

Case Report

Unexpected Scurvy Presenting with Proptosis: A Case Report

Jelvin T. Co*, Timothy Joseph S. Orillaza and Romelito Jose G. Galsim

Department of Radiology, The Medical City, Ortigas Avenue, Pasig City

* Contact Details: jelvin.co@gmail.com

ABSTRACT: Scurvy is a state of dietary deficiency of ascorbic acid, or vitamin C as it is more commonly known. Since the discovery of the link between scurvy and vitamin C, it has become a rare condition. We present a 5 y.o. Filipino female with proptosis. An intraorbital neoplastic process was primarily entertained. Only after magnetic resonance imaging (MRI) revealed bilateral supraorbital subperiosteal hematoma formation was scurvy considered and empiric treatment of vitamin C supplementation started, yielding significant improvement. Given the multiple tools that we have in the information age, MRI plays a role in helping diagnose an easily treatable condition.

Keywords: hemorrhage, MRI, proptosis, scurvy

INTRODUCTION

Since the discovery of the link between scurvy and vitamin C deficiency, scurvy has become an uncommon condition in all age groups¹. Cases of scurvy may still be found in industrialized societies, frequently among the extreme ages of the population². The initial presenting features of scurvy are nonspecific, ranging from fatigue to musculoskeletal complaints³. As scurvy progresses, patients present with lethargy, osteoporosis and impaired wound healing. Laboratory workup may also reveal nonspecific results such as anemia. Left untreated, scurvy may have fatal complications⁴.

In the pediatric population, scurvy usually involves symmetric bone disease and a tendency to hemorrhage⁵. Because of its rarity, other conditions are often considered before its diagnosis², and a high index of suspicion in the proper clinical setting is needed. In the absence of x-ray findings, features of hemorrhage as seen through magnetic resonance imaging (MRI) can lead to its diagnosis. This case highlights a rare case of scurvy, its unique manifestations and findings seen on MRI.

CASE REPORT

A 5 y.o. female presented with a 6-month history of severe constipation associated with bloody stools, loss of appetite and body malaise. Multiple outpatient consultations were unrevealing, and led to assessments of constipation and growing pains. The patient was managed symptomatically with temporary relief. Initial radiologic work-up revealed normal x-rays of the extremities and chest as well as an unremarkable ultrasound of the whole abdomen. In the interim, the patient gradually developed progressive weakness, fatigue, body and joint pains. Three hours prior to consultation, she complained of bilateral eye pain and eyeballs “popping out,” prompting a visit to the emergency room.

The patient’s birth, past medical, developmental and personal/social histories were unremarkable. The patient’s feeding history was noted to be completely deficient in fruits and vegetables, with note of vomiting when eating the latter. Pertinent physical examination of the patient revealed bilateral proptosis with tenderness, generalized abdominal pain, gum swelling and body weakness. Workup involved a complete blood count, urinalysis, chest and skull x-rays, all of which were normal. No other radiographs were done. At this point, the initial impression was that of retinoblastoma. A referral was made to the Ophthalmology service whose assessment was neuroblastoma. She was then referred to the pediatric Hematology/Oncology service for further management.

On the 1st day of admission, MRI studies of the chest, brain, orbits and whole abdomen with contrast enhancement were requested to evaluate for malignancy. The MRI studies of the chest and abdomen were normal. The cranial and orbital MRI examinations (Figures 1 to 3) revealed bilateral orbital subperiosteal foci with features suggestive of hematoma formation and its mass effects. Based on these findings, a range of differentials including trauma, metabolic disorders, hemorrhagic diatheses, vaso-occlusion, and abscess formation, were considered. The unremarkable MRI of the chest and abdomen, along with a normal complete blood count made leukemia or neuroblastoma less likely. There was no history of trauma. The patient had an unremarkable past medical history, ruling out hemorrhagic diatheses and vaso-occlusion. As the patient did not present with fever, an infection was excluded from the differential diagnoses.

A metabolic disorder was considered given the patient’s feeding history, and empiric treatment with vitamin C was initiated. The patient’s clinical picture progressively improved. On the 3rd day of admission, the patient’s pain decreased significantly. By the 6th day, the proptosis resolved completely.

