Osteomyelitis variolosa: a case report

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ABSTRACT

Osteomyelitis variolosa is an infection of bone and joints by smallpox virus variola major, most commonly in the elbows, wrists, ankles, hands, and feet. We report one such case in a 70-year-old woman who presented with deformities of the right knee, both elbows and ankles, and the left hand, and a history of childhood fever with rashes. Her lateral femoral condyle of the right knee was hypoplastic with patella baja. Her right elbow was ankylosed and her left elbow was dislocated with multidirectional instability. Her third and fourth metacarpals on the left hand were shortened. Both ankles were stiff with valgus deformity; both taluses were destroyed.

Key words: joint deformities, acquired; osteomyelitis; smallpox; variola virus

INTRODUCTION

Osteomyelitis variolosa is an infection of bone and joints by smallpox virus variola major, and occur in <5% of those infected by this virus. Although smallpox was eradicated in 1980, its orthopaedic sequelae still occur in countries where it was endemic. During smallpox epidemics, 0.25 to 0.5% of all patients or 2 to 5% of affected children had osteoarticular manifestation, most commonly in the elbows, wrists, ankles, hands, and feet. We report a 70-year-old woman with osteomyelitis variolosa involving the right knee, both elbows and ankles, and the left hand.

CASE REPORT

In July 2010, a 70-year-old woman presented with a 5-year history of pain in the right knee. The pain was dull and increased after household activities. There was no history of trauma. The patient had a hypoplastic lateral femoral condyle with patella baja (Fig. 1). There was no joint line tenderness, warmth, or laxity. Palpable crepitus was noted on knee movement. Knee range of motion was full. She also had bilateral elbow and ankle deformities attributed to a febrile illness at the age of 3 years. Her right elbow was fused and ankylosed in 90° of flexion and in a mid-prone position, with a dysplastic distal
humerus, ulnar shaft, and bowing of the radius (Fig. 2). Nonetheless, dexterity was good. The left elbow was dislocated with multidirectional instability, and the distal humerus was palpable inferiorly and the olecranon posteriorly (Fig. 2). The left hand showed third and fourth brachymetacarpia (Fig. 3). Both her ankles were stiff and displayed valgus deformity. Both taluses had collapsed and resorbed and had osteoarticular changes involving the distal tibia, fibula, and calcaneus (Fig. 4). She had facial and limb stigmata of variola pustules. Her hips, wrists, shoulders and spine were essentially unaffected. The patient declined any treatment other than conservative management for her right knee pain.

DISCUSSION

80% of osteomyelitis variolosa manifest symmetrically. Affected joints may subluxate, dislocate or become fixed in a grossly deformed position. In severe cases, the joint may be totally disorganised leading to abnormal mobility. The bones may be irregular, sclerosed, or thickened. In those who have contracted smallpox in childhood, deformities are usually bilateral and involve the epiphyses at their junction with the diaphyses of long bones. The most frequent sites are the elbows, ankles, wrists, hands and feet, with destruction of perimetaphyseal structures. Subsequent deformities in bone growth resulting from reparative ossification or distortion and/or reduction or cessation are probably due to the destruction of cartilage cells.

Figure 1  Hypoplastic lateral femoral condyle of the right knee, with patella baja.

Figure 2  (a) The right elbow is fused and ankylosed in 90° of flexion and in a mid-prone position, with a dysplastic distal humerus, ulnar shaft, and bowing of the radius. (b) The left elbow is dislocated with multidirectional instability, and the distal humerus is palpable inferiorly and the olecranon posteriorly.
In our patient, both elbows and ankles were involved. The lateral femoral condyle of the right knee was hypoplastic. Metacarpals of the left hand were shortened. All were characteristic of osteomyelitis variola sequelae. Smallpox was rampant in India during the period 1920 to 1975, and the patient’s medical history and clinical scenario retrospectively indicated that she had had smallpox. Unequal involvement of distal femoral epiphyseal plate is not uncommon in variola, which leads to uneven growth and premature fusion. The differential diagnoses include sequelae of bacterial septic arthritis, congenital dysplasias, and Caffey’s disease. Bilateral elbow involvement affecting all 3 bones is rarely seen in simple pyogenic osteomyelitis, and epiphyseal involvement is not a feature of bacterial osteomyelitis. The clinical presentations of Caffey’s disease are similar except that there is no rash. Our patient had stigmata of smallpox pustules on the face and limbs. Caffey’s disease often affects the mandible, clavicle, ribs, scapula, and other bones (which are rarely affected by smallpox), and it does not cause bony ankylosis as occurs with smallpox. Manifestations of osteomyelitis variolosa include short and stubby metacarpals or phalanges, unusual deformities of long bones, flaring of the metaphyses, flail joints, and ankylosis with precocious osteoarthritis.

REFERENCES