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Case Report

Sarcoid dactylitis: A case report of rare presentation of uncommon disease, sarcoidosis—From Ethiopia ☆,☆☆

Zerihun Gelashe Hailu[a,*], *Bemnet Taye Gebregiorgis*[a], *Moges Zenebe Wegayehu*[a], *Becky Abdisa Adugna*[b], *Abdulhamid Mustefa Bedewi*[c]

[a] *Department of Radiology, Black Lion Hospital, Addis Ababa University, Addis Ababa, Ethiopia*
[b] *Department of Internal Medicine, Black Lion Hospital, Addis Ababa University, Addis Ababa, Ethiopia*
[c] *Black Lion Hospital, Addis Ababa University, Addis Ababa, Ethiopia*

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ABSTRACT

We present a rare case of sarcoid dactylitis after an adult female presented with painless swelling of the bilateral hand fingers and toe, with radiographic findings highly characteristic for osseous sarcoidosis and confirmation made by bronchoscopic biopsy of hilar enlarged lymph nodes.

Introduction

Sarcoidosis is a multisystemic disease of young adults due to accumulation of noncaseating granulomas. The most common sites of involvement are the lungs and the mediastinal hilar lymph nodes. Patients present with pulmonary manifestations in 87%-97% of the cases [1]. Skin (15%-18%), eyes (10%-30%), liver (20%-30%), spleen (10%), heart (2%-5%), nervous system (5%), upper respiratory tract (2%), and gastrointestinal tract (approximately 1%) are among the other common sites of involvement [1].

Osseous sarcoidosis is a very uncommon manifestation of the illness (seen in 5% of patients) and most patients with bone involvement have no symptoms. The majority of cases of osseous sarcoidosis are thus discovered by incidentally [1,2]. It is mostly seen in young individuals with multiorgan sarcoidosis, and it is rarely a presenting symptom by itself [2].

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* Corresponding author.
E-mail address: zerihun.gelashe@aau.edu.et (Z.G. Hailu).
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Case presentation

This is a 32-year female patient presented with painless digit swelling of 2-year duration. The swelling involved right middle finger, left middle finger, and right second toe. She has no other systemic symptom. On examination, the involved digits were swollen with sausage appearance. Hand and foot radiograph (Fig. 1) revealed bilateral symmetric third middle phalanx and right foot second proximal phalanx lytic lesions with thickened trabeculae forming typical lace (honeycomb) like lesions with surrounding soft tissue swelling. There is also a geode like lytic lesion in the right middle phalanx with bilateral radial side cortical scalloping. Based on typical imaging appearance finding suggestive of sarcoid dactylitis, chest radiograph was requested and revealed right lower paratracheal and bilateral hilar lymphadenopathy consistent with sarcoid diagnosis. Subsequently bronchoscopic biopsy showed granulomatous inflammation and angiotensin converting enzyme level was found to be elevated to 167 μg/L (normal $<$ 8-52). After settling the diagnosis, the patient is started on prednisolone and she is on follow-up.

Discussion

Musculoskeletal system findings are among the important features of extrapulmonary sarcoidosis involvement and may occur prior to or simultaneously with the pulmonary involvement. Sarcoid musculoskeletal involvement include are myopathies, tendinopathies, synovitis/arthritis, and dactylitis [3].

Among osseous manifestations of sarcoidosis dactylitis is the most common form of presentation appearing in approximately 90 % of patient with osseous sarcoidosis [1,2]. Osteitis tuberculosa multiplex cystica was the word used by Jüngling, a student of Perthes, to describe lesions in the small bones of the hands and feet by 1920 and 1926 and sarcoid dactylitis is also known as Jüngling disease for this reason [2]

Sarcoid dactylitisis typically seen among young adult women between the ages of 30 and 50 years. They commonly co-occur with chest disease and chronic skin lesions in individuals with active multi-systemic sarcoidosis [4]. It typically involves bilateral fingers and toes asymmetrically causing soft tissue swelling of the middle and distal phalanges of second and third digits forming “sausage finger” due to perivascular infiltration by granulomatous lesions [2]. Extension of this granulomatous lesion in the soft tissue to the adjacent bone cause bone destruction and produce characteristic radiographic changes of sarcoid dactylitis [4].Our patient presents with this typical bilateral asymmetric involvement of the fingers with the predominant involvement of the middle phalanges but also the distal part of the proximal phalanges of the right second toe is affected. Such kind of initial symptom in extrapulmonary site is an unusual presentation in sarcoid patients.

