

## Case Report

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# Infantile cortical hyperostosis, masquerading as osteomyelitis: a case report with three year follow up and review of the literature

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## ABSTRACT

Infantile cortical hyperostosis is a self-limiting disease which presents with a classical triad of (1) irritability, (2) soft tissue swelling, (3) cortical hyperostosis of skeleton. Most commonly it affects mandible and its involvement is pathognomonic for this disease. There is no clear cut treatment guideline present in any literature. Here we report a case of Caffey's disease in a 5 month female child initially thought to be osteomyelitis of radius and ulna. We also present a summary of cases reported since 2000.

**Keywords:** Infantile cortical hyperostosis, Osteomyelitis

## INTRODUCTION

Caffey's disease is a rare disorder of skeleton and surrounding soft tissues originally described by Caffey and Silvermann in 1945 as a syndrome of unknown aetiology.<sup>1</sup> It is an episode of massive supperiosteal newbone formation involving mandible, clavicle and diaphysis of long bones.<sup>2,3</sup> It is characterized by bone pain, irritability, fever, pseudoparalysis of affected limbs and soft tissue swelling. The presentation is variable and can mimic that of osteomyelitis but involvement of mandible is characteristic of Caffey's disease.

## CASE REPORT

A five month old female child born to non-consanguineous parents presented to our OPD with complain of swollen and tender left forearm since last 3 weeks and occasional intermittent low grade fever since last 4 weeks. She was being treated outside with parenteral antibiotics since last seven days and referred to our hospital for evaluation of persistent fever.

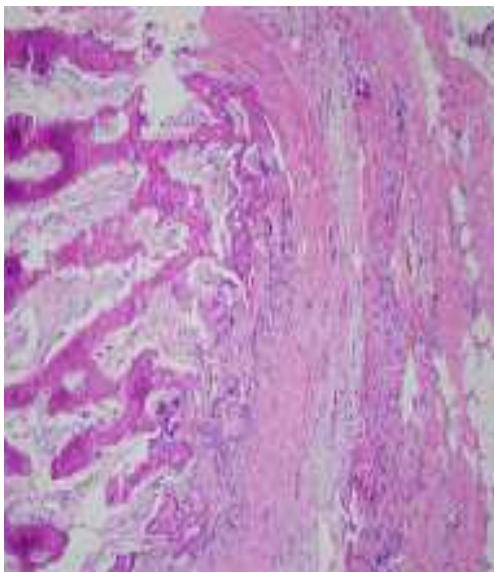
On examination she had diffuse mandibular and submandibular swelling without any facial asymmetry. Swelling of the bilateral cheek and mandible was tender to palpation. Swelling erythema and tenderness were present over left forearm with decreased pronation, supination, elbow and wrist flexion. She was irritable, pyrexic (101°F), tachycardiac (140/min) and tachypnoeic (60/min). She had a normal term delivery with birth weight 3.2 kg. She was immunized to date and was on exclusive breast feeding.

Initial blood investigations revealed anemia (Hb 8.5 gm%), mild leucocytosis (16000/cumm), thrombocytosis, persistently elevated ESR (60-70 mm in 1<sup>st</sup> hour), CRP (20 mg/l), alkaline phosphatase (330-400 units). Blood and urine culture were negative. Liver function, kidney function and electrolytes were normal. VDRL of both mother and child were nonreactive. Radiograph (Figure 1) of the left forearm showed patchy asymmetrical periosteal new bone formation causing cortical thickening in diaphysis of both radius and ulna. Associated perifocal soft tissue thickening was seen. No osteolytic lesion was present.



**Figure 1: Radiograph during presentation showing perifocal soft tissue thickening with asymmetrical cortical thickening and deformed bone.**

Due to inflammatory parameters and associated radiologic pictures, osteomyelitis was initially suspected and parenteral antibiotics continued. As the response was poor we planned for an open biopsy. Histopathologic review (Figure 2) showed normal bone trabeculae coated with osteoblasts and myxoid periosteum. Marrow fibrosis and associated reactive bone formation consistent with early subacute changes in cortical hyperostosis seen.



