Infantile Cortical Hyperostosis (Caffey Disease): A Possible Misdiagnosis as Osteomyelitis

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Abstract

Infantile cortical hyperostosis (Caffey disease) is characterized by spontaneous episodes of subperiosteal new bone formation along one or more bones commencing within the first 5 months of life. It is featured by acute inflammation of periosteum and overlying soft tissue which causes fever. Sometimes it can present as fever without focus. Generally, the diagnosis may be delayed as this disorder mimics a wide range of diseases including osteomyelitis, hypervitaminosis A, scurvy, bone tumors and child abuse. The purpose of this case report is to emphasize the high index of suspicion of this disease in our country, and remind the clinicians about how a full history is important to reach the diagnosis.

Keywords: Caffey disease; Infantile cortical hyperostosis; Osteomyelitis

Introduction

Caffey’s disease is a rare genetic disorder that classically presents with a clinical trial of fever, irritability, and soft tissue swelling, in an infant with a radio graphical picture of cortical thickening or bony expansion, bridging of bones across inter osseous membrane, marginal sclerosis, and hyperostosis of flat bones [1]. The mandible (75%), clavicles, and ulnae are the bones most frequently involved, others being long bones, lateral ribs and iliac bones, the skull being rarest [2]. Diaphyseal involvement and metaphyseal paring of long bones is noted giving a spindle-shaped appearance of bones. The clinical importance stems from the fact that certain pathological conditions such as osteomyelitis, bone tumors, scurvy, hypervitaminosis A, and battered baby syndrome can resemble the radiographic findings of Caffey’s disease, thus warranting exclusion.

Case Report

An 8th month-old Saudi boy presented on July 4, 2017 to the Emergency department of Prince Muhammad Bin Abdulaziz Hospital (National Guard Health Affairs) Medina, Saudi Arabia, with a history of sudden painful right jaws swelling for two days (Figure 1), no fever, history of trauma or insect bite. By physical examination there was mild diffuse firm, fullness and tenderness on the right jaw, the overlying skin was normal and no discharging in us or localized lymphadenopathy. On general examination there was mild tenderness in the right upper limb maximum in for ear mother wise was normal. Plain X-ray of upper limbs how ed no fracture line or bony abnormality. Patient was discharged home.
pain for three days (Figure 2). No history of trauma or fallen down.

On examination, the baby was alert but irritable. Vital signs were normal except for low grade fever. There was swelling in the right forearm with tenderness, hotness but normal range of movement on all joints. Other systemic examination was unremarkable.

He is a baby of medically free mother, full-term, product of uneventful pregnancy, normal vaginal delivery, his birth weight was 2.72kg. He was exclusively on breast feeding, normal growth and development, immunization up-to-date according to Saudi vaccination program. No history for recent travel. Family history was insignificant.

His initial Investigation done in the ER showed WBCs count of 14.6, neutrophils 37.60%, Lymphocytes, platelets 743×10^9/L, Hgb 10.3gm/dl, CRP 141.2, ESR 105mm/hr, alkaline phosphatase 323(normal). Other laboratory tests were within normal range. Blood culture revealed no growth.

X-ray marked ulnar & radial diaphyseal periosteal reaction sparing their metaphases & epiphyses, with soft tissue swelling around the forearm (Figure 3). However, there were no osteolytic lesions.

Orthopedic consultation done and advised to start empirical antibiotic for possible osteomyelitis. So, patient was admitted to hospital and started on amikacin and floxacin after 11 days of admission the patient still not improved, antibiotics changed to vancomycin and Ceftriaxone, and MRI of the right forearm pre & post contrast was done showed revealed marked radial & ulnar shaft periostitis with marked edematous /inflammatory changes in the surrounding structure (Figures 4-6). X-ray of the right mandible was done and it demonstrate marked periosteal reaction. Other skeletal survey to rule out child abuse showed no obvious periosteal reaction around the other flat bones (clavicle, scapula, ribs, clavicles or iliac bones). After what we found along with the non-improvement of the child condition we changed the diagnosis from osteomyelitis to Caffey’s disease based on this clinical background (Bone biopsy and genetic study not done due to unavailability). The baby was discharged on Ibuprofen 10mg/kg PRN then here turned to pediatric clinic for follow up after four weeks of discharge with complete recovery.

