Infantile Cortical Hyperostosis: A Case Report

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ABSTRACT

This is a report of a 6 year old Saudi girl with the rare syndrome of cortical hyperostosis. She had the typical clinical and radiological findings of the disease. In addition, she had unusual long standing history of pseudoparalysis of the lower limbs, failure to thrive and recurrent epistaxis.

Infantile cortical hyperostosis is a rare proliferative bone disease affecting infants under the age of 6 months[1]. It was first reported in Europe in 1930[2]. It usually develops soon after birth, where affected infants present with irritability associated with swelling and tenderness of the long bones, mandible, ribs or scapulae. It is a self limiting disease, clearing spontaneously before the age of one year. It has no known aetiology, but it has familial tendency[3].

THE CASE

A 6 year old Saudi girl presented to our paediatric clinic at the age of three years with a history of inability to walk, crawl or stand since birth. She also had a history of irritability and painful swelling of her both legs since birth. There is no history of fever or trauma. None of her family had the same condition. Her other three siblings were well. The parents were first degree relatives. Her clinical examination revealed a well-looking child, in no distress, and normal vital signs. Her weight was 10.5 kg (5th centile), her height was 86 cms (5th centile) and her head circumferance was 47 cms (5th centile). The general examination was unremarkable. Local examination of her lower legs showed prominent, swollen, tender, echomotic, bony, non-fluctuant areas of both tibial shafts. There was no regional lymphadenopathy. Radiological examination showed periosteal reaction in the form of cortical thickening and sclerosis of the shafts of both tibias (Fig 1). Local skin biopsy was normal. Her initial laboratory testing revealed a haemoglobin of 8.4 gm/dl, haematocrit of 29.6%, MCV - 65.8, MCH - 19.3, White Cell count - 8200/mm^3 with normal differential count. Her erythrocyte sedimentation rate was 77 m/h and her platelets count was 220,000. The haemoglobin electrophoresis was normal and her serum iron was 18 mg/dl. Her PT and PTT were normal.

The patient was treated symptomatically using leg

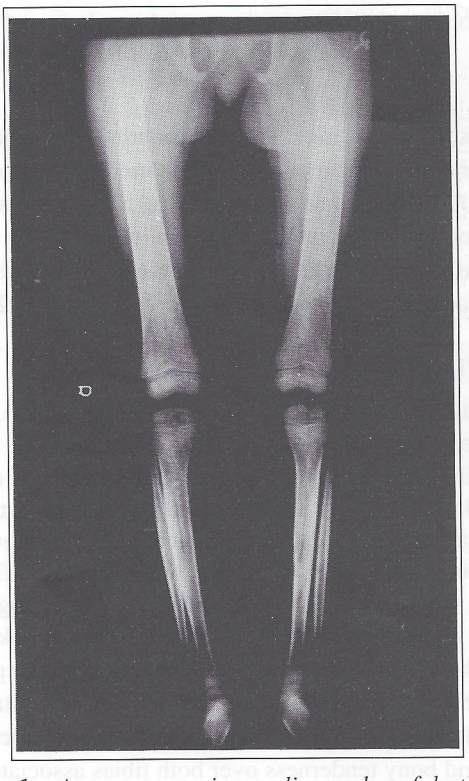

Figure 1: *Antero posterior radiographs of both tibia (age 3 years) shows periosteal reaction in the form of cortical thickening and sclerosis*

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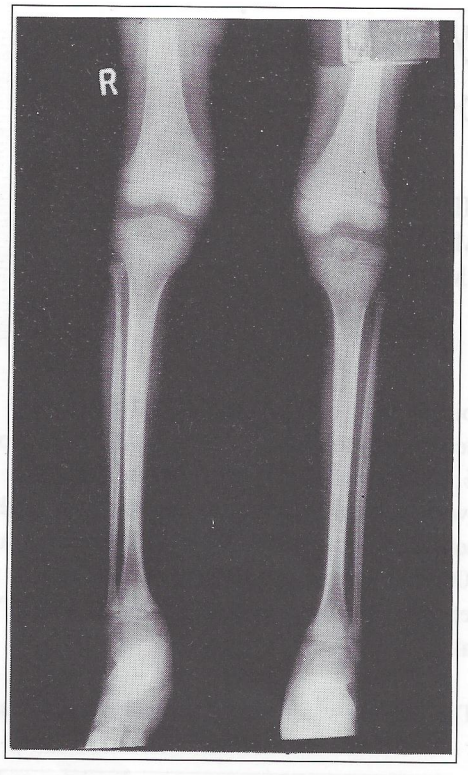

