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INFANTILE SCAPULAR CAFFEY DISEASE-A CASE REPORT

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ABSTRACT

Caffey disease is congenital and genetic musculoskeletal disease that most commonly occur in babies. The condition is self limited without a defined cause. It is marked by an acute inflammation of the periosteum and is characterized by systemic changes of irritability and fever. The diagnosis of Caffey disease is often challenging as it is a genetic abnormality. It impersonate certain diseases mainly osteogenesis imperfecta, Ehlers-Danlos syndrome, hypervitaminosis A and Hurler syndrome. In most cases physician diagnose the disease through physical examination. The condition resolves within six months and no specific treatment is required. NSAID's such as Indomethacin and Acetaminophen may be given to the patient as palliative treatment. We describe a case of one month old infant presented with swelling over the right shoulder.

KEYWORDS: Caffey Disease, Infantile Cortical Hyperostosis, Periosteum.

INTRODUCTION

Caffey's disease or infantile cortical hyperostosis is a benign, rare, proliferating bone disease affecting infants. Caffey and Silverman first reported this disease as a distinct entity in 1945 which is a selflimiting condition.[1,2] The disease mainly affects the jaw, shoulder blades, collarbones and shafts of long bones and by an excess new bone formation (hyperostosis).

Children are usually presented with tender and painful soft tissue swelling, erythema, fever and irritability. Studies shows that 80-99% of the patients with Caffey disease experience

symptoms such as cellulitis and cortical irregularity. 30-79% experiences behavior abnormality, fever, hyperesthesia and periosteal thickening of long tubular bones.

The cause of this condition is undefined but immunologic defects and viral causes are generally explained.[3] The exact etiology is however still unknown.[4] Both sporadic and familial forms exist.[5] The pathological phases are early, sub acute and late.

CASE REPORT

A 1 month old female infant was admitted in the inpatient department with twenty days history of swelling behind the right shoulder (scapula) associated with irritable cry while handling or lifting the baby. The child shows paucity of movement of the right upper limb and flexion of fingers. Patient was presented with low grade intermittent fever. The birth weight of the baby was 2.85 kg and the immunization status was up to date.

On general examination the baby was alert and irritable. The weight of the baby was found to be 3.96 kg and 50 cm height. There were no pallor, icterus, cyanosis, clubbing or pedal edema. The vitals were within the normal range. Laboratory examination showed leucocytosis with thrombocytosis. CRP (20mg/L) and ESR (80mm/hr) levels were also elevated. USG of the right shoulder was normal. MRI scan showed findings suggestive of infantile cortical hyperostosis.

The baby was treated with Inj. Moxclav (amoxicillin and clavulanic acid) 150mg, Syrup Ibugesic plus (ibuprofen and paracetamol) along with other supportives. The child became asymptomatic and the swelling of the right scapula reduced significantly. The child was reviewed for 6 months and showed normal growth.

DISCUSSION

Caffey's disease, also called infantile cortical hyperostosis (ICH), is a bone disorder characterized by excessive bone formation that most often occurs in babies. In 1945 Caffey and Silverman reported this rare syndrome which they called infantile cortical hyperostosis, following an earlier report by Roske, 1930. Epidemiological data revealed that the disease occurs in 3 infants per 1000 worldwide. 75-80% cases affects the mandible and 10% occurs in the scapula.The predominant signs and symptoms of Caffey's disease are irritability, pain, tenderness, hyperaesthesia, soft tissue swelling and redness associated with fever in the early stages. Other reported clinical findings include dysphagia[6] and nasal obstruction.

The gene associated with Caffey's disease is COL1A1. It undergoes mutation and these genes provide instructions for making the type I collagen. Type I collagen is the most abundant form of collagen in the human body. The mutation in COL1A1 gene that causes Caffey's disease replaces the protein building block arginine with the amino acid cysteine at protein position 836, his mutation in turn results in the production of type I collagen fibrils that are variable in size and shape.[7]

A combination of clinical picture, laboratory findings and imaging findings are used for the diagnosis of Caffey disease. Laboratory investigations may show an abnormality in erythrocyte sedimentation rate (ESR), C reactive protein (CRP) and alkaline phosphatease (ALP) levels. Radiographic examination reports of periosteal new bone formation mainly in the bones underlying areas of soft tissue swelling.

Caffey disease is self limiting and hence management is palliative. Painkillers such as Indomethicin or naproxen are usually prescribed for symptomatic treatment. In some cases corticosteroids are administered to speed up bone remodeling.

CONCLUSION

Caffey disease is a self-limiting condition of children. The aim of this case report is to highlight this disease entity to avoid unwanted and invasive testings. Correct diagnosis requires a proper knowledge of this disease along with a high index of suspicion. A notable history, clinical examination, laboratory studies, and radiographs are sufficient enough to confirm a diagnosis of this abnormality in most of the cases.

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