant pallor, spongy bleeding gums, loss of teeth (Figure 2) with tender rosaries in chest along with tenderness in shin region. Child had above knee slab in left leg and swelling in right knee, though there was no crepitus or abnormal mobility (Figure 3). So a possibility of scurvy was kept. On investigations, haemoglobin was 6.3 g/dl, counts and coagulation studies were normal. IV antibiotics were started keeping a possibility of orbital cellulitis. To rule out cavernous sinus thrombosis, MRI brain with orbital sections was planned. USG right knee revealed no effusion. X-ray bilateral knees was also suggestive of scurvy showing wimberger sign, diffuse osteopenia with cortical thinning, metaphyseal irregularity seen at proximal distal femur and proximal tibia, pelkan spurs and white line of Frenkel, findings consistent with scurvy (Figure 4). X-ray left femur revealed anteromedial displacement of distal femur suggestive of salter harris type 1 fracture (epiphyseal separation). MRI revealed bilateral subperiosteal hematoma in orbit and no signs of cavernous sinus thrombosis (Figure 5). Antibiotics were stopped and high dose of vitamin c was initiated. Within ten days of treatment child had significant improvement. Bony tenderness and knee swelling reduced. Improvement in eye symptoms was also documented in form of decreased eyelid swelling and improved eye movements. At discharge, no eyelid swelling in both eyes and eye movements were full and free in all gazes and directions. Bony tenderness resolved.

Discussion

Scurvy is a preventable and treatable disease caused by poor intake or absorption of ascorbic acid. Humans don't have the gene for L-gluconolactone oxidase, enzyme which converts glucose into ascorbic acid. Thus vitamin c is an essential exogenous vitamin. Children fed predominantly heat treated (pasteurized) milk or unfortified formulas and not receiving fruits and fruit juices are at significant risk for symptomatic disease [1]. Best food sources of vitamin c are citrus fruits, fruit juices, berries, guava, tomato and green leafy vegetables. In scurvy there is defective formation of connective tissues and collagen in skin, cartilage, dentine, bone and blood vessels,

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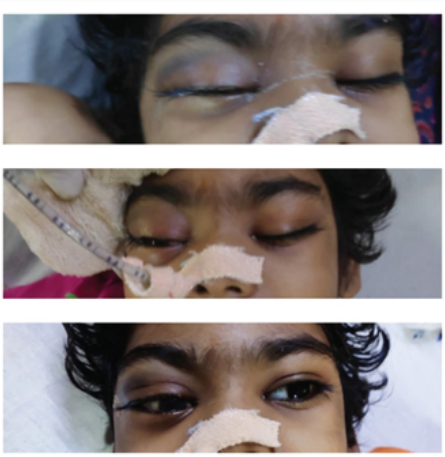

Figure 1: Bilateral eye swelling (a), gradual response of vitamin c supplementation (b,c).

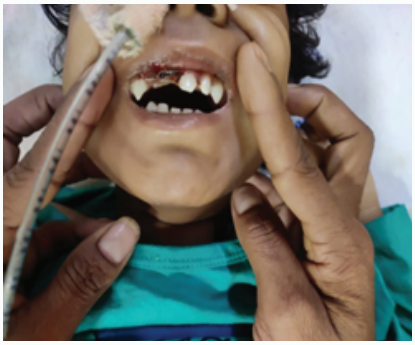

Figure 2: Spongy bleeding gums with loss of teeth.

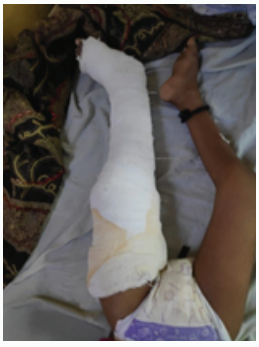

Figure 3: Above knee slabin left leg in-situ.

leading to their fragility. In long bones, osteoid is not deposited by osteoblasts, cortex is thin and trabeculae become brittle and fracture easily [1]. Manifestations of symptoms usually start usually after 8-12 weeks of vitamin c deficiency when stores get exhausted. Low serum ascorbic acid of < 0.2 mg/dl along with significant clinical history and examination features confirms diagnosis of scurvy. Pediatric patients commonly present with musculoskeletal manifestations, present in around 80% patients [4]. With damage to synovial blood vessels and microfractures, there is pain and swelling secondary to subperiosteal haemorrhages and hemarthroses additionally partial and complete separation of the epiphysis and fracture of calcified cartilage of epiphyseal plate are known to occur in scurvy [5,6]. This was also seen in our case. A similar case report from Delhi show epiphyseal

