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PHALANGEAL MICROGEODIC SYNDROME IN CHILDHOOD

A Case Report

Y. MELLER,* J. BAR-ZIV,** J. GOLDSTEIN*** & G. TOROK*

Departments of Orthopedic Surgery,* Pediatric Radiology** and Pathology,*** The Soroka Medical Center, Faculty of Health Sciences, Ben-Gurion University of the Negev, Beer-Sheva, Israel

A rare case of avascular necrosis of bone of unknown etiology affecting several phalanges of one hand in a 12-year-old Israeli girl is presented. This entity has been described in the literature under the title of "Phalangeal microgeodic syndrome in childhood." Forty cases have been reported mostly from Japan. The clinical presentation is that of a sudden onset of chilblain-like appearance of the fingers. Characteristically, the disease occurs in the colder months of the year. The radiological picture is pathognomonic although an inexperienced observer might suspect an aggressive destructive lesion. The prognosis is excellent as the disease is a self-limited one and surgery is unnecessary.

Key words: fingers; osteolysis

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Phalangeal microgeodic syndrome in childhood was first described by Maroteaux in 1970. Since then 40 cases have been published of which 30 were from Japan (Akasaka et al. 1976, Hatanaka et al. 1978, Kaibara et al. 1981, Sugiwra et al. 1976, 1977). The clinical manifestations include: Spindle-shaped swelling, redness and local heat and tenderness of sudden onset occurring in one or several phalanges of one or both hands. Rarely it might affect the toes (Hatanaka et al. 1978). The mean age of afflicted children is 6½ years (range, 4 months to 13 years) and the ratio of boys to girls is 2.3 to 1. The etiology is obscure although one fact common to many of the published cases is the occurrence in the colder months of the year. This prompted Sweet & Smith (1979) to postulate that this entity is similar to frostbite injury but secondary to a less severe degree of cooling.

The purpose of this report is to present a case from Israel which occurred in wintertime. But it is important to note that winter in Israel is usually mild and the temperature almost never falls below freezing point.

CASE REPORT

K.R., a 12-year-old girl was admitted to our department in February, 1980, because of the sudden and simultaneous appearance of swelling of the right middle phalanges of the third, fourth and fifth fingers (Figure 1). Swelling and some pain were first noted 2 weeks prior to admission. There was no history of trauma. On physical examination there were no signs of a systemic illness. The middle phalanges of the right third, fourth and fifth fingers were swollen, slightly tender, red and warm. There was a full range of motion of all the fingers.

Laboratory data included a normal urinalysis, a white blood cell count of 6,500 with a normal differential, 130,000 thrombocytes and a Westergren sedimentation rate of 20 mm in 1 h. The routine biochemical screening tests and all the Rheumatological tests were normal. Tests for Syphilis, Tuberculosis, Brucellosis and other possible causes of osteomyelitis were negative. Chest X-ray and E.C.G. were normal. Tc 99 total body bone scan showed increased concentration of the isotope in the fingers of the right hand. A radiograph of the hand (Figure 2) showed multiple small lacunae in the middle phalanges of the involved fingers. There was also a slight widening of the affected phalanges caused by a periosteal reaction. A biopsy of the middle phalanx of the right third finger was done on February 20, 1980. Cultures of the specimen produced no growth of fungi, mycobacteria or routine bacterial flora.

