Case Report

Multifocal bilateral xanthogranulomatous osteomyelitis

Rahul Sapra, Pankaj Jain, Shubha Gupta[1], Rakesh Kumar

Abstract

Xanthogranulomatous osteomyelitis is a rare chronic inflammatory disorder which clinically resembles bone tumor. It is characterized histologically by the presence of a large number of foamy histiocytes admixed with lymphocytes and plasma cells. Xanthogranulomatous inflammation can involve any organ, with kidneys and gall bladder being the worst affected. Only anecdotal cases of osteomyelitis have been reported in the past with scant medical literature. However, all the earlier reported cases, except one, were unifocal lesions. The authors present a case of multifocal xanthogranulomatous osteomyelitis involving the left medial malleoli, left talus and the right cuboid bones.

Key words: Foamy histiocytes, xanthogranulomatous osteomyelitis, multifocal
MeSH terms: Osteomyelitis, xanthoma, bone diseases, infections

Introduction

Xanthogranulomatous inflammation, first reported in the genitourinary tract, can occur in many other organs such as the colon, appendix, pancreas, lymph nodes, gall bladder, salivary glands, endometrium, ovaries, fallopian tubes and the vagina. It may rarely affect lung, brain or the bone. Delayed type hypersensitivity reaction of cell mediated immunity has been implicated in its pathogenesis. Histologically, it is characterized by abundant foamy periodic acid-Schiff-positive histiocytes, containing abundant granular, eosinophilic cytoplasm in the initial stages, giant cells, fibrosis and calcification together with polymorphonuclear leukocytes, activated plasma cells and lymphocytes of polyclonal origin.

Bone involvement, although rare, generally mimics of bone tumor and all the earlier reported cases have been diagnosed only after biopsy. Radiographic picture is inconsistent and may show lytic or even blastic lesion with rare periosteal reaction, which makes preoperative diagnosis a challenge.

Departments of Orthopedics, S. M. S. Hospital, Jaipur, [1]Pathology, SDM Hospital, Jaipur, Rajasthan, India

Address for correspondence: Dr. Rahul Sapra,
Department of Orthopedics, S. M. S. Hospital, Jaipur, Rajasthan, India.
E-mail: saprarahul@gmail.com

Access this article online	
Quick Response Code:	Website: www.ijoonline.com
	DOI: 10.4103/0019-5413.159682

Xanthogranulomatous osteomyelitis is rare and only 11 cases have been reported so far [Table 1].

Case Report

A 34-year-old male patient, farmer by occupation, attended the clinic with pain in his left ankle and the right foot since last 3 months. It was gradual in onset and increased at night. There was no history of trauma, fever or any significant past medical history. On examination, there was local rise of temperature with normal overlying skin and deep tenderness bilaterally. There was normal range of motion (ROM) at both the ankle and the subtalar joints with terminally painful movements. Laboratory investigations showed that total and differential leukocyte counts within normal range with raised erythrocyte sedimentation rate (38 mm at 1st h). Serum calcium, phosphorus, parathormone, liver and renal function tests were found to be within normal range.

Anteroposterior and lateral radiographs showed a well defined lytic lesion in the medial malleoli and the talus on the left side and on the cuboid bone on the right side [Figure 1]. Bilaterally, there was no cortical breach, margins were selerotic, periosteal reaction or intralesional calcification was absent. Computed tomography findings were consistent with the X-ray features showing lytic lesions without cortical breach in the respective bones. Magnetic resonance imaging findings showed increased signal intensity on both T1- and T2-weighted images without any soft tissue extension of the lesion. With these features a probable diagnosis of benign bone tumor was made. Patient was taken for Jamshedi needle biopsy from the lesion. Yellow cheesy material (caseation like) was aspirated and sent for culture, AFB staining, polymerase chain reaction (PCR) for tuberculosis and histopathological examination. In the meantime, because of

Table 1: Summary of this previous literature

Author	Number of cases	Site	Treatment	Other observations
Cozzutto[1] (1984)	2	Rib and tibial epiphysis	En bloc resection of 1st rib Excision and sequestrctomy of tibial lesion	-
Vankalakunti *et al.*[2] (2007)	1	Ulna	Curettage and bone grafting with internal fixation	Foamy histiocytes are derived from monocytes/macrophages and are positive for KP1, HAM56, CD11b and CD68
Cennimo *et al.*[3] (2009)	1	Index finger	Minocycline, clarithromycin, and ethambutol with radical synovectomy of the lesion	*Mycobacterium marinum* infection
Kashani *et al.*[7] (2010)	1	Femur	Wide spectrum antibiotic therapy	-
Borjian *et al.*[4] (2012)	1	Contralateral humerus and fibula	Patient left without treatment	Positive culture for *Staphylococcus aureus*
Kamat *et al.*[5] (2011)	1	Tibia	Simple curettage	Positive culture for *Staphylococcus aureus*
Lee *et al.*[6] (2013)	1	Ulna	Excision biopsy and curettage	Increased accumulation of FDG in PET scan
Wang *et al.*[8] (2013)	2	Ribs	-	Accumulation of FDG in PET/CT scan
Holmes *et al.*[9] (2013)	1	Tibia	Excision biopsy and curettage	-

