

*Case Report*

Osteomyelitis Variolosa Presenting As Ulnar Neuropathy - A Case Report

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ABSTRACT

Smallpox being eradicated in 1980 has led to the disease and its complications forgotten by the clinicians. Long term complications and sequelae of smallpox may still be evident among population who was affected by the disease earlier in their life. Blindness and joint deformities due to Osteomyelitis variolosa may still be seen though very rarely. We report a 48 year old gentleman who presented with features of compressive neuropathy of ulnar nerve following degenerative arthritis of the elbow joint due to Osteomyelitis variolosa.

Keywords: Smallpox, osteomyelitis variolosa, ulnar neuropathy.

INTRODUCTION

As Smallpox was eradicated in 1980 it is possible that the current generation clinicians may not be aware of the complications or sequelae of this disease. The long term complications / sequelae of smallpox apart from the disfiguring scars include blindness due to optic atrophy, corneal ulcer, retinal hemorrhages or arthritis and deformities due to osteomyelitis variolosa. These long term complications are still seen in survivors of the disease though very rarely but would be historic shortly.

The skeletal abnormality termed as osteomyelitis variolosa has been documented in 2 to 5% of children with smallpox. [1] The primary infection may be subclinical with manifestations being evident subsequently. [1] It commonly affects the elbows, wrists, hands, ankles and feet symmetrically. [2] It may be due to the separation of epiphysis, marked periosteal

reaction and premature closure of the physis leading to deformities and limb length inequalities. The same may lead to ankylosis of joints and malformed bones causing restriction of limb functions. Osteomyelitis variolosa has been rarely documented to cause fractures and neurological sequelae. Here we report a gentleman who presented with osteomyelitis variolosa and its sequelae resulting in ulnar entrapment neuropathy due to narrowing of cubital tunnel.

CASE REPORT

A 48 year old gentleman presented to the hospital outpatient department with complaints of pain in the right elbow with radiation to hand and deformity & numbness in the 4th and 5th fingers. His both elbow joints were deformed since his infancy for which surgical correction though advised was not performed. He was able to carry on with his routine activities in spite of the

deformity. He was diagnosed to have diabetes at the age of 43 years and was on oral medications. Clinical examination revealed bilateral fixed flexion deformity of 70° with further flexion only upto 110° in midprone position (Fig 1) of elbows. He had only jog of movements in pronation / supination. He had evidence of bilateral elbow joint arthritis and high ulnar nerve palsy with claw hand on the right side. Tenderness was noticed at the joint line and periarticular region. Investigations revealed poorly controlled diabetes and entrapment neuropathy of the ulnar nerve on right side at the elbow on Nerve conduction studies. Radiographs of the elbows showed features of degenerative arthritis with bilaterally symmetrical involvement and deformed lower end of humerus, upper end of ulna and radius (Fig 2 & 3). Other joints were spared. Further evaluation of history revealed that the patient had smallpox in the neonatal period and had recovered subsequently. His elder brother also had smallpox simultaneously but succumbed to the illness. Our patient was noted to have progressive deformity following the illness since infancy. Radiography of the hands of the patient confirmed the diagnosis of osteomyelitis variolosa documenting shortening of the third and fourth metacarpal bones bilaterally (Fig 4 & 5).

The ulnar nerve was found to be secondarily involved by the long standing elbow deformity. Patient was treated surgically with anterior transposition of ulnar nerve following control of blood sugar with insulin. The joint deformity was not corrected surgically. Intraoperative findings showed narrowing of the cubital tunnel with osteophytes and a ganglion arising from the joint capsule compressing the ulnar nerve. Postoperative period was uneventful and the patient was discharged on the third post operative day. At the time of discharge his radiating pain along the distribution of ulnar nerve was completely relieved. At 12 weeks

follow up, his claw hand has improved to near normal status.



Fig 1: Clinical picture showing both elbows with flexion deformity in midprone position.



