A rare case of osseous sarcoidosis presenting as lytic lesions 20 years after initial diagnosis

John Shumar , Tyler Church, Arthur Holtzclaw, Joseph Zeman

Pulmonary and Critical Care Medicine, Walter Reed National Military Medical Center, Bethesda, Maryland, USA

Correspondence to

Dr John Shumar;
John.n.shumar.mil@mail.mil

Accepted 5 December 2020

DESCRIPTION

A 70-year-old African-American woman with a history of biopsy-proven sarcoidosis with pulmonary and cutaneous manifestations diagnosed 20 years ago presented to her primary care physician (PCP) for an evaluation after two recent falls at home. Given the acute nature of these changes, her PCP ordered a CT scan of the head to evaluate for any intracranial abnormalities, which revealed numerous lytic lesions within the calvarium (figure 1). With these acute findings, a prior CT scan of the chest from 2 weeks prior was reviewed retrospectively and was also significant for multifocal lytic lesions in the cervical and thoracic spine. All previous imaging studies prior to these CT scans were reviewed without any evidence of osseous changes.

Given these new lytic lesions, there was significant concern for malignancy, specifically multiple myeloma or metastatic disease. Serum protein electrophoresis, urine protein electrophoresis, serum-free light chains and a peripheral smear were ordered and the result was negative. She was up to date with age-appropriate cancer screenings with no significant findings. Ultimately, she underwent a positron emission tomography (PET) scan, which revealed innumerable hypermetabolic mixed sclerotic and lytic lesions in both the axial and the appendicular skeleton concerning for osseous sarcoidosis given her otherwise negative malignancy workup (figure 2). A right iliac crest biopsy was performed, which was significant for non-caseating granuloma formation consistent with osseous sarcoidosis. She was treated with prednisone and transitioned to methotrexate with repeat PET imaging showing a decrease in the activity and number of the

Figure 2 Positron emission tomography CT at the time of diagnosis showing innumerable hypermetabolic osseous mixed sclerotic and lytic lesions. These findings were concerning for malignancy versus osseous sarcoidosis given her history of biopsy-proven pulmonary sarcoidosis.

hypermetabolic osseous lesions, consistent with a good treatment response (figure 3).

Sarcoidosis is a systemic inflammatory disease defined by the development of non-caseating granulomas affecting any organ system.¹ Osseous involvement of sarcoidosis is relatively rare and infrequently described in the literature. In previous analyses, there is an estimated prevalence of osseous sarcoidosis among all patients with sarcoidosis of around 3%–13%, with the majority of cases having axial skeleton involvement.² Only around 50% of patients with osseous involvement will be symptomatic, and of those patients, most present with dactylitis or low back pain.² Osseous involvement is often identified at the time of initial diagnosis of sarcoidosis, but can also be diagnosed incidentally on imaging many years later.² Osseous sarcoidosis

Figure 1 Non-contrast CT scan of the head showing numerous lytic lesions within the calvarium, initially raising concern for multiple myeloma versus metastatic disease.

Figure 3 Positron emission tomography CT obtained after treatment showing an interval decrease in the activity and number of previously seen hypermetabolic lytic lesions within the axial and appendicular skeleton, consistent with a good response to treatment.

© BMJ Publishing Group Limited 2020. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: Shumar J, Church T, Holtzclaw A, et al. *BMJ Case Rep* 2020;**13**:e239319. doi:10.1136/bcr-2020-239319

Images in...

on imaging can be broadly described as permeative, destructive or lytic. The PET scan has further revolutionised the imaging evaluation of osseous sarcoidosis by adding metabolic activity information and by allowing for a more sensitive evaluation of the entire axial and appendicular skeleton in one scan.³ This is important given how many patients with osseous sarcoidosis remain asymptomatic.² Ultimately, given the radiographic similarity to malignancy, a tissue biopsy is often needed for confirmation and delineation of the underlying pathology.⁴ There is no consensus for the treatment of osseous sarcoidosis, although glucocorticoids are often used initially with transition to methotrexate or hydroxychloroquine.² Given our patient's history of disease, her osseous involvement nearly 20 years after initial

diagnosis represents an exceedingly rare presentation of a rare disease. This case is a pertinent reminder of the pervasive nature of sarcoidosis and its potential to affect any organ system at any time, particularly as treatment can alter the clinical course.

The views expressed are those of the authors and do not reflect the official policy or position of the US Army, Air Force, Navy, Department of Defense or the US Government.

Contributors JS wrote the manuscript and is the corresponding author and article guarantor. TC helped to edit the manuscript. AH evaluated the patient initially, and edited and approved the manuscript. JZ approved the manuscript.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

ORCID iD

John Shumar <http://orcid.org/0000-0001-8913-6139>

REFERENCES

- 1 Iannuzzi MC, Rybicki BA, Teirstein AS. Sarcoidosis. *N Engl J Med* 2007;357:2153–65.
- 2 Ben Hassine I, Rein C, Comarmond C, et al. Osseous sarcoidosis: a multicenter retrospective case-control study of 48 patients. *Joint Bone Spine* 2019;86:789–93.
- 3 Demaria L, Borie R, Benali K, et al. ¹⁸F-FDG PET/CT in bone sarcoidosis: an observational study. *Clin Rheumatol* 2020;39:2727–34.
- 4 Sparks JA, McSparron JJ, Shah N, et al. Osseous sarcoidosis: clinical characteristics, treatment, and outcomes—experience from a large, academic hospital. *Semin Arthritis Rheum* 2014;44:371–9.

Learning points

- New lytic osseous lesions should primarily raise concern for a malignant process, such as multiple myeloma or metastatic spread, but also include benign entities such as sarcoidosis. Differentiating between them often requires a biopsy and tissue specimens.
- Sarcoidosis can affect nearly any organ in the body, manifesting at the beginning or late in the disease course. This must be considered in any new clinical change in a patient with a history of sarcoidosis, and should always be considered a disease of exclusion.

Copyright 2020 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <https://www.bmj.com/company/products-services/rights-and-licensing/permissions/>
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

Customer Service

If you have any further queries about your subscription, please contact our customer services team on +44 (0) 207111 1105 or via email at support@bmj.com.

Visit casereports.bmj.com for more articles like this and to become a Fellow