FDG=Fluorodeoxyglucose, PET=Positron emission tomography, CT=Computed tomography

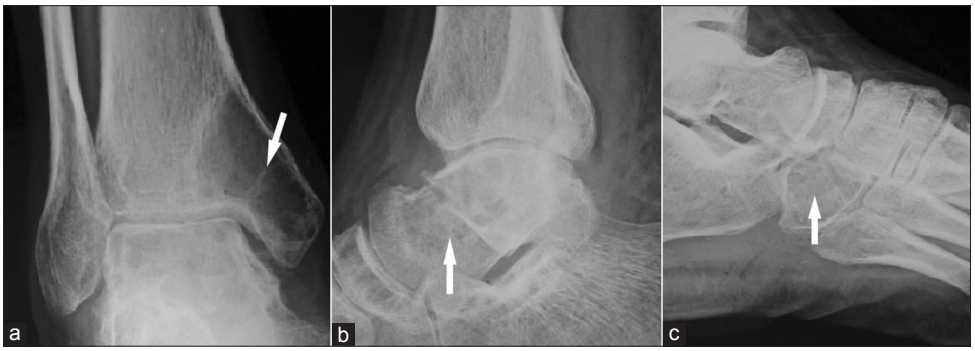

Figure 1: Anteroposterior (a) and lateral (b) radiographs of left ankle showing osteolytic lesion in medial malleoli and talus (arrow) (c) oblique view radiograph of right foot showing involvement of cuboid bone (arrow)

macroscopic findings of biopsy material and high prevalence of tuberculosis in our country, antitubercular treatment was started with isoniazid, rifampicin, pyrazinamide, and ethambutol emperically. However these drugs were stopped after 5 days because of a negative culture for other bacterias, AFB stain, PCR and histological picture consistent with xanthogranulomatous osteomyelitis. After a thorough review of literature at this stage, no definitive management protocol of the disease was found and the decision of curettage and bone grafting was taken. For left medial malleoli and right cuboid. Direct incision over the diseased areas were used. However, for left talus, anterolateral approach was used. Autogenic bone graft was taken from the right iliac crest. Curetted material was again sent for histopathological and microbiological assessment.

On gross examination, the specimen consisted of multiple yellowish white tissue pieces. Histopathological examination on both the occasions revealed dense infiltration of lymphocytes, large number of plasma cells and abundant foamy histiocytes in islands and sheets [Figure 2]. Again no microorganisms were grown on culture and PCR study for *Mycobacterium tuberculosis* was negative. After 1.5 years of followup, patient is relieved of his symptoms. His visual analog scale score has improved from a preoperative

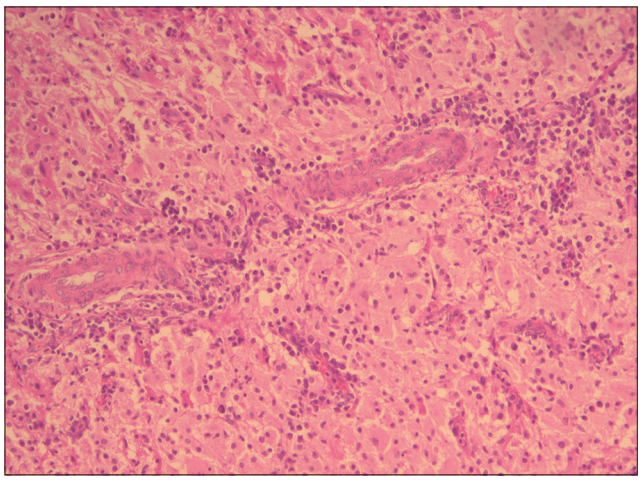

Figure 2: Histopatology of lesion showing characteristic foamy histiocytes along with abundant lymphocytes and plasma cells