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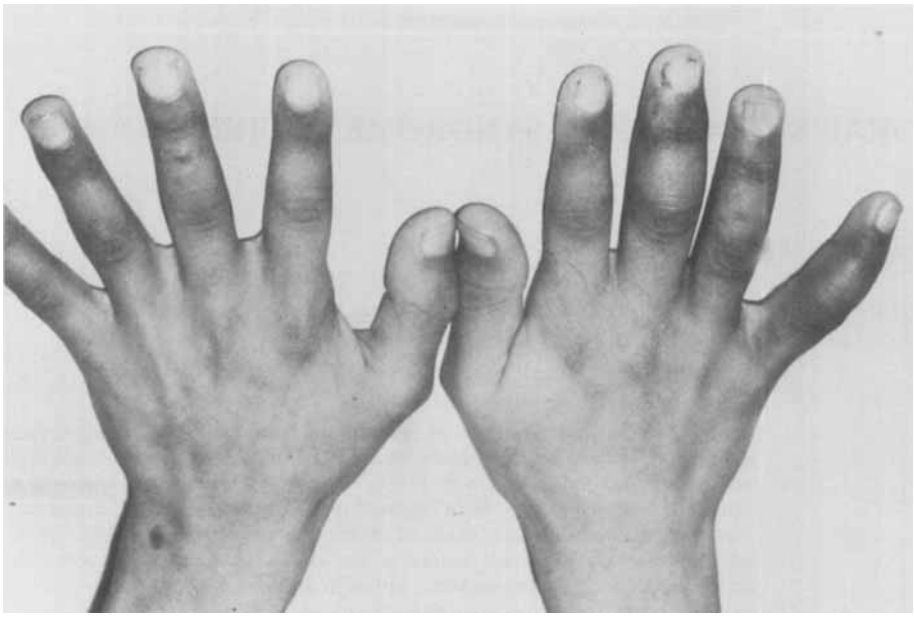

Figure 1. Swelling of the middle phalanges of the third, fourth and fifth fingers of the right hand.

Figure 2. Multiple small lacunar "microgeodic" osteolytic lesions in the cortex of the middle phalanges of third, fourth and fifth fingers of the right hand. Periosteal new bone formation and sclerosis are also present.

Pathology

Histologic sections of the two fragments of bony tissue obtained showed necrotic changes affecting both bone and bone marrow. Bone spicules were irregular, variably stained, and fragmented. Most lacunar spaces lacked nuclei. The edges of the bone spicules were frayed (Figure 3). The bone marrow seen between the spicules of necrotic cancellous bone showed changes consistent with coagulative necrosis (Figure 4). The histologic findings are those seen in aseptic necrosis of bone.

The patient became asymptomatic in a few weeks and when last seen, 1 year after the biopsy, her clinical condition is excellent and the radiological radiograph is almost normal (Figure 5).

DISCUSSION

The term "Microgeodic" was coined because of the occurrence of small lacunar-like osteolytic lesions of 1 mm in diameter simulating a "geode" in the radiographs. Apart from this feature, the affected phalanx shows some sclerosis in the diaphysis with rarefaction in the metaphysis, and slight widening due to periosteal reaction. Usually it involves the middle or proximal phalanges, but has never been described in the distal

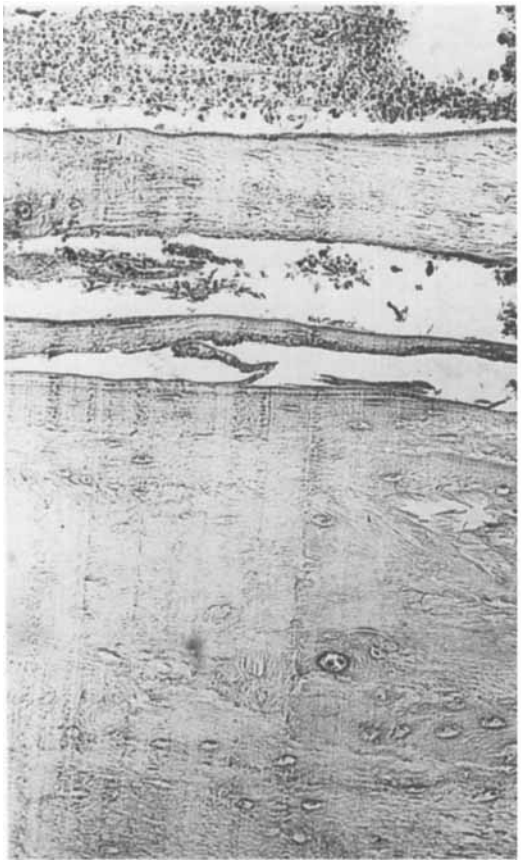

Figure 3. Bone spicules with numerous empty lacunar spaces showing loss of nuclei. The edges of the spicules are frayed and irregular (H + E × 40).