High erythrocyte sedimentation rate (ESR), increased acetylcholinesterase levels (ACE), and energy to tuberculin are the laboratory findings [2]. ACE is a recognized marker for sarcoidosis and is produced by epithelioid cells originating from active macrophages. Despite ACE increase in multiple other disease entities, it aids in sarcoidosis diagnosis, used to tell disease activity, and is helpful for monitoring therapy [5]. This patient also showed increased ACE level suggesting an active sarcoidosis.

Ultrasound, radiograph, scintigraphy, CT and MRI can be used in sarcoid imaging. High-resolution ultrasound is useful to distinguish soft tissue infiltration, which will appear hypoechoic, from a joint effusion and also useful to guide a diagnostic puncture [4]. Bone scintigraphy precede radiographic abnormalities making it a more sensitive modality for lesion detection and with an additional benefit of being a whole-body investigation for detecting multiple musculoskeletal sites [4].

Three types of sarcoidosis bone lesions of the small hand bones are described based on their radiographic appearance. Type I (bullous lesion), the rarest with appearances of large bone geodes. Type II (small pseudocystic lesions), which are the most common and appears multiple well delineated, rounded and occasionally confluent polycyclic geodes. Type III: “lace-like” or “bees nest” or “grid” reticulated appearance with thickened bone sheets and thin cortex. These types of disease may co-exist in the same bone or in several bones in the same person. Typically, no periosteal reaction or joint involvement is seen on standard radiography. CT is particularly useful for detail evaluation of bone changes seen on radiograph [4]. Pathologic fractures with bone collapse and misalignment may occur because of sarcoid osteolysis [6]. Our patient radiograph showed a combination of radiographic abnormalities mentioned above. Other than the adjacent tissue swelling there is no periosteal reaction seen in the radiograph of our patient.

When it comes to detecting osseous lesions and determining the degree of marrow, soft tissue, and tendon involvement, MR imaging is the most sensitive, but it is not very specific. Osseous sarcoid can show up on imaging as well-defined, focal T1-hypointense, T2-hyperintense, and enhancing intramedullary lesions or as poorly defined infiltrative processes in the bone marrow [6].

In such patient with sole or predominant hand symptoms of dactylitis and lytic bone lesions on radiograph, psoriatic arthritis, gout, enchondroma, acrometastases, and septic osteitis, are the key differential diagnoses to be taken into consideration [2].

There is no agreement on how to treat osseous sarcoidosis. Patients who are asymptomatic do not require treatment. Corticosteroids can be used as first-line therapy for symptomatic individuals, albeit their usefulness on radiologic alterations has not been proven. If there is no response or the lesions progress, immunosuppressive medication (methotrexate, TNF inhibitor) is the second line choice [2].

