Florid reactive periostitis ossificans of long bones and digits associated with reaction in a patient with leprosy

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Introduction and Summary

The first case of reactive periostitis was reported by Akarman in 1958\(^1\) and florid reactive periostitis/florid reactive periostitis ossificans was first described by Spjut & Dorfman in 1981.\(^2\) This entity of periostal reaction associated with soft tissue swelling, presents as a painful bone producing mass of the affected bone/digit and usually appears similar to a bone lesion that radiologically and clinically mimics an infectious or neoplastic disorder. The other names given to this condition by different authors include ‘pseudo malignant osseous tumour’ of the digits\(^3\) and ‘fibro-osseous pseudo tumour’ of the digits.\(^4\) However, Landsman \textit{et al.} recommended that authors adopt the term ‘Florid reactive periostitis’.\(^5\) The cause of florid reactive periostitis is not exactly known, but approximately one half of the patients give a history of trauma and only one case each of spontaneous florid reactive periostitis of the tibia and distal part of ulna has been reported in literature.\(^6,7\)

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The tubular bones of the hands and feet are the most common location, and periostitis ossificans in the long bones is anecdotal. Here we are reporting a case of reactive periostitis ossificans that was found in a patient with lepromatous leprosy and erythema nodosum leprosum (ENL)/Type 2 lepra Reaction. The case was managed conservatively (medical treatment) with complete recovery, and did not require surgical intervention as described previously by various authors.\textsuperscript{2,5,8} This is probably the first case of ‘Florid reactive periostitis ossificans’ that is being reported in association with leprosy reaction (Type 2/ENL).

The Case Report

A 15 year old boy of average built (weight 43 kg) was diagnosed with lepromatous leprosy of about 2 years duration in July 2006. His slit skin smears were examined for acid fast bacilli (AFB) and the bacillary index (B.I) was 3+ (mean of the 4 sites examined). He was started on WHO multi drug therapy (MDT) for multi-bacillary (MB) leprosy adult regime i.e. once a month on day 1; 2 capsules of Rifampicin (300 mg × 2), 3 capsules of Clofazimine (100 mg × 3) and 1 tablet of Dapsone (100 mg) and then once a day from day 2; 1 capsule of Clofazimine (50 mg) and 1 tablet of Dapsone (100 mg). After about 4–5 months of WHO-MDT, the patient presented in the outpatient department (OPD) of the institute with erythema nodosum leprosum (ENL)/Type 2 lepra reaction, of about 10 days to 2 weeks duration. The ENL lesions were the typical reddish brown skin eruptions which were painful and warm to the touch over many parts of the body. He also had mild to moderate fever (oral body temperature 99 to 100° F). He was then admitted in the ward and thoroughly examined and investigated for any possible precipitating cause of the ENL reaction and then put on oral corticosteroids: 15 mg per day of prednisolone equivalent along with anti-inflammatory analgesics. WHO-MDT was continued. His reaction subsided in about a week to 10 days and steroids were tapered off gradually over 4 to 6 weeks.

The second episode of reaction occurred about 3 months after the first episode, in about 3–4 weeks after withdrawal of corticosteroids with similar clinical presentation and was managed similarly.

The patient had another episode of reaction about 2 and half months after the second episode and this time he presented with the same coetaneous ENL eruption and fever along with bone and joint pain with mild soft tissue swelling of upper and lower limbs and also fingers and toes which developed after the onset of the reaction. This time, he required a slightly higher dose of corticosteroids i.e. Prednisolone 20 mg per day, to suppress the reaction, along with other anti-inflammatory drugs like aspirin 300 mg per day and chloroquine phosphate 250 mg twice daily. Supplementary calcium and Vitamin-D\textsubscript{3} was also given. His reaction and bone pain subsided gradually and steroids were tapered @ 5 mg every 2 weeks. However, he required a maintenance dose of 10 mg per day of Prednisolone for the suppression of ENL reaction and aspirin 300 mg to 500 mg on and off for bone pain. On September 2007, he presented with severe bone pains and swellings of left leg (shin) (Figure 1a), right forearm (medial aspect) below the elbow joint (Figure 1b) and some fingers and toes, of about 2 to 3 weeks duration, along with moderate to severe ENL reaction and fever.