Fig 2: Radiograph of elbow showing bony sclerosis and arthritis

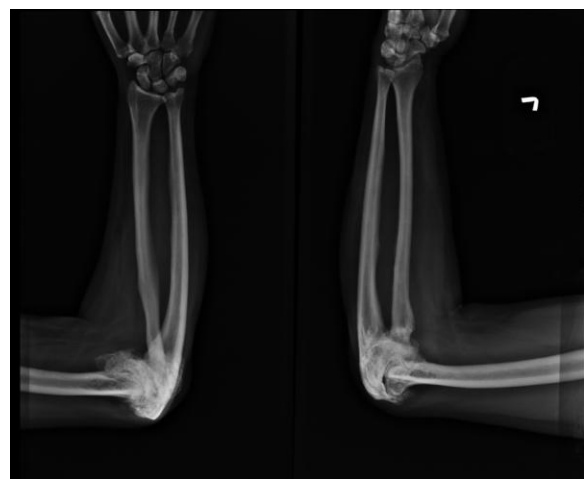


Fig 3: X Ray elbow – Lateral view



Fig 4: X Ray Left hand showing shortening of third and fourth metacarpals (Brachymetacarpia)



Fig 5: X Ray Right hand showing shortening of third and fourth metacarpals

DISCUSSION

The bony sequelae of smallpox following osteomyelitis variolosa affect most commonly the elbows (80%) and less commonly the hands, ankles or feet (20%).^[3] Axial joint involvement and other large joint involvements are extremely rare. Various studies have documented the predilection to elbows but the reasons are not clearly understood.^[4] It is postulated that the unique articular anatomy of the elbows with all the epiphysis around the elbow joint being intra articular might be responsible for deformities. Cockshott and MacGregor (1958) documented variolar inclusion bodies in sterile joint effusion postulating true viral affection of bone.^[5]

Viral invasion of bone may be rarely seen in smallpox, vaccinia and rubella.

The severity of affection of joints is not correlated with the severity of primary infection. Mild infection or subclinical course in childhood may present with joint deformities in later life. Our patient also had a mild infection with minimal cutaneous manifestations and sequelae but later with significant affection of bilateral elbows resulting in deformities. Retrospective assessment of patient's history with smallpox affection and death among family member in the relevant period in early life is in keeping with the diagnosis of osteomyelitis variolosa with its long term sequelae.

Hands may show short stubby fingers due to dactylitis.^[3] Shortening of the third metacarpal (Brachymetacarpia) with arthritis of third metacarpo-phalangeal joint is classically seen in osteomyelitis variolosa. However other metacarpals and phalanges may also be affected. Our patient though the physical appearance of hands was not showing any deformity other than clawing; on radiological examination demonstrated shortening of the third and fourth metacarpal bone. Figure 4 & 5 show that the third metacarpal is shortened than the second and the fourth is equating in its length to the fifth. It is noteworthy that normally in any individual the third metacarpal is the longest of all metacarpal bones and the fourth will be longer than the fifth.

Osteomyelitis variolosa should be considered for following reasons: (1) Symmetrical bone changes mostly affecting the elbows. All three bones of the elbow joint are involved. (2) Separation of an epiphysis due to transverse metaphysitis in varying degree. (3) Marked periosteal reaction. But all these typical findings may be seen in acute situation unlikely to be documented in the patients now a day. The radiography may show sclerosis of bone

surrounding the joint with relative preservation of the joint space in long standing cases. The deformities may be due to a result of reparative ossification or distortion as a result of destruction of cartilage cells. [6]

The differential diagnosis includes pyogenic osteomyelitis, salmonella osteitis in sickle cell anemia, leprosy, Caffey's disease and congenital dysplasias. Symmetrical affection is not common in pyogenic conditions. Leprosy with its characteristic radiological and neurological manifestations is distinguishable from variola affection. Caffey's disease affects mandible, clavicle etc which are not usually a manifestation of osteomyelitis variolosa. Bony ankylosis seen with osteomyelitis variolosa is not seen in patients with Caffey's disease.

Treatment for the condition may be surgical with attempted arthroplasty or osteotomy, especially to improve the function of the limb. Often it may not be treated aggressively due to limited benefits; unless there is severe impairment of limb functions. Cockshott and MacGregor (1958) described the condition as "an uncomfortable interference with function rather than an illness". Our patient underwent no bony correction but for the anterior transposition of the ulnar nerve to relieve the symptoms.

CONCLUSION

The case has been reported not because of its importance in diagnosis but for its clinical interest as its seldom seen and identified. Recognition of this etiology will prevent a great deal of unnecessary and unrewarding investigations. Osteomyelitis variolosa like any other manifestation of smallpox will become extinct in near future as the primary infection has been already eradicated.

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