Figure 3: Radiograph of the right forearm showing marked ulnar & radial diaphyseal periosteal reaction sparing their metaphases & epiphyses, with soft tissue swelling around the forearm (Caffey disease).

Figure 4: MRI. Axial T2 image at mid right forearm with fat suppression revealed marked radial & ulnar shaft periostitis with marked edematous /inflammatory changes in the surrounding structure.

Figure 5: MRI. Axial image at mid right forearm, pre contrast.

Figure 6: MRI. Axial image at mid right forearm post contrast. Demonstrate marked enhancement of the radial and ulnar periosteum (periostitis) as well as the surrounding soft tissue reaction (Caffey disease).
**Discussion**

Caffey disease, also known as Infantile Cortical Hyperostosis is a self-limiting disorder. It was first reported as a disease entity by Caffey and Silverman in 1945 [3]. The manifestations include irritability, swelling of the overlying soft tissue and cortical thickening of the underlying bones [1]. The swelling is painful with a wood like in duration but with no redness or warmth, thus suppuration is absent. There are usually nother signs and symptoms. Mandible is the most commonly involved site followed by scapula, clavicle, ribs and long bones. There are usually nother signs and symptoms [4-5]. The pain can be severe and canals result in pseudoparalysis. Other rare clinical findings included dysphagia, nasal obstruction and proptosis [6-8]. Laboratory findings include elevated ESR, and in some patients’s high alkaline phosphatase, thrombocytosis, anemia and raised immunoglobulin levels [9,10].

The exactetiology of this condition is still unknown [11]. Most cases are sporadic, but a few familial cases with autosomal dominant and recessive patterns have been described [12]. Among the proposed causes are, infections, immunological defects and genetic abnormalities. Others studies discovery of a gene in 3 unrelated families with autosomal dominant inheritance (geneCOLIAL, 17q21) which encodes Alpha-1 chain of TypeI collagen, has raised some doubts whether some cases area type of Collagenopathy, like Osteogenesis imperfect [13,14]. Similar conditions have also been reported following prolonged treatment with Prostagland in (PGE1) for maintaining ductal patency in infants with cyanotic heart disease [15,16]. Two forms of Caffey disease have been described in literature, a classical mild infantile form (ICH) delineated by Caffey and Silver mananda severe form with prenatalonset. The incidence of the disease is unknown. A total of 44 cases have been reported with the severe prenatal on set of Cortical Hyperostosis [17].

**Figure 7:** Radiograph AP & lateral view of the right mandible demonstrate marked periostral reaction.

Radiography is the most valuable diagnostic study in Caffey disease. Cortical new bone formation (Cortical Hyperostosis) beneath the regions of soft tissues welling in the diaphysis, sparing metaphysis and epiphysis is the characteristic feature [15]. While no laboratory tests are specific for diagnosis of ICH, the important differential diagnosis are to be excluded are osteomyelitis, chronic hyper vitaminosis A, bone tumor; scurvy, child abuse and prolonged Prostagland in (PGE1) infusion [15-19]. Consciousness of the presence of the Caffey disease is an uncommon condition but its regular clinical radiological outline will help patients. So, the Patients will no longer be forced to undergo needless investigation and medication Figures 7 & 8.

**Conclusion**

The condition should be taken under consideration when diagnosing this disease, that too with high level of uncertainty. A comprehensive history, clinical estimation, requisite laboratory inspection is needful to set aside the differential diagnoses. Plain radiography is adequate to assure the diagnosis in roughly every case.

**References**


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