

Received: 2013.06.02  
Accepted: 2013.06.04  
Published: 2013.08.09

ISSN 1507-6164  
© Am J Case Rep, 2013; 14: 304-307  
DOI: 10.12659/AJCR.889422

## A case report of xanthogranulomatous osteomyelitis of the distal ulna mimicking a malignant neoplasm

Authors' Contribution:  
Study Design A  
Data Collection B  
Statistical Analysis C  
Data Interpretation D  
Manuscript Preparation E  
Literature Search F  
Funds Collection G

ABEF 1 **Seung Hyun Lee**  
ABEF 1 **Young Han Lee**  
D 2 **Heejung Park**  
C 3 **Yong Jin Cho**  
F 1 **Ho-Taek Song**  
A 2 **Woo Ik Yang**  
ACF 1 **Jin-Suck Suh**

1 Department of Radiology, Research Institute of Radiological Science, Medical Convergence Research Institute, Severance Biomedical Science Institute, Yonsei University, College of Medicine, Seoul, Republic of Korea  
2 Department of Pathology, Severance Hospital, Yonsei University, College of Medicine, Seoul, Republic of Korea  
3 Department of Orthopedic Surgery, Severance Hospital, Yonsei University, College of Medicine, Seoul, Republic of Korea

**Source of support:** Basic Science Research Program through the National Research Foundation of Korea(NRF) funded by the Ministry of Education, Science and Technology (2012R1A1A2042165)

**Corresponding Author:** Young Han Lee, e-mail: [radiologie@gmail.com](mailto:radiologie@gmail.com)

**Patient:** Male, 59  
**Final Diagnosis:** Xanthogranulomatous osteomyelitis  
**Symptoms:** Painful swelling in the wrist  
**Medication:** Drug history of antifungal agents  
**Clinical Procedure:** Excisional biopsy  
**Specialty:** Radiology

**Objective:** Rare disease  
**Background:** Xanthogranulomatous inflammation is a chronic inflammatory disease in which bone involvement is extremely rare. Bone involvement of xanthogranulomatous inflammation, termed xanthogranulomatous osteomyelitis (XO), often presents as a mass-like lesion extending to adjacent structures, which can mimic infiltrative carcinoma.

**Case Report:** We present a case of XO in the ulna, which mimicked a neoplasm. A 59-year-old man presented with a 2-month history of painful swelling in the right wrist. Plain radiography and CT showed an osteolytic lesion at the distal ulna. MRI revealed a soft-tissue mass with lobulated margins and contrast enhancement. Intense uptake in bone scan and PET suggested malignancy. An excisional biopsy from the representative area resulted in a pathology diagnosis of XO.

**Conclusions:** Gross and radiologic manifestations of XO can mimic neoplasm. XO generally has benign prognosis, contrary to malignant bone tumor. Therefore, biopsy and histopathological confirmation are necessary for proper management.

**Key words:** xanthogranulomatous osteomyelitis • inflammatory • ulna • image

Full-text PDF: <http://www.amjcaserep.com/download/index/idArt/889422>



1432



1



1



13

## Background

Xanthogranulomatous inflammation is a chronic inflammatory disease histologically characterized by collection of foamy macrophages admixed with polymorphonuclear leukocytes, activated plasma cells, and lymphocytes of polyclonal origin, in a mosaic-like pattern [1,2]. The most common sites of involvement are the kidney and gallbladder [3,4]; however, it can occur in many other organs, including the lymph nodes, colon, appendix, pancreas, urinary bladder, prostate, testis, epididymis, endometrium, ovaries, fallopian tubes, vagina, and salivary glands. Xanthogranulomatous inflammation rarely occurs in lung, brain, or bone [1,5].

