Case Study

Caffey Disease mimicking as an osteomyelitis of ulna: A Case Report

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ABSTRACT

Caffey disease or Infantile Cortical Hyperostosis (ICH) is a rare and mostly self-limiting condition affecting young infants. A 5-month-old baby boy presented to our hospital with complaints of fever, swelling in left forearm and irritability for 3 days. Initially he was treated for osteomyelitis and later it turned out to be Caffey disease. It is characterized by acute inflammation of the periosteum and the overlying soft tissue and is accompanied by systemic symptoms of irritability and fever. Diagnosis may be delayed as this disorder mimics a wide range of diseases including osteomyelitis, hypervitaminosis A, scurvy, bone tumors and child abuse.

Keywords: Caffey disease, Osteomyelitis, Infantile cortical hyperostosis

INTRODUCTION

Caffey disease or Infantile Cortical Hyperostosis (ICH) is a rare disease of unknown etiology and mostly self-limiting condition affecting young infants. It is characterized by acute inflammation of the periostium and the overlying soft tissue and is accompanied by systemic symptoms of irritability and fever. Diagnosis may be delayed as this disorder mimics a wide range of diseases including osteomyelitis, hypervitaminosis A, scurvy, bone tumors and child abuse. The emphasis here is to remind pediatricians about the various presentations of the disease. A high index of suspicion is required in a typical clinical setting to identify the disease.

CASE REPORT:

A 5-month-old baby boy was admitted to our hospital in January 2018, with complaints of swelling in left forearm, fever and irritability for 3 days. There were no accompanying respiratory, gastrointestinal or urinary symptoms. He was born at term by lower segment caesarean section due to breech presentation. The birth weight was 3 kg with no history of perinatal complications. His immunization status was up to date. He was on breast feeding and vitamin D supplementation as per recommendation. There was no history of recent travel or trauma. There was history of similar complaints in last month which lasted for 2 days and resolved, spontaneously.

On examination, the baby was active, alert and playful. Pallor was present. His temperature was 101'F, pulse was 135/minute, respiratory rate was 25/minute and BP was 80/54 mm of Hg. There was no facial dysmorphism. His weight was 7kg, length was 62cm and his head circumference was 40.5cm (around the 50th centile). The overlying skin of left elbow was warm and the swelling was tender. Rest of the systemic examination was unremarkable.

The child was admitted in the ward, relevant laboratory investigations including blood culture were sent and IV antibiotics were started. Results of investigations revealed Hb 6.9gm/dl, WBC count 15,900/cmm with 35% of neutrophils and 59% of lymphocytes, platelet count was 6,82.000/cmm and CRP 66.5mg/l. A peripheral blood smear showed microcytic hypochromic red cells and thrombocytosis. His X ray of left forearm and local part ultrasonogram showed moderate degree of periosteal thickening of ulna with irregular cortex and soft tissue swelling, suggestive of acute infantile osteomyelitis of ulna (Figure 1).