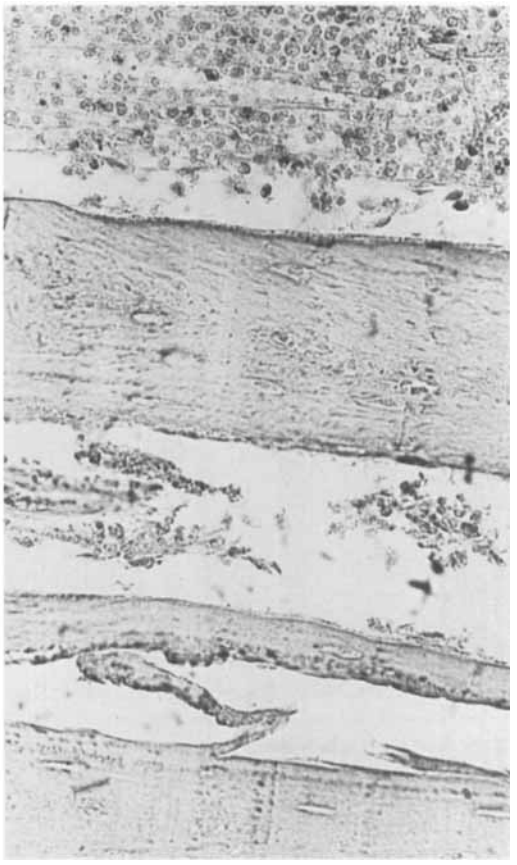

Figure 4. Photomicrograph demonstrating ischemic necrosis of both bone and bone marrow elements (H + E × 100).

phalanx. Although the radiological picture is quite characteristic, several other conditions should be considered in the differential diagnosis. These include: osteomyelitis, tuberculosis, syphilis, sarcoidosis and sickle-cell anemia (Kaibara et al. 1981, Sennara & Gorry 1978).

The etiology of this syndrome is not clear. Most of the cases have been described as occurring in wintertime in Japan and Europe where the temperature drops to low levels often below freezing point. Sweet & Smith (1979) postulated that this condition is analogous to frostbite injury, but secondary to a less severe degree of cooling. This is an appealing hypothesis but some features are contradictory. In frostbite injury, the distal phalanges are the most commonly afflicted parts of the hand excluding the thumb which may be protected by the fist. This location has never been described in phalangeal microgeodic syndrome in childhood. Furthermore, it seems logical that cold injury would affect any exposed phalanges but indeed this injury involves particular phalanges in an unexpected distribution. The mild climate in the Negev area of Israel where the temperature never drops below freezing point does not favor the cold-injury hypothesis.

Phalangeal microgeodic syndrome in childhood would seem to be more universal in distribution than previously thought, and is not necessarily confined to Japan and Europe. The case presented here demonstrates the occurrence of this condition in a more moderate climate such as is found in the Middle-East.

The clinical presentation is quite characteristic

Figure 5. The destructive process has disappeared. There is residual widening of the affected phalanges, sclerosis of the medullary canal and apparent partial closure of the epiphyseal plate.

and consists of the sudden appearance of spindle-shaped swelling of particular phalanges with minimal local signs of inflammation, pain, redness, tenderness and heat. As a rule there are no systemic signs present and laboratory findings are non-contributory.

The disease is a benign self-limiting process which subsides in a few weeks and results in complete recovery. We feel that biopsy is unnecessary in view of the typical clinical and radiological manifestations. If biopsy is performed the histologic findings consist of non-specific bone and bone marrow necrosis.

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Correspondence to: Y. Meller, M.D., Orthopedic Department, Soroka Medical Center, Beer-Sheva, Israel.