Bone involvement of xanthogranulomatous inflammation, termed xanthogranulomatous osteomyelitis (XO), often presents as a mass-like lesion extending to adjacent structures, and can mimic infiltrative carcinoma [6]. XO may also accompany inflammatory signs of pain, fever, and leukocytosis, due to a delayed-type hypersensitivity reaction of cell-mediated immunity. Therefore, distinguishing infection from neoplasm is an important first step in diagnosing XO. The differential diagnosis of xanthogranulomatous osteomyelitis includes Langerhans cell histiocytoses, Erdheim-Chester disease, and chronic recurrent multifocal osteomyelitis [2,5,7–9]. Clinical manifestation and radiologic findings are important in the decision, as well as histopathological diagnosis, which can confirm the diagnosis of XO.

XO is rare and only 6 cases have been reported: 2 in 1984 by Cozzutto et al. [1], 1 in 2007 by Vankalakunti et al. [5], 1 in 2009 by Cennimo et al. [10], 1 in 2011 by Borjian et al. [2], and by 1 Kamat et al. [11] in 2011. To the best of our knowledge, this is the seventh reported case of xanthogranulomatous inflammation in bone. We report this rare XO case with the radiologic features of simple radiography, PET/CT, and MRI.

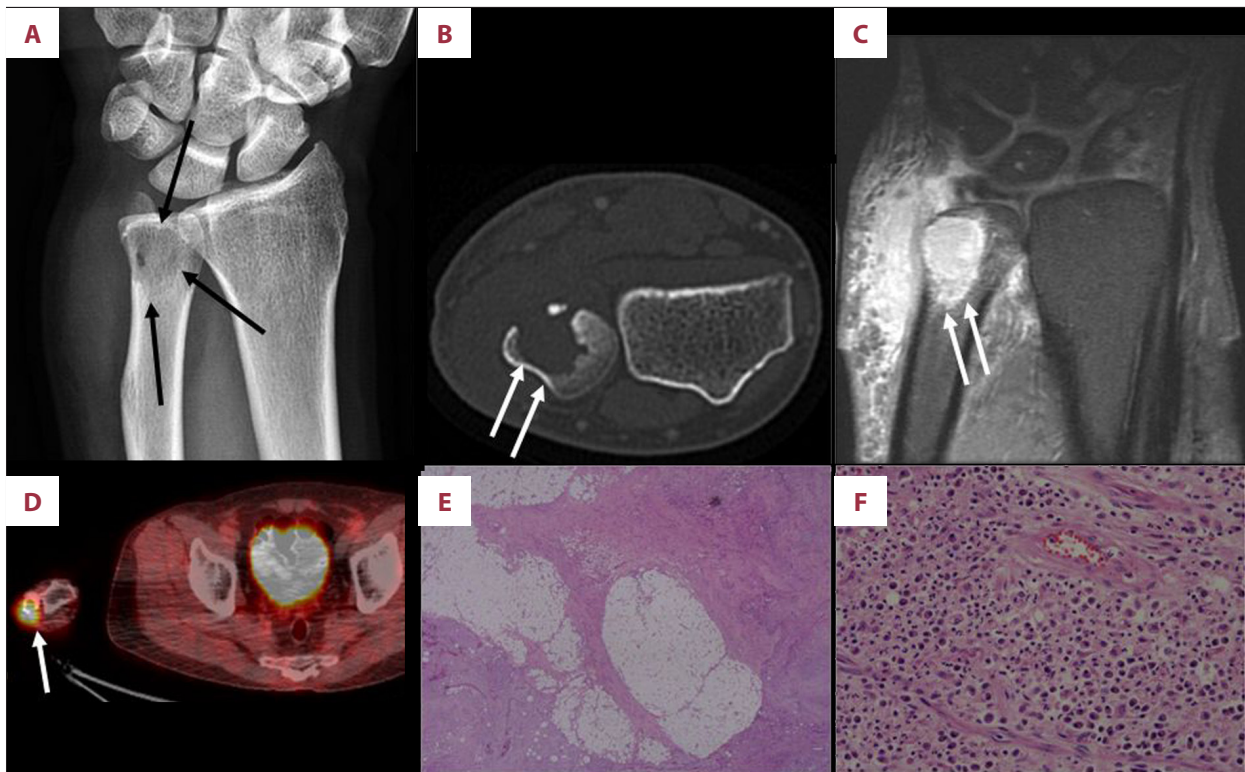
## Case Report

A 59-year-old man was referred to the orthopedics department of our institution, presenting a 2-month history of right wrist pain with swelling. He had no trauma history or previous surgical history of the right wrist. He had a past medical history of myelodysplastic syndrome (MDS) and rheumatoid arthritis, diagnosed 6 years before. He had been taking antifungal medicine (itraconazole) for 2 months and vitamin A and D for 3 months for multiple furunculosis of the face, neck, and axilla. Upon systematic review, the patient's right wrist had mild limitation of motion and swelling. Physical examination of his right wrist revealed tenderness with local heat and without motor or sensory impairment. Lab studies showed leukocytopenia (white blood cell count of 2120 cells/mL with a normal distribution of neutrophils [42.9%], lymphocytes [43.4%], and

eosinophils [0%], and slightly elevated monocytes [13.7%]) and mild thrombocytopenia (platelet count of 10 800 cells/mL) that had been consistently low due to his history of MDS. Other lab studies, including liver function tests and renal function tests, were within normal limits, and his vital signs were stable, without fever. The clinical impression according to his symptoms, signs, and laboratory data could suggest metastasis or myeloma involvement, considering his age and prior history of MDS.

