Pediatric Radiology

Predominant or Exclusive Orbital and Facial Involvement in Infantile Cortical Hyperostosis (de Toni-Caffey's Disease)

Report of Four Cases and a Review of the Literature

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Abstract. Four patients who presented with predominant or exclusive face and orbital involvement by de Toni-Caffey's disease are reported. Facial manifestations of infantile cortical hyperostosis may be the first and sometimes the only manifestation of the disease. A mandibular involvement is almost always present and its characteristic appearance leads to the diagnosis of de Toni-Caffey's disease.

Key words: Cortical hyperostosis - Mandible

Although de Toni-Caffey's disease is well known and its diagnosis is usually easy, predominant or exclusive cephalic involvement is sufficiently uncommon, and may pose diagnostic difficulty. Four cases of such involvement form the basis of this report.

Cases Report

Case 1: A male infant (full term, normal pregnancy and normal delivery) of normal parents at six weeks of age had sudden onset of swelling of the lower part of his face with slight fever, normal blood count, normal serum amylase and negative complement fixation reaction for mumps. The family physician made a diagnosis of parotitis and prescribed Ampicillin. Spectacular improvement was obtained within 48 hours, but 6 days later, a relapse occurred with the same symptoms leading to admission to Hôpital Trousseau. Eighteen days after the onset of the disease his face was swollen, mainly on the right side without any modification of color or heat of the skin. This swelling was tender, extended posteriorly and did not fill the retromaxillary groove. It stopped anteriorly about 2 cm from the labial commissura and extended to the cheek. A firm, nonfluctuant swelling was palpated along the maxilla in the submaxillary fossa, mainly on the left side. The oral cavity was normal and Stenon's ducts apertures were not inflamed. The infant's general condition was good. There was no fever and the weight gain was satisfactory. The consulting physician ruled out parotitis and favored masseter myositis or mandibular angle exostosis. Electromyographic examination of masseter muscles was normal. Blood count showed 3,360,000 red blood cells 10,300 white blood cells with 41% neutrophilic (Polymorphonuclear lymphocytes) and 6% eosinophilic (Polymorphonuclear lymphocytes). Erythrocyte sedimentation rate was 50 mm at the first hour. Other laboratory studies were normal.

The radiological examination of the head revealed corticoperiosteal thickening of the mandible mainly in the vertical parts of the bone and also to a lesser degree in the horizontal parts; a characteristic picture of infantile cortical hyperostosis (Fig. 1). A radiological survey of the rest of the skeleton was negative. The infant was given a 3 weeks course of corticosteroids and left the hospital after several days of treatment. Forty-five days later, repeat radiographs showed almost the same mandibular findings but with better definition; there was no other bony involvement. When re-examined at 10 months of age, the child's mandible was normal. The final diagnosis was infantile cortical hyperostosis with only mandibular involvement.

Case 2: A three months old girl, without previous history, was admitted to Hôpital Trousseau. She had fever and a bulging fontanelle. She was discharged two weeks later without diagnosis. The only study available is a sedimentation rate of 60 mm at the first hour. During the following months she had periods of fever and variable swelling of the mandibular angles. When she was 5 1/2 months old, a physical examination showed a large head with swollen cheeks and a small body. Both mandibular angles were thick and hard. The rest of the examination was normal. The clinical diagnosis was that of infantile cortical hyperostosis. The blood count was normal and the sedimentation rate was 44 mm the first hour. The radiological examination of the head showed the same findings as the previous case. During the following months, she had several periods of low fever and the sedimentation rate remained high. At the age of 3, her general status was good and there were no clinical or radiological abnormalities.

Case 3: An Algerian girl was well until the age of 1 month when she had several episodes of bilateral edema of the eye-
Fig. 1. Case 1: Typical corticoperiosteal thickening of the mandible.

Fig. 2. A Case 3: Soft tissue swelling around the mandible with discrete cortical thickening. The upper part of the right orbit also is thickened. B (Same patient as Fig. 2 A - 3 weeks later) More prominent hyperostosis of the mandible and hyperostosis of the right maxillary bones.