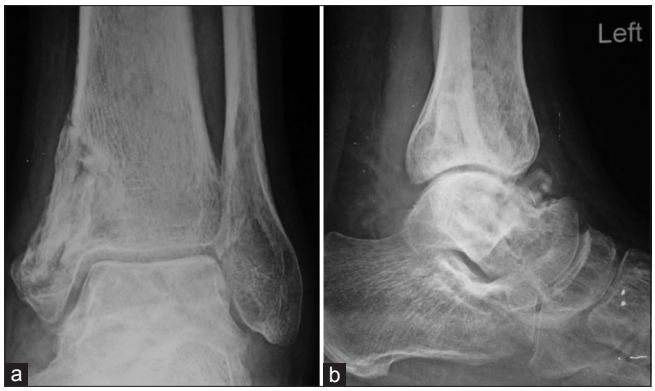

Figure 3: Followup radiographs after 1.5 years anteroposterior (a) and lateral views (b) showing reasonably good incorporation of graft tissue

value of 6 to a postoperative value of 2. There is no pre and postoperative loss of ROM. Followup X-rays show reasonably good incorporation of the bone graft without any signs of recurrence of the disease [Figure 3].

Discussion

Xanthogranulomatous osteomyelitis has variable age of presentation occurring in adolescents to post menopausal women, according to previously reported cases.[1-6] Except for the case described by Borjian *et al*. where they reported simultaneous lesions in the humerus and fibula, no other case report describes multifocal involvement. Furthermore, all of the previously mentioned cases have been reported in long bones namely femur, humerus, ulna, ribs, phalynx and tibia.[1-9] Involvement of small bones in the form of talus and cuboid makes this case unique. Borjian *et al*. and Kamat *et al*. isolated *Staphylococcus aureus,* while Cennimo reported *Mycobacterium marinum* from the lesion. However, no organism has been isolated in our case.

Differential diagnosis for xanthogranulomatous osteomyelitis includes Langerhan's cell histiocytosis, Erdhiem–Chester disease, lipid storage disorders such as gaucher's disease, chronic recurrent multifocal osteomyelitis, infiltrative storage disorder, malakoplakia, fibrohistiocytic tumor and metastatic renal cell carcinoma.[4] Being a farmer, fungal osteomyelitis was also considered as a possibility. However, presence of pathognomic cells in the form of foamy histiocytes on biopsy ruled out this possibility.

Differentiating between neoplastic process and inflammatory process remains the cornerstone in the management of this rare disorder. Unfortunately until date, only histopathological examination has shown promise in this regard. In the absence of any previously set treatment protocol for this disorder, the authors would like to recommend curettage with bone grafting as an effective method of treatment for this disease.

This rare entity should always be kept in mind as differential diagnosis by the treating surgeon and the radiologist.

References

1. Cozzutto C. Xanthogranulomatous osteomyelitis. Arch Pathol Lab Med 1984;108:973-6.
2. Vankalakunti M, Saikia UN, Mathew M, Kang M. Xanthogranulomatous osteomyelitis of ulna mimicking neoplasm. World J Surg Oncol 2007;5:46.
3. Cennimo DJ, Agag R, Fleegler E, Lardizabal A, Klein KM, Wenokor C, *et al. Mycobacterium marinum* Hand Infection in a "Sushi Chef". Eplasty 2009;9:e43.
4. Borjian A, Rezaei F, Eshaghi MA, Shemshaki H. Xanthogranulomatous osteomyelitis. J Orthop Traumatol 2012;13:217-20.
5. Kamat G, Gramapurohit V, Myageri A, Shettar C. Xanthogranulomatous osteomyelitis presenting as swelling in right tibia. Case Rep Pathol 2011;2011:257458.
6. Lee SH, Lee YH, Park H, Cho YJ, Song HT, Yang WI, *et al.* A case report of xanthogranulomatous osteomyelitis of the distal ulna mimicking a malignant neoplasm. Am J Case Rep 2013;14:304-7.
7. Kashani MM, Zakerian BZ, Shayan K, Riyasi F. Xanthogranulomatous osteomyelitis of femur. Iran J Orthop Surg 2010;8:187-90.
8. Wang YL, Yu H, Zhang H, Cui XJ, Han JK. Xanthogranulomatous osteomyelitis of rib mimicking malignant lesions in (18) F-FDG PET/CT imaging: A report of two cases. Clin Nucl Med 2014;39:e315-7.
9. Holmes BJ, Castelino-Prabhu S, Rosenthal DL, Ali SZ. Xanthogranuloma of bone: A challenging imitator of malignancy. Acta Cytol 2013;57:198-202.

How to cite this article: Sapra R, Jain P, Gupta S, Kumar R. Multifocal bilateral xanthogranulomatous osteomyelitis. Indian J Orthop 2015;49:482-4.

Source of Support: Nil, **Conflict of Interest:** None.