Infantile Cortical Hyperostosis, Caffey's Disease, Involving Two Cousins

David K. Yousefzadeh, M.D. 1, Patrick Brosnan, M.D. 2, and Joseph H. Jackson, Jr., M.D. 2

1 The University of Iowa Hospitals and Clinics, Iowa City, Iowa, USA
2 Driscoll Foundation Children's Hospital, Corpus Christi, Texas, USA

Abstract. Two cases of infantile cortical hyperostosis are presented in the hope of eliciting word of increased incidence seen by others. These two children are cousins born within 24 h of each other in separate cities, and their disease followed similar clinical courses. Their 14-year-old maternal uncle appeared to have had symptoms identical to those of Caffey's disease at three months of age. The apparent immunity of premature infants to infantile cortical hyperostosis is a previously unnoticed feature that we have come across in reviewing the literature. In analyzing the many theories proposed by different investigators as the pathogenesis of Caffey's disease, the following conclusions are reached.

I) Multiple causal factors may have to combine to produce Caffey's disease.
II) There may be a spectrum of Caffey's diseases with different pathogeneses.
III) Should the incidence of infantile cortical hyperostosis increase, all the previously suspected causal factors must be reexamined.

Key words: Cortical hyperostosis - Caffey's disease.

In recent years, there has been a widespread conviction that Caffey's disease has become rare. While not wishing to challenge this assumption, we are presenting two cases to elicit correspondence on an increased incidence.

Historical Review

An infant suffering from hyperirritability, thickening of the bones, and painful swelling of the overlying soft tissues was first described by Roske in 1930 [22]. In 1934, Caffey described an undiagnosed bone lesion, which, with Silverman, he reported in 1945 together with three additional cases as the classic description of what they named 'infantile cortical hyperostosis' [5, 6].

In 1888, West [32] reported siblings with a disorder that might be explained by symptoms of infantile cortical hyperostosis.

Incidence

In 1950, Sherman and Hellyer [25] reviewed all the roentgenograms in their files for the previous 20 years and could not find a single case of infantile cortical hyperostosis. The rarity of the disease prior to 1946 was not entirely due to mistaken diagnoses, and the disease is either an old one with increasing frequency or a new entity. Cayler and Peterson [8] in their report of 1956 suggested an incidence of three cases per 1,000 patients under six months of age.

In the United States of America and Europe, the prematurity rate has been shown to be between 5-15% [24]. Prior to 1967, at least 200 cases of infantile cortical hyperostosis had been reported [15], so that one would expect to find between 10 and 30 cases among the premature infants in the literature. Interestingly, we failed to find any such frequency. There have been only one or two cases seen in near term infants (Caffey, personal communication). The reason for this apparent immunity is not clear.

The recent gradual decrease in incidence is another mysterious feature of Caffey's disease. From 1964-1968, there were five to six cases every year diagnosed in the pediatric emergency area of Cook County Children's Hospital, Chicago, Illinois. During 1969-1976, the number per year gradually decreased from two to zero patients yearly [16].

It has been almost a generation since the epidemic
years of Caffey’s disease. If the familial predisposition reported in infantile cortical hyperostosis was not due to nongenetic factors such as infection, toxic reaction, etc., an upturn in the incidence of the disease should logically be expected.

Pathogenesis

Infectious Theory

Caffey et al. [7] describe the condition under infections of bone, because many patients have severe and