Case Report

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DOI: 10.4103/ijh.ijh_62_23

# Sarcoidosis presenting with bicytopenia due to bone marrow granuloma

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**Abstract:**

A 55-year-old homemaker presented with low-grade fever, fatigue, and loss of weight for 2 months. She had hepatosplenomegaly, mediastinal lymphadenopathy, cytopenias, hypercalcemia with normal parathormone, and noncaseating granulomas in bone marrow and lymph node biopsy. The patient was diagnosed to have sarcoidosis with bone marrow involvement (histopathology showing noncaseating granuloma and high serum angiotensin-converting enzyme after ruling out other causes of granulomatous disorders by appropriate tests). She was managed with prednisolone 20 mg daily which was reduced and stopped after 3 months. On follow-up, she was asymptomatic and her blood counts were normal. Diagnosis of sarcoidosis should be considered always after ruling out other causes since it can mimic lymphoproliferative disorders and granulomatous infections. The initial presentation of sarcoidosis with bicytopenia due to bone marrow granulomas is extremely rare and physicians should have awareness of such atypical presentations.

**Keywords:**

Bicytopenia, bone marrow granuloma, sarcoidosis

## Introduction

Sarcoidosis is an inflammatory multisystem disorder of unclear etiology characterized by noncaseating epithelioid cell granulomas. Pulmonary involvement and mediastinal lymphadenopathy are the common features associated with sarcoidosis. The extrapulmonary manifestations can cause involvement of the skin, eyes, and heart. Bone marrow granulomas secondary to sarcoidosis is rare.

## Case Report

A 55-year-old homemaker presented with low-grade fever, fatigue, and loss of weight (8-kg) for 2 months. She had type 2 diabetes mellitus (on insulin) for 10 years. She had no sick contacts and had no history of addictions. On examination, she had pallor, hepatomegaly (6 cm below the costal margin, firm in consistency, smooth surface, and nontender), and splenomegaly (3 cm below the costal margin and firm in consistency). The rest of the examination was unremarkable.

Hemoglobin was 8.4 g/dL (normocytic normochromic anemia), total leukocyte count 2910/μl (50% neutrophils, 39% lymphocytes, 4% eosinophils, and 7% monocytes), platelet count 195,000/μl, and erythrocyte sedimentation rate 68 mm in 1st h. Peripheral smear showed normocytic normochromic anemia with mild neutropenia, but there were no abnormal cells or blasts. Urinalysis was normal. Serum calcium level was 12.6 mg/dL, serum albumin 3.6 g/dL, and serum phosphorus was 4.2 mg/dL. Other electrolytes were normal. An intact parathormone level was 35.9 pg/mL (reference range, 15–65 pg/mL). The 1, 25 dihydroxyvitamin D3 level was 86 (19.9–79.5). The rest of the biochemical parameters were normal.

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**Submission:** 05-08-2023
**Revised:** 26-08-2023
**Accepted:** 27-08-2023
**Published:** 16-10-2023



**How to cite this article:** Abdulla MC. Sarcoidosis presenting with bicytopenia due to bone marrow granuloma. Iraqi J Hematol 2023;12:210-2.

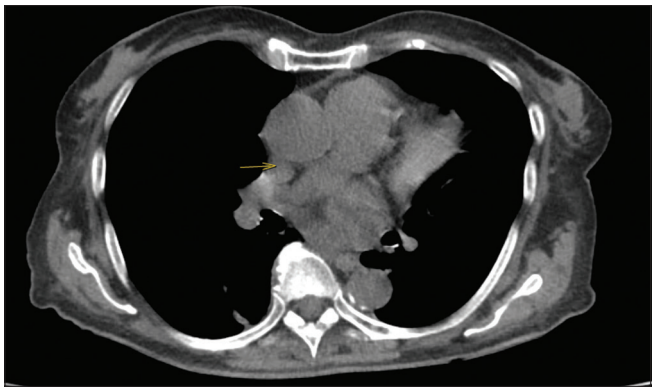

**Figure 1:** Thorax contrast-enhanced computed tomography showing mediastinal lymphadenopathy

Iron studies showed high ferritin (715 mcg/L), low serum iron (22 mcg/dL), and low total iron-binding capacity (180 mcg/dL). The plasma levels of cobalamin and folate were normal. The chest X-ray showed a widening of the mediastinum. Ultrasonography of the abdomen showed hepatosplenomegaly. Thorax contrast-enhanced computed tomography showed mediastinal lymphadenopathy [Figure 1]. C3 and C4 complement levels were normal. Serological testing for hepatitis B surface antigen, hepatitis C antibody, antinuclear antibody, antineutrophilic cytoplasmic antibodies, and human immunodeficiency virus antibody were all negative. Toxoplasma IgG and IgM antibody titers were normal.

