

## Caffey's Disease Affecting the Ribs

Nayek K<sup>1</sup>, Barik K<sup>2</sup>, Chaki B<sup>3</sup>, Adhikary A<sup>4</sup>

<sup>1</sup>Dr. Kaustav Nayek, MBBS, MD, Professor, <sup>2</sup>Dr. Kanailal Barik, MBBS, MD, Associate Professor, <sup>3</sup>Dr. Bhusan Chaki, MBBS, PG Trainee, <sup>4</sup>Dr. Amit Adhikary, MBBS, PG Trainee, Department of Paediatrics, Burdwan Medical College and Hospital, West Bengal, India.

### Abstract

Caffey's disease is a rare disorder affecting the bones in infancy. It is characterized by irritability, fever, cortical hyperostosis and adjacent soft tissue swelling. Here is report of an uncommon case where the ribs were significantly affected including the clavicle.

### Introduction

Caffey's disease is a rare disease of unknown etiology. It is characterized by cortical hyperostosis and inflammation of the contiguous muscle and fascia. Mandible is the most common site involved. Here we are reporting a case which has affected the ribs including the clavicle.

### The Case

A six month old male child was admitted to this hospital with the history of high grade intermittent fever since 26 day of age. He was treated with different medications (including iv antibiotics) but fever did not resolve. Patient did not have any history of contact with tuberculosis.

At four months of age patient developed swelling at lower left chest. We had done the routine investigations including a chest X-ray which showed cortical hyperostosis of the ribs of left side. Hyperostosis initially affected the lower ribs and it gradually involved the upper ribs. Hyperostosis also affected the medial end of the clavicle. Initially there was mild pleural effusion also which resolved gradually.



Fig 1: Chest x-ray showing cortical hyperostosis of the ribs of left side.

Complete haemogram showed a decreased hemoglobin level, very high ESR, leucocytosis, and thrombocytosis. Hb was 8.2gm%, ESR 85mm, WBC count 20000/ $\mu$ l with Neutrophil 40%, platelet count was 722000/ $\mu$ l, RBCs morphology were microcytic hypochromic anemia.

A provisional diagnosis of Caffey's disease was made based on the clinical and investigational study. Patient was put on steroid (Prednisolone 1mg/kg/day) and responded dramatically. His fever subsided, swelling decreased and patient became well looking.

### Discussion

Caffey's disease of bone is a rare disorder of unknown etiology. It is characterized by cortical hyperostosis along with inflammation of the adjacent muscle and fascia. Usually sporadic in nature but may be autosomal dominant or autosomal recessive<sup>1</sup>. A linkage to mutations of the *COL1A1* gene (codes for the  $\alpha_1$  chain of type I collagen) has been reported<sup>2</sup>. It was first reported as a disease entity by Caffey and Silverman in 1945<sup>3</sup>.

Bones commonly affected are the mandible, rib lesions and by remissions and relapses. Clavicle, scapula and the ulna, but any tubular hones except the phalanges may also be affected. Mandible is the most common site involved<sup>4</sup>.

The disease manifests as irritability and fever around 10 weeks of age. Soft tissue swelling may precede the onset of bone changes. Swelling is nonsuppurative, have wood like induration with minimal warmth<sup>1</sup>.

Laboratory finding may show a high ESR, low hemoglobin level, increased thrombocyte count. Serum alkaline phosphatase level may also be elevated. Some patients may have raised prostaglandin level<sup>4,5</sup>.

Radiographically, there is periosteal proliferation and cortical thickening under the soft tissue swelling. Hyperostosis may be massive.

Differential diagnosis includes chronic vitamin A intoxication, prolonged prostaglandin E infusion in children with ductal dependent flow, primary bone tumors, and scurvy<sup>1,6,8</sup>.

Treatment includes indomethacin and corticosteroids. Corticosteroids should be used if there is a poor response to indomethacin.

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## How to cite this article ?

Nayek K, Barik K, Chaki B, Adhikary A. Caffey's Disease Affecting the Ribs. *J Nepal Paediatr Soc* 2012;32(3):250-251.