**Figure 2: Histopathology showing normal bone trabeculae coated with osteoblasts and myxoid periosteum and also presence of marrow fibrosis.**

On review literature, these clinical findings along with anemia, thrombocytosis, increased alkaline phosphatase,

ESR, radiologic features and biopsy findings were found to be consistent with Caffey's disease. During hospital stay only supportive anti-inflammatory measures given. Forearm was splinted to prevent pathological fracture. Patient was discharged with advice of regular follow up and analgesic during acute phase. At 6 month and 1 year follow up she was asymptomatic with bony swellings both in forearm and cheeks significantly reduced. Routine blood investigations were within normal limit. Radiography showed corticomedullary differentiation (Figure 3, 4). On three year follow up no swelling, tenderness, hyperesthesia over left forearm. Full range of motion regained at wrist, elbow and pronation, supination. No complain over mandible. Radiography showed normal bone with good corticomedullary differentiation (Figure 5).



**Figure 3: Radiograph at 6 month follow up showing healing and bone remodelling.**



**Figure 4: Radiograph at 1 year follow up showing corticomedullary differentiation.**



**Figure 5: Radiograph at 3 year showing near normal bone.**

## DISCUSSION

George Roske in 1930 first described this disease in a patient after exclusion of TB, rickets, scurvy etc. But John Caffey with Silvermann recognized this as a special clinical entity and the name infantile cortical hyperostosis was assigned to it.<sup>1</sup>

Existence of two forms of this disease have been reported. One is in utero form and another is classic form. The in utero form has two sub types on the basis of age of onset. Onset before 35 week of gestation is highly lethal and after 35 week gestation is milder variety without much complication. USG can detect it prenatally but can be confused with osteogenesis imperfect.<sup>5,9</sup> The classic form has the average age of onset about 9 weeks, and cases in which the onset occurs after first five months of age are considered invalid by Caffey and others.<sup>10,11</sup>

Various mechanisms have been proposed for the manifestation of the disease. For prenatal form autosomal recessive and for post natal form autosomal dominant form has been suggested.<sup>6</sup> Allergic and infectious theories have been postulated. Another theory says inherited defects of arterioles of periosteum results in hypoxic periosteal damage.<sup>12</sup>

However the initial step is inflammation of periosteum and soft tissues surrounding it followed by subperiosteal

bone formation. Affected bones are thickened by subperiosteal new bone deposition. Early stage is called acute inflammatory stage with loss of periosteal subperiosteal boundary. Osteoblastic reaction with overlying muscle involvement is seen. Subperiosteal deposition with round cell infiltration to overlying edematous muscle and soft tissue seen. Subacute phase reestablishes periosteum as entity itself. Removal of extra bone from subperiosteal surface with medullary cavity dilatation is remodeling stage.

Diagnostic features are (1) Age between birth and 5 months, (2) Triad of irritability, swelling of overlying soft tissue and (3) associated mandible inflammation.<sup>13</sup> Associated fever, and anorexia are present. Swelling is painful with induration but absence of redness, warmth and suppuration. Mandible is most commonly involved site followed by clavicle, ulna and other long bones.<sup>2,3</sup> Isolated cases of entrapment neuropathy causing Erbs palsy and facial palsy have been reported in literature.<sup>14,15</sup> Sometimes pain is severe and movement of involved part is limited causing pseudoparalysis. Even torticollis, pleural effusion mandible asymmetry has been reported.<sup>16</sup>

Laboratory findings include raised ESR and Alkaline phosphatase level. Leucocytosis is common. Thrombocytosis, anemia, increased immunoglobulin level are seen.<sup>17</sup> No laboratory tests are specific, but diagnoses those are to be excluded are osteomyelitis, syphilis, scurvy, hypervitaminoses, Ewings sarcoma, metastatic tumors etc. Radioimaging is most valuable diagnostic study in Caffey's disease. Cortical hyperostosis that is new bone formation beneath soft tissue swelling are characteristic feature.

