Case Report

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 Caffeys disease in a 5 month female child initially thought to be osteomyelitis of radius and ulna. We also present of summary of cases reported since 2000.

Keywords: Infantile cortical hyperostosis, Osteomyelitis

INTRODUCTION

Caffeys disease is a rare disorder of skeleton and surrounding soft tissues originally described by Caffeys and Silvermann in 1945 as a syndrome of unknown aetiology. It is an episode of massive superiostel newbone formation involving mandible, clavicle and diaphysis of long bones. 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 Caffeys 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 sub mandibular swelling without any facial asymmetry. Swelling of the bilateral cheek and mandible was tender on palpation. Swelling erythem 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 1st 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 periostal 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.
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.

ESR, radiologic features and biopsy findings were found to be consistent with Caffeys 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 1: Radiograph during presentation showing perifocal soft tissue thickening with asymmetrical cortical thickening and deformed bone.

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

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 follow up showing normal bone with good corticomedullary differentiation.
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. 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.\(^1\)\(^2\) Isolated cases of entrapment neuropathy causing Erbs palsy and facial palsy have been reported in literature.\(^14\)\(^15\) Sometimes pain is severe and movement of involved part is limited causing pseudoparalysis. Even torticollis, pleural effusion mandible asymmetry has been reported.\(^16\)

Laboratory findings include raised ESR and Alkaline phosphatase level. Leucocytosis is common. Thrombocytosis, anemia, increased immunoglobulin level are seen.\(^17\) 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 regress without treatment within 6 months to one year.\(^8\) 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).

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.\(^5\)\(^9\) 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.\(^10\)\(^11\)

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.\(^6\) Allergic and infectious theories have been postulated. Another theory says inherited defects of arterioles of periosteum results in hypoxic periosteal damage.\(^12\)

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

**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.\(^1\)

Figure 5: Radiograph at 3 year showing near normal bone.
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 disease are also presented.

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

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REFERENCES