Figure 2: *After 18 months partial resolution of periosteal reaction with cortical thickening*

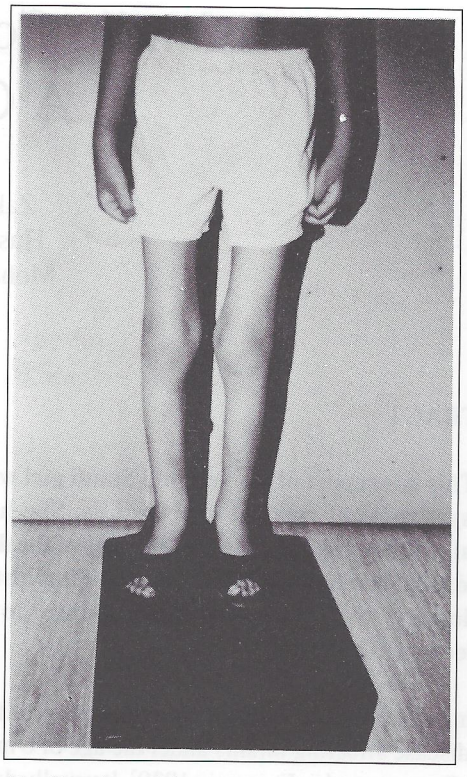

Figure 3: *Bowing of legs of the patient at the age of 6 years*

braces with analgesics. After 18 months of physiotherapy and rehabilitation programme, she started to walk normally. Her follow-up course was uneventful apart from mild few echomotic patches on both legs. Follow-up X-ray of both legs showed marked improvement in the cortical hyperostosis, with some bowing of her legs (Fig 2).

DISCUSSION

Infantile cortical hyperostosis is a rare disease. It was first reported by Roske in a seven weeks old baby[2]. Caffey and Silverman accounted the first definition of the disease in 1945[4]. This was followed by many other reports. The familial form of the disease has been recently reviewed by Newburg and Tampas[5]. In both the sporadic and familial types, there is a world-wide distribution affecting many racial groups. The condition usually starts weeks after birth, but may develop in utero and therefore can be present at birth[6]. In our index case, the history of irritability, inability to stand or walk, with recurrent echomotic swellings and bony tenderness over both tibias associated and the typical radiological findings which improved with time, the diagnosis of infantile cortical hyperostosis was established. None of the family members were affected indicating a sporadic incidence rather than the familial type. What was interesting in our case was the chronicity of the disease, since in most of the patients reported, the clinical as well as the radiological cause usually did not progress beyond the first or the second year of life[1]. The only late complications that could be encountered were bowing of legs with some discomfort (Fig 3).

CONCLUSION

We conclude that although this is a rare and benign disease, it has to be included in the differential diagnosis of other diseases with similar clinical and radiological picture such as osteomyelitis, rickets and scurvy[7].

REFERENCES

1. Maclachlan A, Gerrard JW. Familial Infantile Cortical Hyperostosis in a large Canadia Family. Can Med Assoc J 1984;130:1172-4.
2. Roske G. Eine Eigenartige Knochenerkrankung im Sauglingsalter. Monatsschr Kinderheilkd 1930;47:385-400.
3. Gerrard JW, Holman GH, Gorman AA, Morrow IH. Familial Infantile Cortical Hyperostosis. J Pediatr 1961;59:543-8.
4. Caffey J, Silverman WA. Infantile Cortical Hyperostosis. Preliminary report of a new syndrome. Am J Roentgenol 1945;54:1-16.
5. Newburg AH, Tampas JP. Familial Infantile Cortical Hyperostosis. An update. Am J Roentgenol 1981;137:93-6.
6. Barba WP, Freviks DJ. The Familial Occurrence of Infantile Cortical Hyperostosis in Utero. J Pediatr 1953;42:141-50.
7. Talab YA, Mallouh A. Hyperostosis with Hyperphosphatemia: A Case Report and REview of the Literature. J Pediatr Orthop 1988;8:338-41.