X-ray findings include mandible hyperostosis and margin of hyperdensity below the periosteum when thickens and produce cortical hyperostosis. Marginal irregularity is noted. Absence of metaphyseal changes seen in all hyperostotic bones. Chronic form shows medullary dilatation due to resorption of hyperostotic bones producing thin cortical walls. Bone scintigraphy should be done to distinguish between mono and poly ostotic disease. The scintigraphy image is characteristic.

Caffey's disease is mostly self-limiting disease and active manifestation regresses without treatment within 6 months to one year.<sup>8</sup> Indomethacin /ibuprofen/naprosyn is used in symptomatic cases. Caffey advocated the use of steroid for several cases. Prednisolone 20 mg daily for 3 days rescues a desperately ill patient in few days. So steroid use is restricted to non-responding cases.

After description of Caffey's disease in 1945 there were more than 100 cases reported in between 1945 and 1960. But after 1970 there was a sudden unexplained drop in incidence. Now this disease is very rare with only one case series and 20 case reports between 2000 and till date (Table 1).

**Table 1: Caffey's disease with 20 case reports between 2000 and till date.**

Author	Year	Presentation	Location	Treatment
Cooper et al.	2001	Fever with normal blood picture	Extremities, clavicle and mandible	Steroid and indomethacin
Caksen	2001	No fever, increased ESR	Both arms, both legs	Observation
Verma	2002	Fever with cheek swelling	Mandible	Oral steroid
Bykov	2003	Fever	Mandible	
Hasegaw	2003	Fever, increased TLC, CRP, ALP	Left tibia and femur	
Dutta	2004	Fever, increased CRP, platelet	Mandible, tibia	Ibuprofen, steroid
Suri	2004	No fever, pseudo paralysis, increased ESR, ALP	Mandible, forearm and leg	
Ly-Ba	2007	No fever, increased inflammatory markers	Left clavicle	
Shannon	2006	No fever, increased TLC, ESR	Tibia	
Kovacic	2007		Mandible	
Wong	2007	NO fever, increased TLC, ESR	Left mandible	Indomethacin
Kamoun-Goldrat	2008	No fever	Mandible, humerus, tibia	Observation
Kumar	2008	Increased immunoglobulin, platelet		
Kaissi	2009	No fever	Mandible, right radius, leg	
Lo	2010	Increased ESR, platelet	Extremities, mandible	Indomethacin
Parnell	2010	No fever, increased CRP	Both scapulae, mandible	Indomethacin
Kessler	2010	No fever	Mandible, extremities	Only follow up
Rangnath	2010	Increased ALP, ESR	Both tibia and femur	
Krishnamurthy	2011	Increased ESR, TLC	Mandible, both tibia	Ibuprofen
Agrawal	2011	Fever, swelling of limbs, thrombocytopenia	Diaphysis of all long bones with sparing of mandible and clavicle	Symptomatic treatment
Mahalingam	2013	Fever, anemia, thrombocytosis, increased TLC, ESR, CRP	Arm and clavicle	Anti-inflammatory measures
Pagano	2015	Presented with neuroblastoma from local site of diagnosed Caffey's disease	Clavicle	Supportive measure

## CONCLUSION

Infantile cortical hyperostosis, though a rare disease, is self-limiting and can mimic osteomyelitis. Presentation may be as in our case fever, soft tissue swelling, irritability. Keeping this condition in mind, a good clinical examination, plain radiograph and if necessary biopsy are sufficient for diagnoses. A review of cases of Caffey's disease are also presented.

## Abbreviations

ESR - Erythrocyte sedimentation rate  
CRP - C reactive protein  
ALP - Alkaline phosphatase

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