Plain radiography of the wrist revealed a bulging mass-like lesion located at the distal ulna with a round radiolucent lesion of the distal ulna, suggesting an osteolytic mass (Figure 1A). CT of the wrist showed a lobulated soft-tissue-density mass about 2×2×4 cm involving the ventral-medial side of the distal ulna, with a focal cortical defect of the bone (Figure 1B). On MRI, the lobulated mass on the distal ulna displayed low signal intensity on T1-weighted image, and high signal intensity on T2-weighted image, and was strongly enhanced by gadolinium injection (Figure 1C). The lobulated contour, with the extending pattern of the lesion into the adjacent soft tissues and location of the lesion itself, suggested infection, but the adjacent bone marrow edema was relatively minimal. Therefore, a tumorous lesion, such as primary bone tumor, metastasis/myeloma, or lymphoproliferative disease (Castleman's disease), seemed more probable, and infection seemed less likely. Whole body bone scan (WBBS) and PET-CT scan were performed to detect any other lesions. The WBBS revealed increased uptake in the right distal ulna, a finding suspicious for a bony tumor. The PET-CT also showed focal intense fludeoxyglucose (FDG) uptake in the distal part of right ulna, compatible with malignancy, with no other abnormal FDG uptakes to indicate metastasis (Figure 1D).

The radiological diagnosis of the wrist lesion focused more on a malignant lesion than infection, but, controversial as this diagnosis is, confirmation by pathology was recommended. A surgical incisional biopsy of the distal part of right ulna was performed, and microscopic findings showed acute and chronic inflammation, with extensive infiltration of foamy macrophages, most likely consistent with xanthogranulomatous osteomyelitis. The excisional biopsy was used to completely rule out malignancy, since radiologic findings suggested neoplasm rather than infection. In the operating room, the exposed distal ulna had a mass lesion that manifested malignant behavior aggressively invading upon enclosing tissue. The lesion was completely excised and curetted out, and the soft tissue and bony tissue from the ulna were sent to the pathology department for histopathologic inspection. Upon gross examination, the tissue presented as an ill-defined mass-like lesion. The specimen had diffuse infiltration of mixed inflammatory cells, including neutrophils, small lymphocytes, and plasma cells admixed with aggregated foamy macrophages on light microscopy. The foamy macrophages had a distinct cytoplasmic border, abundant pale eosinophilic granular cytoplasm, and a small round nucleus (Figure 1E and 1F).



