Caffey's Disease (Infantile Cortical Hyperostosis)

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Introduction

A one year three month old girl, of non-consanguineous parentage admitted due to low grade fever, irritability, excessive cry, gradual swelling and bowing of both the lower limbs lasting four months. The swelling had a wood like induration but no redness or warmth or suppuration. Both the lower limbs were painful with pseudoparesis (Figure 1). Her perinatal and neonatal period was uneventful and no history of physical abuse. Investigations showed, Hb%: 10gm/dl, TLC: 8,200/cumm, Platelet count: 3 lakhs/cumm, ESR: 60 mm in 1st hour. Blood culture revealed no growth. Serum alkaline phosphatase was 730 IU/l, Ca++ 9.6 mg/dl and phosphorus: 4 mg/dl. VDRL serology of the mother was negative. Bone radiographs revealed periosteal elevation, new bone formation, cortical hyperostosis and fusion of tibia and fibula of both sides (Figure 2). The child was treated with syrup ibuprofen and there was remarkable improvement in pain, tenderness and swelling within two weeks.

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How to cite
Paria P, Haldar RC, Ghosh S. Caffey's Disease (Infantile Cortical Hyperostosis). J Nepal Paediatr Soc 2015;35(3):312-313.

doi: http://dx.doi.org/10.3126/jnps.v35i3.13080

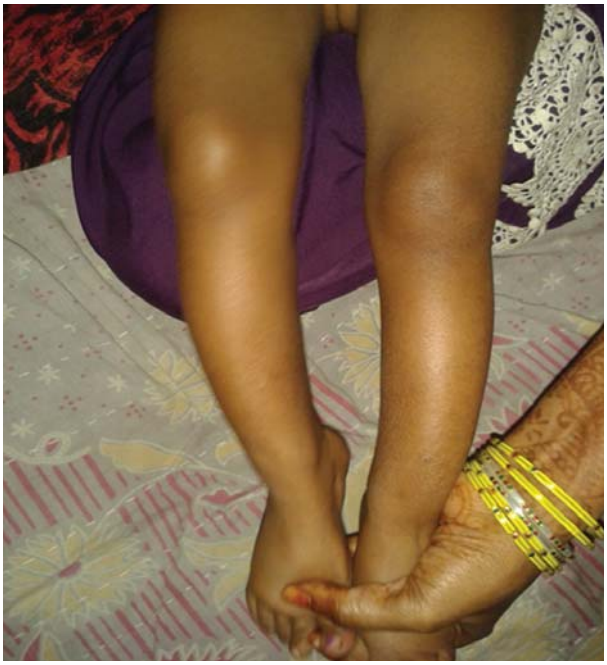

Fig 1: Showing swelling and bowing of both the lower extremities

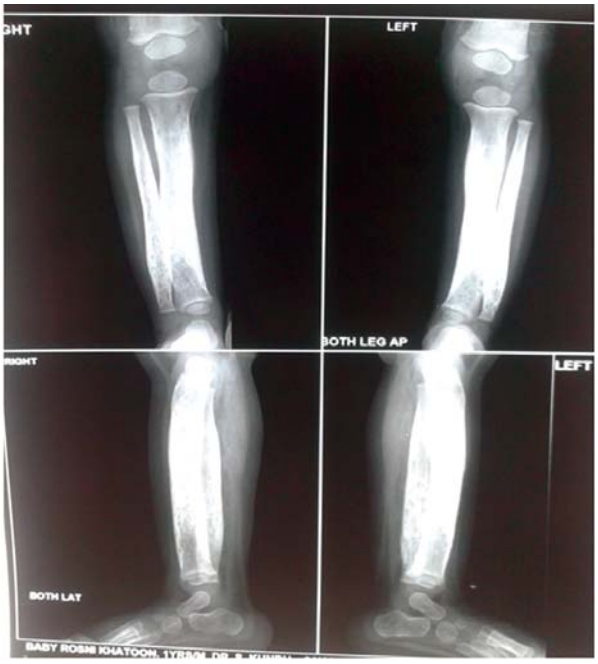

Fig 2: X-ray showing cortical hyperostosis, periosteal elevation, new bone formation and fusion of tibia and fibula of both sides.

Discussion

Caffey disease or Infantile Cortical Hyperostosis (ICH) is a rare and mostly self limiting condition affecting young infants less than five months of age, though has also been reported to occur in utero. It is often sporadic, but both autosomal dominant and autosomal recessive inheritance have been reported[1]. The most common bone involved include the mandible (75%), clavicle and ulna. Prenatal disease is characterized by typical bone lesions, polyhydramnios, hydrops fetalis, prematurity, and high mortality. Onset in infancy is manifested by systemic changes of irritability, fever and anorexia, along with inflammation of the periostium and the overlying soft tissue[2,3]. Diagnosis may be delayed as this disorder mimics a wide range of diseases including osteomyelitis, congenital syphilis, hypervitaminosis A, scurvy, bone tumors and child abuse[1]. Complications are unusual but include psudoparalysis, torticollis, mandibular asymmetry, bone fusion etc. Treatment is palliative with NSAIDs, and sometimes steroids [4]. Prognosis is good and disease resolves without any sequelae in 6–9 months. Early recognition helps to avoid unnecessary treatment.

References

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