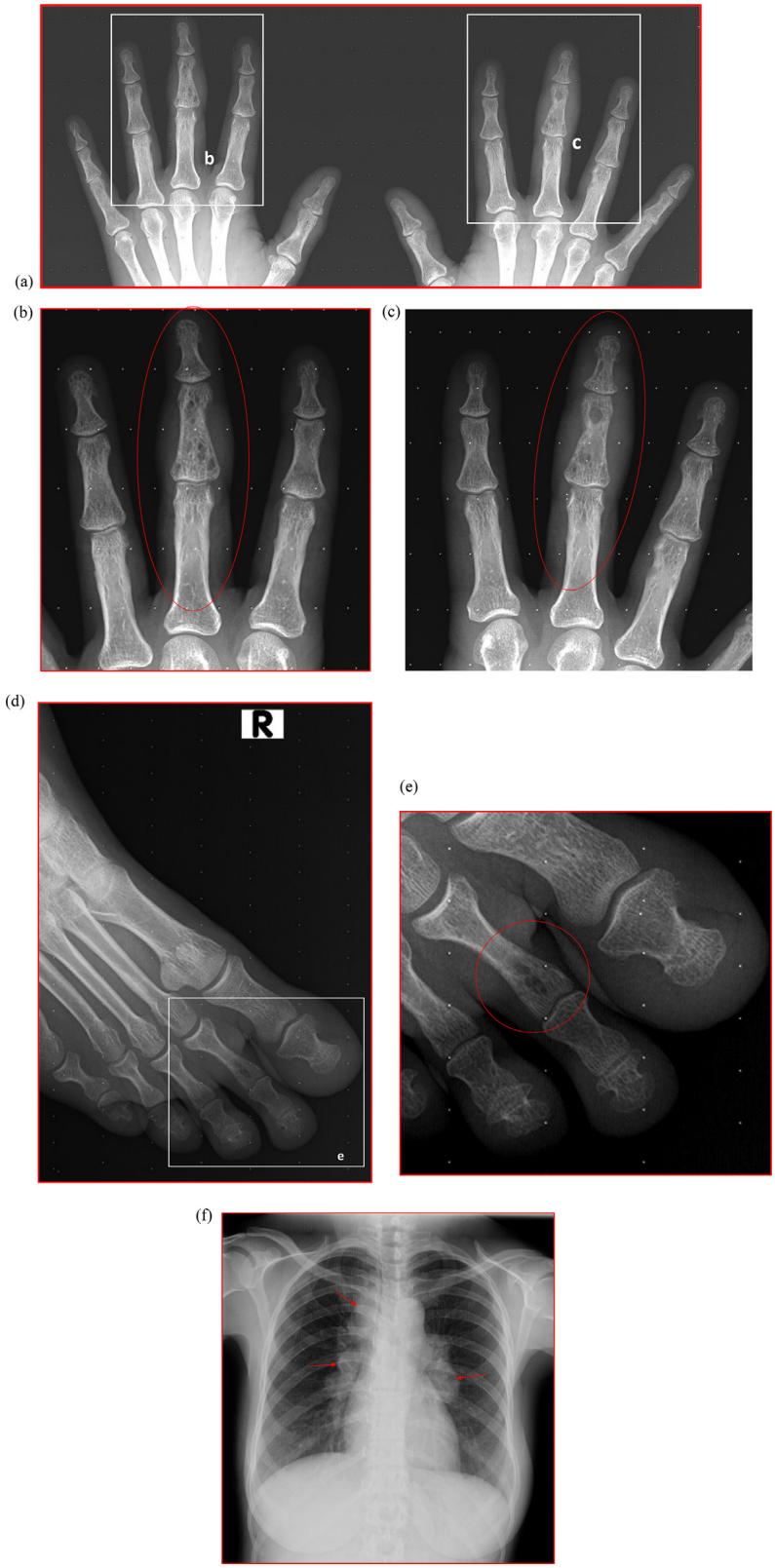

Fig. 1 – Bilateral hand, right foot, and chest radiograph—lace-like lytic lesion seen in the middle phalanx of bilateral middle finger and distal part of middle phalanx of right second toe. (Note the oval circle with red color outline in b, c and e). The middle finger of bilateral hands has also soft tissue swelling suggesting dactylitis (compare middle finger with un-affected index and ring finger for better appreciation of soft tissue swelling). Right paratracheal and bilateral hilar enlarged lymphadenopathies (red arrows) seen forming typical "123 Sign" used to describe sarcoid mediastinal lymphadenopathy.

Conclusions

The rheumatologic symptoms and imaging appearance of the sarcoidosis are not as well understood and well known as the imaging findings of thoracic presentations of sarcoidosis. The bony lesions of sarcoid dactylitis are classical, very characteristic and well known, especially useful to avoid biopsy in a known sarcoid patient presenting with dactylitis. Such characteristic lesion is also quite useful in directing further investigation in patient who present initially with dactylitis to arrive at the definitive diagnosis of sarcoidosis, similar to the case presented here.

Patient consent

Written informed consent was taken from the patient and confidentially is ascertained.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2023.09.065.

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