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Infantile cortical hyperostosis of the mandible. Caffey's disease

RESUMEN

Introducción: La enfermedad de Caffey o hiperostosis cortical infantil es un padecimiento auto limitable de etiología desconocida que se presenta durante los primeros meses de vida y cursa con formación de hueso nuevo perióstico. El hueso más involucrado es la mandíbula.

Material y Método: Se describe la presentación clínica, la cual incluye hiperirritabilidad, aumento de volumen de partes blandas y dolor de partes blandas, en los primeros seis meses de vida. Los hallazgos por imagen incluyen un incremento en la densidad del hueso, bien definida con engrosamiento de la cortical y bordes lobulados.

Conclusiones: La enfermedad de Caffey se demuestra mejor por TC, a pesar de que existen hallazgos descritos por IRM.

Palabras clave: Enfermedad de Caffey, tomografía computarizada, hiperostosis.

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Caffey's disease or infantile cortical hyperostosis is a self-limited disease of unknown etiology, which presents during the first months of life and courses with periosteal new bone formation. The mandible is the most frequently affected bone and its involvement is virtually pathognomonic.[1]

Most cases of the disease were reported from 1945 to 1968, with a gradual, but significant decrease in reported cases during the last decades.[2] The etiology of the disease is still unknown and multiple theories have been proposed, including: infections, immunologic defects, allergic reactions in collagen tissues, vascular, and genetic causes. The theory of genetic transmission by an autosomal dominant inheritance mechanism is supported by a few familial cases, but this is not the most common scenario.[2,6]

The clinical presentation includes a triad of hyperirritability, soft tissue swellings and painful firm soft tissue masses, occurring usually during the first six months of life. Some patients may develop fever, anemia and elevated erythrocyte sedimentation rate and serum alkaline phosphatase. The usual course of the disease is towards complete recovery and regression of the radiographic findings within a variable period of time that ranges between some weeks and several months.[2]

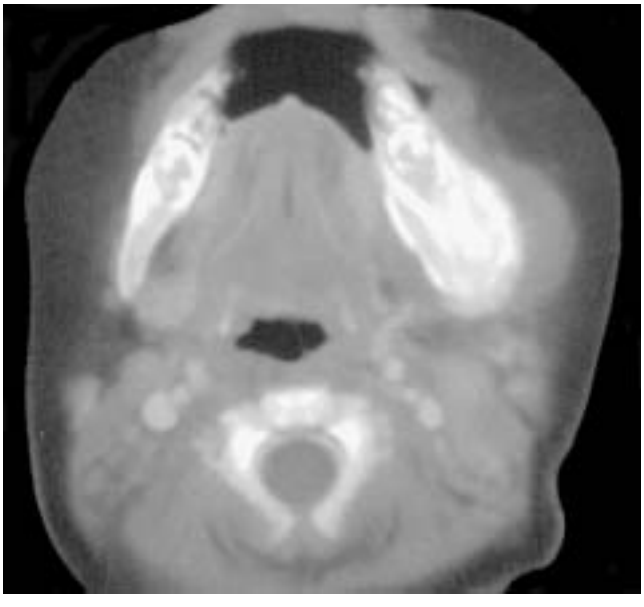

Figure 1. CT axial section of the mandible in a 6 month-old infant with Caffey's Disease demonstrated a localized periosteal laminated reaction in the angle of the mandible on the left (arrow). Associated adjacent soft tissue edema is also noted.

The mandible is involved in approximately 75%-80% of cases, either alone or with the involvement of other bones.[3] Pathologically, there is acute inflammatory reaction in the periosteum with infiltration of the connective tissues by polymorphonuclear leukocytes. The process then

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ABSTRACT

Introduction: Caffey's disease or infantile cortical hyperostosis is a self-limited disease of unknown etiology, which presents during the first months of life and courses with periosteal new bone formation. The mandible is the most frequently affected bone.

Methodology: The clinical presentation includes a triad of hyperirritability, soft tissue swellings and painful firm soft tissue masses, occurring usually during the first six months of life. Imaging findings include an ill-defined increased density of the bone with marked cortical thickening and a wavy contour.

Conclusions: This disease is best demonstrated on Computed Tomography (CT), although in MRI there are several changes.

Key words: CT, Caffeys disease, hyperostosis.

extends to the adjacent soft tissues and muscles, resulting in hyperplasia of collagen and fibrinoid degeneration. Osteoid trabeculae are formed and posteriorly, during a subacute phase, the periosteum reforms around both the old and new bone, the osteoid calcifies and appears subperiosteally. The last stage is of remodeling, with removal of the peripheral hyperostotic bone by resorption.[3,4]

Imaging findings include an ill-defined increased density of the bone with marked cortical thickening and a wavy contour. In the healing phase, a laminated periosteal reaction can be present.[3] This is best demonstrated on Computed Tomography (CT) *(Figures 1 and 2)*.

The Magnetic Resonance (MR) characteristics have also been described as thickening of the periosteum with low to intermediate signal intensity on T1 Weighted sequences and high signal intensity on the T2 Weighted sequences in the adjacent soft tissues, including muscles, secondary to edema.[5,6]

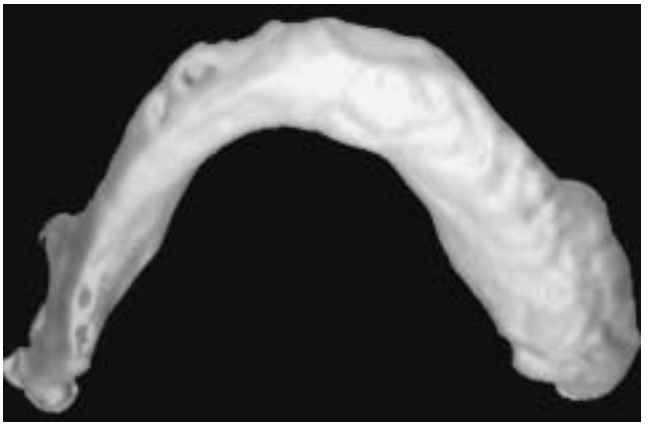

Figure 2. CT axial projection with 3-D reconstruction at the same level as in *Figure 1* demonstrated marked thickening of the angle of the mandible from periosteal reaction (arrow).

Continuing Medical Education Question

Which statement related to aggressive fibromatosis of the head and neck is incorrect?

Which of the following statements are correct as they relate to Caffey's Disease?

1. Most commonly occurs in the mandible.
2. The etiology is unknown, but is a self-limited disease.
3. Computed Tomography is the best diagnostic modality to confirm diagnosis.
4. Inflammatory periosteal reaction is the main pathological feature.

Answer: All of the above

References

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