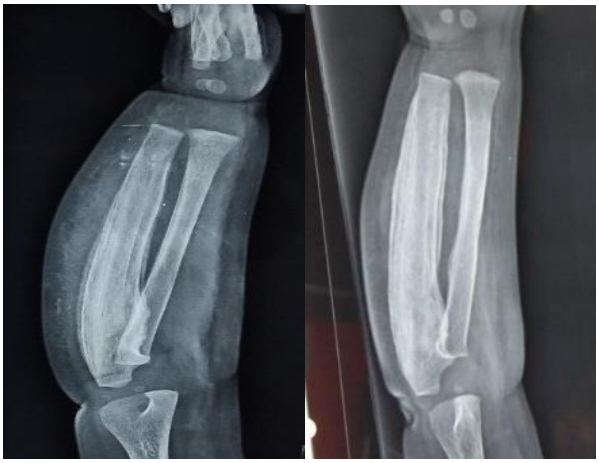

Figure 1 Figure 2

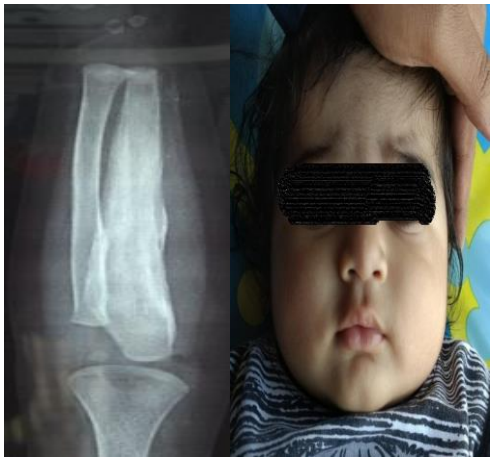

Figure 3 Figure 4

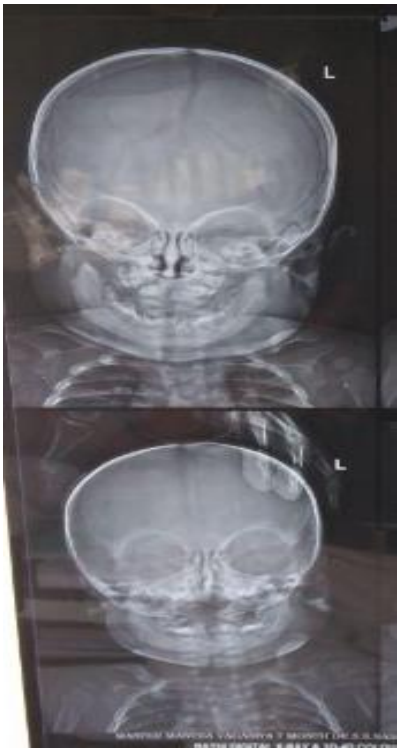

Figure 5

So, child was treated for osteomyelitis. He became afebrile on the third day of admission, His blood culture was sterile. His repeat laboratory investigation showed improvement with Hb 7gm/dl, WBC count 12,900/cmm with 28% neutrophils and 67% of lymphocytes, platelet count 7,00,000/cmm, CRP 36.6mg/l, procalcitonin 0.12ng/ml (Normal range 0.1-0.5ng/ml),ESR 48mm/hr, Sickling test negative, SGPT 19iu/l, Serum creatinine 0.44md/dl and ALP 236 iu/l.

In view of repeated swelling of the frorearm, thrombocytosis and negative procalcitonin we thought of the possibility of Caffey disease. So, we stopped IV antibiotics and carried out regular follow up of the child.

His laboratory investigation repeated after four weeks showed Hb 7.5gm/dl, WBC count 15,000/cmm with 28% of neutrophils and 65% of lymphocytes, platelet count 5,95,000/cmm and CRP 22mg/l. X-ray of the forearm done again also showed marked regression of the periosteal lesion of the ulna (Figure 2 &3).

After 6 weeks the child again came with low grade fever and bilateral swelling of the cheeks (figure 4). His x-ray of the mandible and skull showed periosteal thickening of mandible (right>Left side) and cortical thickening of left clavicle (figure 5). So, now the diagnosis of Caffey disease was confirmed and he was commenced on

NSAIDS(Ibuprofen) for 5days and gradually the swelling of the cheeks subsided. .Follow up after 2 years showed a normally growing child without any illness or relapse.

Discussion

Caffey disease, also known as Infantile Cortical Hyperostosis is a self-limiting disorder. It is characterized by a triad of systemic symptoms (irritability and fever), soft tissue swelling and underlying cortical bone thickening. It was first reported as a disease entity by Caffey and Silverman in 1945[1]. The exact etiology of this condition is still unknown[2]. Most cases are sporadic, but a few familial cases with autosomal dominant and recessive patterns have been described[3]. Among the proposed causes are infections, immunological defects and genetic abnormalities.

The existence of two forms of Caffey disease has been suggested, a classical mild infantile form (ICH) delineated by Caffey and Silverman and a severe form with prenatal onset[4, 5]. The condition has been described as rare with no sex or racial predilection. The classic form has an onset within the first 6 months of life. The manifestations include irritability, swelling of the overlying soft tissue that precedes the cortical thickening of the underlying bones, fever and anorexia. The swelling is painful with a wood like induration with mild redness or warmth and no suppuration. Mandible is the most commonly involved site followed by scapula, clavicle, ribs and long bones. All bones may be affected except the phalanges or vertebral bodies[5].

Laboratory findings include elevated ESR, high alkaline phosphatase, thrombocytosis, anemia and raised serum prostaglandin E levels[5,6]. Radiography is the most valuable diagnostic study in ICH. Cortical new bone formation (Cortical Hyperostosis) beneath the regions of soft tissue swelling is the characteristic feature. While laboratory tests are nonspecific for the diagnosis of ICH, the important differential diagnoses that are to be excluded are osteomyelitis, chronic hypervitaminosis A, bone tumour, scurvy, child abuse and prolonged PGE1 infusion[5,6,7,8].

Caffey disease is mostly self-limiting and resolves within six months to one year and may not need any treatment[9]. However, Ibuprofen, Indomethacin or Naproxen could be used in symptomatic cases. Steroids can be administered if there is poor response to these drugs. In our case, Ibuprofen was used and the outcome appeared to be satisfactory. In some cases, the bone lesions can recur suddenly at their original sites or at newer sites and can have an unpredictable clinical course with remissions and relapses[5,9,10].

Conclusion: Caffey disease or infantile cortical hyperostosis, though a rare entity, is self-limiting and can mimic osteomyelitis. Presentation may be as in our case with fever, soft tissue swelling and irritability. Keeping this condition in mind, a through clinical examination and plain radiography are sufficient for a definitive diagnosis.

Abbreviations: BP-Blood pressure, IV- Intra venous, Hb-Hemoglobin, WBC-White blood cells, ESR - Erythrocyte sedimentation rate, CRP - C reactive protein, SGPT- Serum glutamic pyruvic transaminase, ALP - Alkaline phosphatase

Conflict of interest: The authors declared that they have no conflict of interest.

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