Bone marrow aspirate showed trilineage maturation with suppression of myeloid series, hyperplasia of erythroid series, and megakaryocytic series. Bone marrow trephine biopsy showed well-formed epithelioid granulomas, which contained Langhans-type giant cells without necrosis and lacked surrounding inflammatory cells [Figure 2]. Acid-fast bacilli staining, culture for mycobacterium tuberculosis, atypical mycobacteria, and fungal staining were negative. The purified protein derivative test was negative. The serum angiotensin-converting enzyme was 214 U/L (normal level < 40 U/L). Endoscopic ultrasound-guided fine-needle aspiration from the mediastinal node also revealed noncaseating granuloma and was negative for lymphomas, tuberculosis, and fungal infections.

The patient was diagnosed to have sarcoidosis with bone marrow involvement (histopathology showing noncaseating granuloma and high serum angiotensin-converting enzyme after ruling out other causes of granulomatous disorders by appropriate tests). She was managed with prednisolone 20 mg daily, which was reduced and stopped after 3 months. On follow-up, she was asymptomatic and her blood counts were normal.

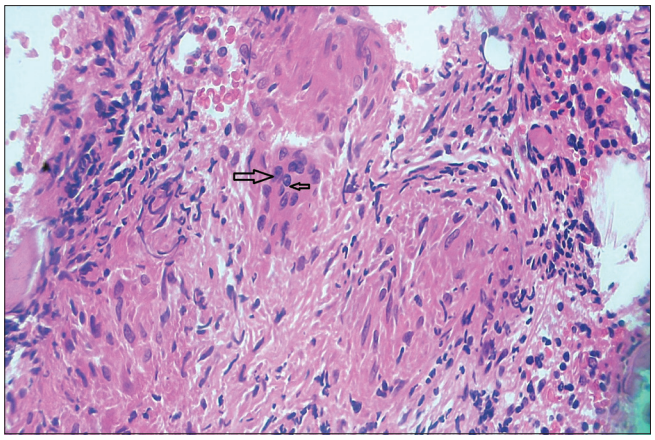

**Figure 2:** Bone marrow trephine biopsy showing well-formed epithelioid granulomas

## Discussion

Extrapulmonary manifestations of sarcoidosis can present before, concurrent with, or after the onset of pulmonary disease.[1] Since it is a multisystem disorder, the treating physicians should be aware of the atypical presentations. Bone marrow involvement in sarcoidosis is rare and was reported previously.[2-7]

Differential diagnoses in a woman with hepatosplenomegaly, mediastinal lymphadenopathy, and cytopenias include lymphoproliferative disorders or granulomatous infections like tuberculosis, atypical mycobacterium, and fungi with marrow infiltration. Bone marrow granulomas can be due to malignant, infectious, and autoimmune causes. This patient had hepatosplenomegaly, mediastinal lymphadenopathy, cytopenias, hypercalcemia with normal parathormone, and noncaseating granulomas in bone marrow and lymph node biopsy. These manifestations can be again due to lymphoproliferative disorders or granulomatous infections. Hypercalcemia can be associated with both Hodgkin's and non-Hodgkin's lymphomas due to raised circulating levels of 1,25(OH) 2D3 or parathyroid hormone-related protein.[8] Granulomatous infections like tuberculosis can be associated with hypercalcemia due to the extrarenal production of 1,25(OH) 2D3.[9] The patient was evaluated and was diagnosed to have sarcoidosis as the cause for her problems because she had no evidence of lymphoproliferative disorders or granulomatous infections on extensive evaluation and had a high serum angiotensin-converting enzyme level.

## Conclusion

Diagnosis of sarcoidosis should be considered always after ruling out other causes since it can mimic lymphoproliferative disorders and granulomatous infections. The initial presentation of sarcoidosis with bicytopenia due to bone marrow granulomas is

extremely rare and physicians should have awareness of such atypical presentations.

## Compliance with ethical standards

All procedures performed in studies involving human participants were by the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

## Informed consent

Informed signed written consent was taken from the patient involved.

## Acknowledgment

I would like to thank Dr.Asik Siddik, Department of Pathology, Maanu Memorial Hospital, Manjeri, Malappuram, Kerala, India, whose comments and suggestions were immensely valuable in improving this case report.

## Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

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