**Figure 1.** (A) Radiograph of an antero-posterior view of the right wrist displays an osteolytic lesion and soft tissue with bulging contour and cortical breakdown in the distal ulna. (B) CT scans of the right wrist in bone window setting presenting a 2×2×4 cm sized soft tissue density bulging mass and cortical breakdown on the ventral-medial side of the distal ulna. This mass showed no periosteal reaction and no sclerotic rim. (C) Coronal contrast enhanced T1-weighted MRI after gadolinium administration showed a soft tissue bulging mass on the ventral-medial side of the distal ulna. (D) PET-CT fusion axial images show focal intense FDG uptake in right distal ulna suggesting malignancy. No other abnormal FDG uptake was found to suggest distant metastasis. (E) Light microscopy with H&E staining (×12.5) showed a mass-like lesion with ill-defined border (F) H&E staining (×200) revealed diffuse infiltration of mixed inflammatory cells, including neutrophils, small lymphocytes, and plasma cells admixed with aggregated foamy macrophages. The foamy macrophages had a distinct cytoplasmic border, abundant pale eosinophilic granular cytoplasm, and a small round nucleus.

The final pathologic diagnosis was confirmed as xanthogranulomatous osteomyelitis based on the acute and chronic inflammation, with extensive infiltration of foamy macrophages and formation of granulation tissue.

## Discussion

Xanthogranulomatous osteomyelitis (XO) is a notable disease in terms of its rarity. Only 6 cases of XO have been reported in the literature (Table 1). Cozzutto et al.<sup>1</sup> first reported 2 cases of involvement of the first rib in a 5-year-old boy and the proximal metaphysis of the tibia in a 14-year-old boy in 1984. In 2007, Vankalakunti et al. [5] reported a case of 50-year-old woman presenting XO in diaphysis of ulna. Cennimo et al. [10] reported a case that manifested as a xanthogranulomatous reaction with culture results positive for *Mycobacterium marinum* in the index finger and wrist of a 41-year-old sushi chef in 2009. Recently, a case of XO invading 2 separate bones, the metaphysis of the

right humerus and the medulla, metaphysis, and diaphysis of the left fibula, in a 14-year-old boy was published by Borjian et al. [2]. Another case of XO, which presented as swelling in the right distal tibia, in a 13-year-old boy was reported by Kamat et al. [11]. Here, we report the seventh case of xanthogranulomatous inflammation in bone, a case of a XO in the distal ulna.

We report this case due to its imaging findings on simple radiograph, CT, MRI, WBBS, and PET-CT, and the importance of differential diagnosis with malignancy, therefore we could present various radiologic features of XO. XO is generally benign and curable, but it can mimic malignant bone tumor clinically and radiologically. Our case of XO is remarkable in that the CT and MR images showed an enhancing mass in the ulna, with focal uptake in WBBS and PET-CT, which should be distinguished from malignant tumors. At present, imaging XO does not suggest a specific finding to make a convincing diagnosis. Therefore, histopathological examination seems to be necessary to accurately diagnose XO.

**Table 1.** Summary of the previous literatures.

Case reports	Region	Culture results
Cozzutto C. [1]	Case 1 (1 <sup>st</sup> rib)	No organisms
	Case 2 (proximal metaphysis of tibia)	No organisms
Vankalakunti M., Saikia U.N., Mathew M. et al. [5]	Diaphysis of ulna	No organisms
Cennimo D.J., Agag R., Fleegler E. et al. [10]	Index finger and wrist	<i>Mycobacterium marinum</i>
Borjian A., Rezaei F., Eshaghi M.A. et al. [2]	Metaphysis of the humerus and the medulla, metaphysis, and diaphysis of the fibula	<i>Staphylococcus aureus</i>
Kamat G. et al. [11]	Distal tibia	<i>Staphylococcus aureus</i>