He was again admitted and thoroughly investigated and radiographs were taken of the tender bones with swellings (left leg, right forearm and both feet). His blood investigations revealed normal serum phosphorus and alkaline phosphatase and serum calcium was slightly low: 7.2 mg/dl (normal range 8.4–10.2 mg/dl). His radiograph showed radio-opaque
masses (reactive periostitis ossificans) along the periosteum of left tibia (Figure 2a), right ulna (Figure 2b) and some of the digits and toes of hands and feet (Figure 2c).

He was put on complete bed rest with immobilisation of the affected limbs. MDT was continued along with anti-reactional drug corticosteroids 10 mg to 15 mg per day of prednisolone equivalent and inflammatory analgesics aspirin (300 mg to 900 mg) per day and nimesulide (100 mg to 200 mg) per day for the relief of pain and inflammation. Calcium supplements along with vitamin-D₃ were continued. His condition gradually improved, bone pains subsided and swellings disappeared. His ENL reaction also subsided and the dose required for its suppression could gradually be tapered off @ 5 mg to 2.5 mg every 2 to 4 weeks, over 16 to 20 weeks. The calcium and vitamin-D₃ was continued and serum calcium repeated after 12 weeks was found to be within the normal range. The radiographs were repeated after the remission of his condition and showed almost complete resolution of the radio opaque masses (Figures 3a, 3b and 3c).

He was released from treatment after the completion of the WHO-MDT regimen. The patient is still on regular follow-up and has not had a recurrence of reaction or bone problems.

**Discussion**

Florid reactive periostitis/florid reactive periostitis ossificans is the term used to describe a benign disease characterised by aggressive periosteal reaction and benign soft-tissue inflammation. Though it is said to be an uncommon bone lesion there are numerous reports in literature that refer to this disease with different names such as periosteal fasciitis,⁹ fasciitis ossificans,¹⁰ benign fibro-osseous pseudotumour,⁴ pseudomalignant osseous tumour of soft tissue,¹¹ and bizarre periosteal osteochondromatous proliferation.¹²
Typically, florid reactive periostitis is more common in adolescents and young adults (range 5–70 years) and women are more commonly affected than men. It presents as a swollen, painful lesion and appears to be a reactive process rather than a true neoplasm.\textsuperscript{2,6} The lesion is most common in the tubular bones of the hands and feet.\textsuperscript{6,7} In the hand, most lesions occur in the proximal phalanx; lesions in the distal phalanx, the metacarpals, and the thumb are rare.\textsuperscript{13,14} The etiopathology of the condition is not exactly known. However, some authors have proposed trauma as the basis of a hypothesis to explain the relationship between
florid reactive periostitis ossificans, periosteal osteochondromatous proliferation, and turret exostosis, suggesting there is a continuum among the three lesions with each representing a different stage of a proliferative process. According to this hypothesis an initial stimulus, possibly trauma, would cause subperiosteal bleeding, resulting in florid reactive periostitis.

**Figure 3.** (a) The post treatment radiograph of the left tibia showing complete resolution of the radio opaque mass and periostitis. (b) The post treatment radiograph of the right ulna showing resolution of the radio opaque mass and periostitis. (c) The post treatment radiograph of both feet showing resolution of the radio opaque mass and periostitis.
ossificans. In time this lesion would develop into periosteal osteochondromatous proliferation and later into turret exotosis. However, only approximately 50% of patients give a history of trauma. Cases of spontaneous florid reactive periostitis of the tibia and distal part of ulna have also been reported in literature. In this case, the possible hypothesis could be that erythema nodosum leprosum (ENL)/Type 2 lepra reaction, which causes severe vasculitis, could have caused some subperiosteal bleeding resulting in florid reactive periostitis ossificans as was explained by the previous authors, and the possible reason for the proliferative process could be recurrent episodes of ENL reaction. The clinical presentation was typical of the cases reported earlier with pain and soft-tissue swelling of the affected bones and associated tenderness and redness, indicating an inflammatory process. The case was managed conservatively with complete rest and immobilisation of the affected limbs, immediate medical intervention with anti-inflammatory analgesics, oral corticosteroids for the suppression of the ENL reaction and calcium supplementation along with vitamin-D as discussed above. The patient completely recovered and the bone density became almost normal after about 12 weeks of treatment (Figures 3a, 3b and 3c) and there is no recurrence of the condition and also of ENL reaction in a 1.5 year of follow-up after stopping treatment.

References