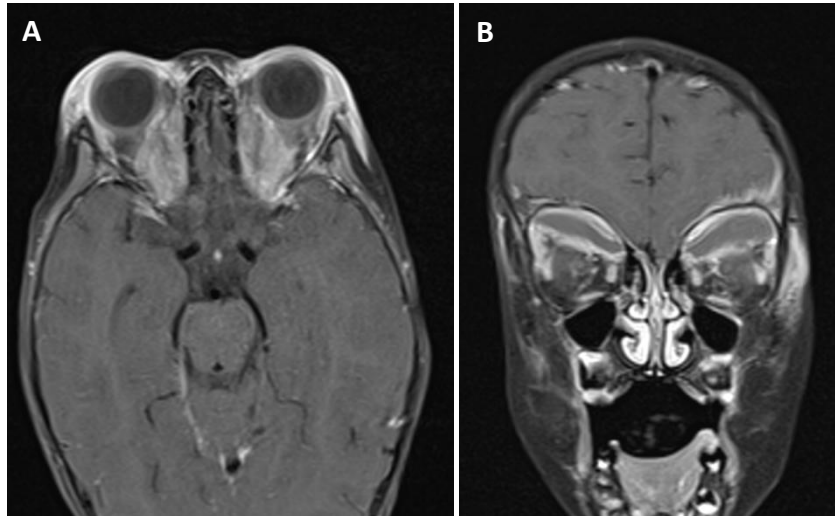


Figure 1. T1 weighted fat suppressed gadolinium-enhanced MRI sequences of the (A) axial and (B) coronal planes of the head and orbits showing rim enhancing spindle-shaped predominantly hypointense masses occupying the superior extraconal regions of both orbits.

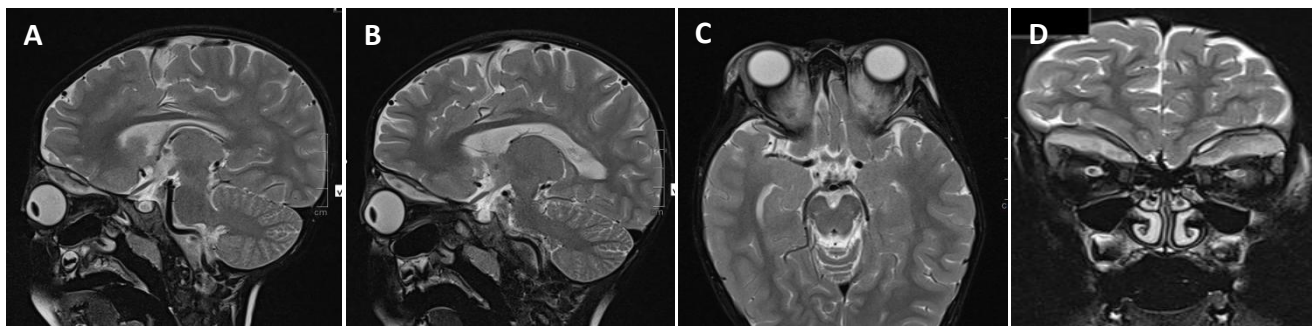


Figure 2. T2 weighted MRI sequences of the sagittal planes of (A) right and (B) left eyes, and (C) axial and (D) coronal planes of the head and orbits show the predominantly hypointense masses occupying the superior extraconal regions of both orbits.

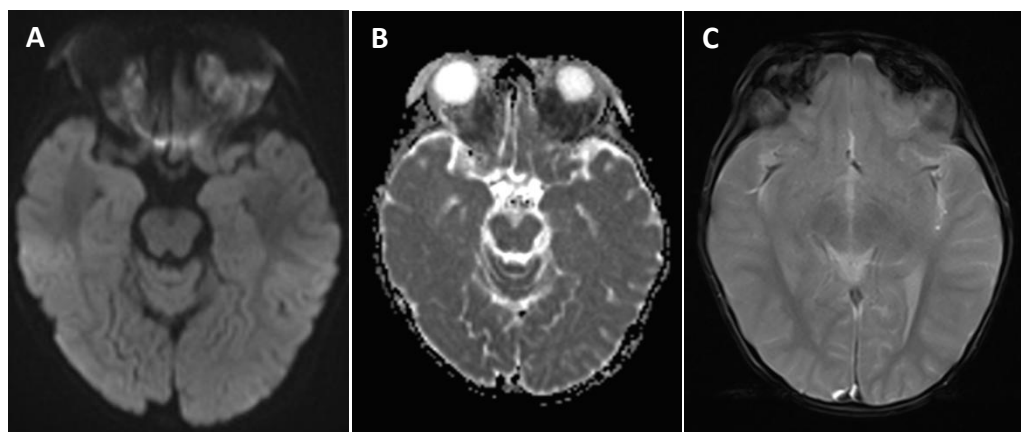


Figure 3. (A) Diffusion weighted imaging (DWI) with (B) apparent diffusion coefficient (ADC) maps and (C) susceptibility weighted imaging (SWI) MRI sequences showing restricted diffusion with loss of signal in SWI of the masses occupying the superior extraconal regions of both orbits

DISCUSSION

Scurvy is one of the oldest diseases known to humankind, with evidence of its existence as early as 1500 BC⁶. It is a metabolic disorder specific to a deficiency in ascorbic acid⁴. Ascorbic acid is an essential nutrient needed in the synthesis of collagen. Deficiency leads to impaired wound healing as well as impaired skin, bone and connective tissue formation⁷.