Microscopic differential diagnosis for xanthogranulomatous osteomyelitis includes Langerhans cell histiocytosis (LCH) and Erdheim-Chester disease (ECD) [2,5,7–9]. Unifocal LCH may present as a single osteolytic lesion, generally affecting long or flat bones [2]. However, bone involvement usually occurs in pediatric patients, with a median age of 5, and Birbeck granules, a diagnostic hallmark [12,13], were missing in our case, therefore LCH could be ruled out. ECD is a systemic condition characterized by multisystemic xanthogranulomatous infiltration, including bone involvement affecting middle-aged adults [2]. Our patient did not show any other site of inflammation on WBBS and PET; therefore, ECD was ruled out. Accordingly, histopathology should distinguish pseudotumoral XO from bone tumors or other benign conditions.

## Conclusions

Xanthogranulomatous osteomyelitis is extremely rare and its preoperative diagnosis is a challenge because it can

radiographically mimic a malignant tumor. XO generally has a benign prognosis, contrary to malignant bone tumor; therefore, biopsy and histopathological confirmation are necessary for proper management.

## Teaching point

Xanthogranulomatous osteomyelitis (XO) is extremely rare, and its preoperative diagnosis is a challenge because it can mimic a malignant tumor on radiologic examinations, including simple radiograph, CT, MR, and PET. Gross and radiologic manifestations of XO can mimic neoplasm; therefore, XO needs differential diagnosis from malignancy and histopathological confirmation is necessary for proper management. Although image features can mimic the malignancy, the radiologist must be aware of the possibility of the XO.

## Conflicts of interest and source of funding

There is no conflict of interest.

## References:

- Cozzutto C: Xanthogranulomatous osteomyelitis. Arch Pathol Lab Med, 1984; 108: 973–76
- Borjian A, Rezaei F, Eshaghi MA, Shemshaki H: Xanthogranulomatous osteomyelitis. J Orthop Traumatol, 2012; 13(4): 217–20
- Franco V, Aragona F, Genova G et al: Xanthogranulomatous cholecystitis. Histopathological study and classification. Pathol Res Pract, 1990; 186: 383–90
- Antonakopoulos GN, Chapple CR, Newman J et al: Xanthogranulomatous pyelonephritis. A reappraisal and immunohistochemical study. Arch Pathol Lab Med, 1988; 112: 275–81
- Vankalakunti M, Saikia UN, Mathew M, Kang M: Xanthogranulomatous osteomyelitis of ulna mimicking neoplasm. World J Surg Oncol, 2007; 5: 46
- Unni KK, McLeod RA, Dahlin DC: Conditions that simulate primary neoplasms of bone. Pathol Annu, 1980; 15: 91–131
- Hamada T, Ito H, Araki Y et al: Benign fibrous histiocytoma of the femur: review of three cases. Skeletal Radiol, 1996; 25: 25–29
- Girschick HJ, Huppertz HI, Harmsen D et al: Chronic recurrent multifocal osteomyelitis in children: diagnostic value of histopathology and microbial testing. Hum Pathol, 1999; 30: 59–65
- Kayser R, Mahfeld K, Grasshoff H: [Vertebral Langerhans-cell histiocytosis in childhood – a differential diagnosis of spinal osteomyelitis]. Klin Padiatr, 1999; 211: 399–402
- Cennimo DJ, Agag R, Fleegler E et al: Mycobacterium marinum Hand Infection in a “Sushi Chef”. Eplasty, 2009; 9: e43
- Kamat G, Gramapurohit V, Myageri A, Shettar C: Xanthogranulomatous osteomyelitis presenting as swelling in right tibia. Case Report Pathol, 2011; 257458
- Favara BE, Feller AC, Pauli M et al: Contemporary classification of histiocytic disorders. The WHO Committee On Histiocytic/Reticulum Cell Proliferations. Reclassification Working Group of the Histiocyte Society. Med Pediatr Oncol, 1997; 29: 157–66
- Weitzman S, Egeler RM: Langerhans cell histiocytosis: update for the pediatrician. Current Opin Pediatr, 2008; 20: 23–29