

## Unilateral proptosis and extradural hematoma in a child with scurvy

Sumit Verma · Sindhu Sivanandan ·  
Mangalasseril K. Aneesh · Vaibhav Gupta ·  
Rachna Seth · Shushil Kabra

Received: 30 April 2007 / Accepted: 18 May 2007 / Published online: 7 July 2007  
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**Abstract** We report a 3-year-old boy with unilateral proptosis, painful swelling of the right thigh and aphasia. He had radiographic evidence of scurvy in the limbs and bilateral frontal extradural hematomas with a mass lesion in the left orbit on MRI. He was treated with vitamin C and on follow-up 8 weeks later had recovered with no evidence of the orbital mass on clinical or radiological study. Scurvy manifesting as proptosis and extradural hematoma is rare.

**Keywords** Proptosis · Extradural hematoma · Scurvy · Child

### Introduction

In 1753, Sir James Lind gave the first clinical description of vitamin C deficiency disease known as scurvy. In the last 50 years, the number of cases of scurvy has decreased remarkably, and so has the awareness among physicians. It is not surprising to find scurvy and its complications being misdiagnosed as malignancy or infection. One such case with extremely rare complications is presented.

### Case report

A 3-year-old boy presented with proptosis of the left eye for 15 days. He had been unable to walk for the last 2 months and had a poor nutritional history. His diet had consisted of diluted cow's milk and rice since late infancy and lacked fruit and vegetables. On examination he was extremely irritable and was undernourished with weight, height and head circumference well below the third percentile. He had pallor, but no bleeding gums or petechiae. The left eye was proptotic with ulceration and displayed seropurulent discharge and no perception of light. The right eye, including fundus evaluation, was normal. Costochondral junctions were sharply angulated. The right thigh was swollen and tender to the touch. The boy barely moved the affected limb and had motor aphasia.

Laboratory investigation revealed a hemoglobin level of 4.8 g/dl (normal range 8–12 g/dl), a total leukocyte count of  $12,300/\text{mm}^3$  (neutrophils 70%, lymphocytes 30%), a platelet count of  $4.3 \times 10^5/\text{mm}^3$ , and a prothrombin time of 14 s (control 13 s). Peripheral smear showed microcytic hypochromic anemia. His renal function test and electrolytes were within normal limits. Liver function tests showed hypoproteinemia and raised alkaline phosphatase. Radiographs of the right femur demonstrated a thin cortical outline of the epiphysis, splayed metaphysis with evidence of a Pelkan spur, submetaphyseal lucencies in the distal femur and subperiosteal hemorrhage (Figs. 1 and 2). MRI of the brain showed bilateral extradural hematomas compressing the frontal lobes. There was a subperiosteal hematoma in the left orbit along with subretinal hemorrhage and retinal detachment. An additional finding was an ill-defined soft-tissue mass lesion in the superior aspect of the orbit, which was hypointense to the extraocular muscles on T1-weighted images and slightly

S. Verma (✉) · S. Sivanandan · R. Seth · S. Kabra  
Department of Pediatrics,  
All India Institute of Medical Sciences (AIIMS),  
Ansari Nagar,  
New Delhi, India 110029  
e-mail: drsumitverma@yahoo.com

M. K. Aneesh · V. Gupta  
Department of Radiodiagnosis,  
All India Institute of Medical Sciences (AIIMS),  
New Delhi, India

**Fig. 1** Right femur with a thin cortical outline of the epiphysis with a splayed metaphysis



hyperintense on T2-weighted images and showed inhomogeneous enhancement on fat-suppressed images after gadolinium administration (Figs. 3 and 4).

Based on clinical findings and investigations, scurvy was the initial clinical diagnosis and the child was started on vitamin C, 300 mg daily. The extradural hematoma was evacuated. On close follow-up 8 weeks later, the child could stand, speak words and his general condition had improved. The left eye swelling had regressed clinically and radiologically with no evidence of a mass lesion in the orbit.

## Discussion

Vitamin C is an essential micronutrient in the diet required for synthesis of supporting tissue of mesenchymal origin such as osteoid in bone, chondroitin in cartilage, dentin in teeth and collagen on capillary walls. It enhances iron absorption from the gastrointestinal tract, acts as an antioxidant and is required in the coagulation pathway. Deficiency of vitamin C leads to scurvy. The common presenting features are irritability, pain, tenderness and pseudoparalysis of the limbs; bleeding gums; and sharply-

**Fig. 2** Right femur with subperiosteal hemorrhage



**Fig. 3** T1-weighted axial MR image shows bilateral extradural hematoma compressing the frontal lobes



**Fig. 4** Coronal, T1-weighted fat-suppressed image after gadolinium administration shows an ill-defined soft-tissue mass in the left orbit

angulated scorbutic rosary. Diagnosis of scurvy is mainly based on clinical and radiological features with rapid clinical improvement within 2 days to 1 week of starting vitamin C therapy [1]. Unusual manifestations include hematuria, melena, subdural hemorrhage, subarachnoid hemorrhage and retroorbital hemorrhage causing proptosis. Extraocular muscle palsy can occur due to intraocular hemorrhage. Hence, it is not uncommon to find scurvy being mistaken as a more common disease such as malignancy or infection [2]. Scurvy manifesting as unilateral proptosis associated with frontal extradural hematoma is an extremely rare complication [3].

**Acknowledgement** We thank Prof. Arun Gupta, Department of Radiodiagnosis, for assistance and guidance during the preparation of this paper.

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