Classic signs and symptoms include asthenia, bleeding disorders and gum abnormalities. These symptoms, however, are uncommon and are seen in its later stages⁸. Patients will usually have nonspecific signs and symptoms. Musculoskeletal symptoms such as arthralgia, myalgia and hemarthrosis are present in up to 80% of cases of scurvy. Among children, lower extremity pain, limping and inability to walk are frequently encountered⁹. Oral symptoms present as swelling, bleeding gums and loosening of teeth. Dermatologic manifestations include petechiae, ecchymoses, hyperkeratosis and perifollicular hemorrhage. Initially, our patient presented with severe constipation, loss of appetite and body malaise. Of these nonspecific signs and symptoms, our patient eventually developed musculoskeletal complaints, with oral findings of gum swelling only noted in the emergency room.

Scurvy in children presents with a wide spectrum of clinical manifestations. These include rashes and weakness¹⁰, difficulty walking¹¹, musculoskeletal pain, and even bilateral proptosis¹². According to Agarwal et al, Vitamin C deficiency generally leads to symmetric bone disease and a tendency to hemorrhage. Of the two, the former frequently attracts the attention of parents and caregivers prompting medical consultation⁵. In our patient, proptosis as a manifestation of intraorbital subperiosteal hemorrhage prompted the ER visit.

Diagnosis of scurvy is made through clinical manifestations of a scorbutic state, biochemical indices and supportive history of vitamin C deficiency⁴. Serum ascorbic acid levels may be low to normal in the presence of scurvy. Recent Vitamin C supplementation, may lead to normal serum ascorbic acid levels in patients with scurvy. The best confirmation of the diagnosis of scurvy is its resolution following vitamin C administration². In the Philippines, testing for serum ascorbic acid is not available. For this patient, the diagnosis of scurvy was confirmed when the patient had drastic improvement upon empiric treatment with ascorbic acid.

Our patient presented with features of painful proptosis and nonspecific symptoms. Scurvy can mimic malignancy, especially leukemia. MRI findings in scurvy have been described to reflect the underlying pathophysiology of hemorrhage in the periosteum^{2,5}, which can be seen in the shafts of long bones^{13,14}. When seen along with periostitis, metaphyseal changes and heterogeneous bone marrow signal intensity, scurvy should be included in the differential

diagnosis¹⁴, but these were absent in our patient. MRI findings of scurvy in the orbits can present with hematoma formation as previously reported^{12,15,16}. The diffuse marrow changes expected of malignancy such as leukemia are typically absent⁵.

Orbital subperiosteal hematoma is not pathognomonic for scurvy. It may be associated with a history of facial trauma or barotrauma¹⁷ as well as Valsalva maneuver, bleeding diathesis, anticoagulation therapy and other systemic diseases. Rupture of the diploic veins between the periosteum and the bony orbit, results in local hemorrhage with subsequent hematoma formation. Left untreated, this may spontaneously resolve or enlarge to cause proptosis.

Our patient presented with a 6-month history of severe constipation accompanied by crying episodes and bloody stools. Prolonged duration of recurrent Valsalva maneuver prompted by severe constipation can cause orbital hemorrhage owing to the increased vascular fragility caused by defective collagen formation in vessel basement membranes. We thus establish the connection between constipation, scurvy, orbital subperiosteal hematoma formation and proptosis. Ocular lesions are uncommon manifestations of scurvy and most present with subconjunctival or orbital hemorrhages with the latter typically located in the superior and subperiosteal areas²⁹. With bilateral extraorbital MRI findings suggestive of subperiosteal hematoma formation, and given the other features described above, scurvy was considered in our patient and empiric treatment with ascorbic acid was started. Our patient had dramatic improvement and resolution of symptoms, strongly reflecting the need to be aware of this easily missed deficiency

CONCLUSION

Scurvy is a rare condition, is often associated with nonspecific symptoms, and can easily be missed. Obtaining a thorough history including dietary habits is imperative. Although scurvy often presents with radiographic findings of bone changes in the extremities, it can present with signs of hemorrhage in the orbits. MRI findings of extraorbital subperiosteal hematoma formation seen in the absence of other radiologic findings should prompt consideration of scurvy in the differential diagnosis.

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