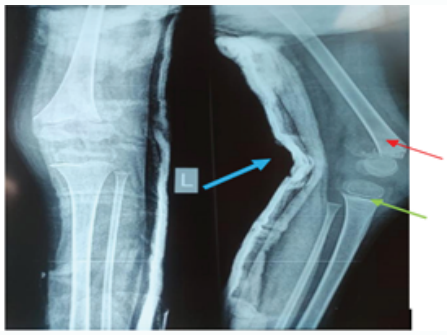

Figure 4: X-ray bilateral knee showing anteromedial displacement of distal end of femur suggestive of epiphyseal separation (Red arrow). Tibia shows wimberger sign (green arrow), pelkan spurs (blue arrow), pencil thinning of cortex and white line of frankel.

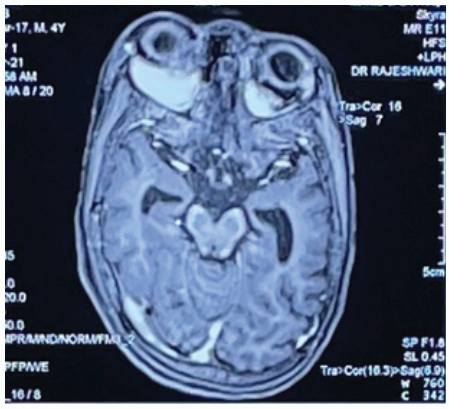

Figure 5: MRI scans showing bilateral subperiosteal orbital hematoma.

separation due to scurvy in 6 yr old child with cerebral palsy [7]. Anemia which is a common manifestation of scurvy is related to impaired iron absorption and co-existent micronutrient deficiency of iron, vitamin b12 and folate. Scurvy has classic radiographic findings. The typical radiographic features occur at the distal ends of long bones and are particularly common at knees. The shaft of long bones has a ground glass appearance because of trabecular atrophy. The cortex is thin and dense, giving appearance of pencil outlining of the diaphysis and epiphysis. The white line of Frankel, an irregular but thickened white line at the metaphysis, represents the zone of well-calcified cartilage. The epiphyseal centres of ossification also have a ground-glass appearance and are surrounded by a sclerotic ring. The more specific but late radiologic feature of scurvy is a zone of rarefaction (trummerfeld zone). A pelkan spur is a lateral prolongation of white line and maybe present at cortical ends. Epiphyseal separation along the line of destruction can also be found. Occular manifestations include subconjuctival hemorrhage, lid hematoma, anterior chamber bleeding, retinal hemorrhage and proptosis [8]. Proptosis as a manifestation of scurvy is rare. In about 10% of patients with infantile scurvy, proptosis occurs as a result of hemorrhage in the orbital bones [9]. Orbital haemorrhages are typically superior and subperiosteal as they were in our patient. Similar presentation was found in case report of 13 year old girl with 18 hour history of right proptosis with periorbital discomfort, improved on vitamin c administration [10]. Another case report also reveals spontaneous orbital hematoma in an infant due to scurvy [11]. Similar case findings were observed in case report published in Malaysia [12]. Antiepileptic therapy can also affect

vitamin levels in patients who are on long term medications. Anti-epileptics increase catabolism of nutrients by inducing cytochrome p-450 enzymes. A study done in Chinese epileptic patient's reveals that vitamin c levels are below reference range in majority of them [13]. Mainstay of treatment in scurvy is replacement of vitamin c in a dose of 100-200 mg daily for 3 months. Spontaneous bleeding usually improves within few days after initiation of therapy. Complete resolution takes place in 3 months. During regular follow-up on OPD basis, bony tenderness along with orbital swelling resolved completely on vitamin c supplementation

Conclusions

This child with West syndrome being on restricted diet demonstrated features of scurvy needed a high index of suspicion and highlighted the importance of dietary history and to keep a keen eye for signs of micronutrient deficiency. Clinical presentation was initially misleading so it is important to keep the diagnosis in mind in case of unexplained bleeding associated with bone fractures. Patients who are on long-term antiepileptic drugs are prone to vitamin c deficiency along with vitamin D and B. Therefore we should also supplement vitamin c in these kinds of patients on marginal diets who are at high risk of developing deficiency.

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