

Image TJH-2020-0357.R1

doi: 10.4274/tjh.galenos.2020.2020.0357

## Giant Cell Arteritis with Concomitant Chronic Myelomonocytic Leukemia

Mert Öztaş<sup>1</sup>, İlkin Muradov<sup>2</sup>, Abdülkadir Erçalışkan<sup>3</sup>, Ahu Senem Demiröz<sup>4</sup>, Şebnem Batur<sup>4</sup>, Ahmet Emre Eşkazan<sup>3</sup>, Serdal Uğurlu<sup>1</sup>

<sup>1</sup>Istanbul University-Cerrahpaşa, Cerrahpaşa Faculty of Medicine, Department of Medicine, Division of Rheumatology, İstanbul, Turkey

<sup>2</sup>Istanbul University-Cerrahpaşa, Cerrahpaşa Faculty of Medicine, Department of Internal Medicine, İstanbul, Turkey

<sup>3</sup>Istanbul University-Cerrahpaşa, Cerrahpaşa Faculty of Medicine, Department of Hematology, İstanbul, Turkey

<sup>4</sup>Istanbul Üniversitesi-Cerrahpaşa, Cerrahpaşa Faculty of Medicine, Department of Pathology İstanbul, Turkey

Serdal Uğurlu, İstanbul University-Cerrahpaşa, Cerrahpaşa Faculty of Medicine, Department of Medicine, Division of Rheumatology, İstanbul, Turkey  
+90 212 414 30 00

June 29, 2020

July 23, 2020

A 62-year-old man with no previous medical condition, was hospitalized with constitutional symptoms, which started within the past 2 months. He described recurrent fever ( $>39^{\circ}\text{C}$ ), weight loss, headache. Initial physical examination was unremarkable except for pallor of conjunctiva and fever ( $39.3^{\circ}\text{C}$ ). His hemoglobin was 7.5 g/dL (normal range, 13 –17.2 g/dL), his white blood cell (WBC) and platelet counts were  $22.8 \times 10^9/\text{L}$  (normal range,  $4.3 - 10.3 \times 10^9/\text{L}$ ) and  $58.3 \times 10^9/\text{L}$  (normal range,  $150 - 400 \times 10^9/\text{L}$ ) respectively. He also had remarkable monocytosis  $6.2 \times 10^9/\text{L}$  (normal range,  $0.3 - 0.9 \times 10^9/\text{L}$ ). He had an elevated CRP level at 150 mg/L (Normal $<5$  mg/L) and the erythrocyte sedimentation rate was 140 mm/h. His blood and urine cultures were sterile. For the suspicion of a vasculitis and in order to exclude concomitant malignancy, PET-CT was ordered, which showed both arcus and ascending aorta involvement without any signs of solid tumors (Figure 1). A diffuse bone marrow FDG uptake was also observed on PET-CT. Fragmentation of lamina elastica interna was detected in temporal artery biopsy without any sign of active vasculitis. Indeed, he was diagnosed as giant cell arteritis (GCA) according to his age, initial erythrocyte sedimentation level, headache, fever, and large vessel involvement in PET-CT. The patient was diagnosed as chronic myelomonocytic leukemia (CMML) with peripheral blood monocytosis ( $\geq 1 \times 10^9/\text{L}$  and

monocytes accounting for  $\geq 10\%$  of the WBC differential count (1). Peripheral blood smear revealed 8% blasts and bone marrow aspiration and biopsy were consisted with CMML-1 (Figure 2). Prednisolone 40 mg/daily was initiated, and his constitutional complaints were resolved. Azacytidine was planned to be initiated for CMML, but the patient decided to receive this treatment in his hometown.

While the pathogenic link between GCA and CMML has not been clearly established so far, the association is commonly observed between GCA and myelodysplastic syndromes (2).

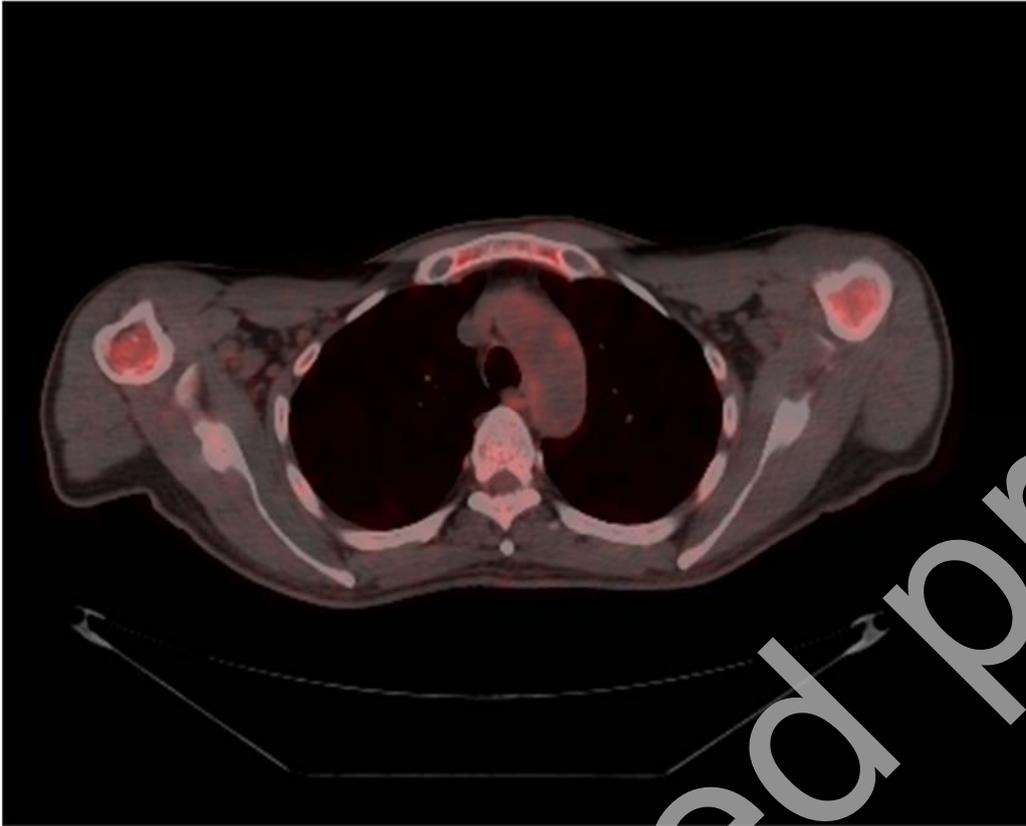
### **Author Contributions**

Concept: M.O., A.E.E.,S.U; Design: M.O.; Data Collection or Processing: I.M.,M.O,A.E,A.S.D, S.B.; Interpretation: M.O., A.E.E., S.U.; Literature Search: M.O.; Writing: M.O.,A.E.E.,S.U .

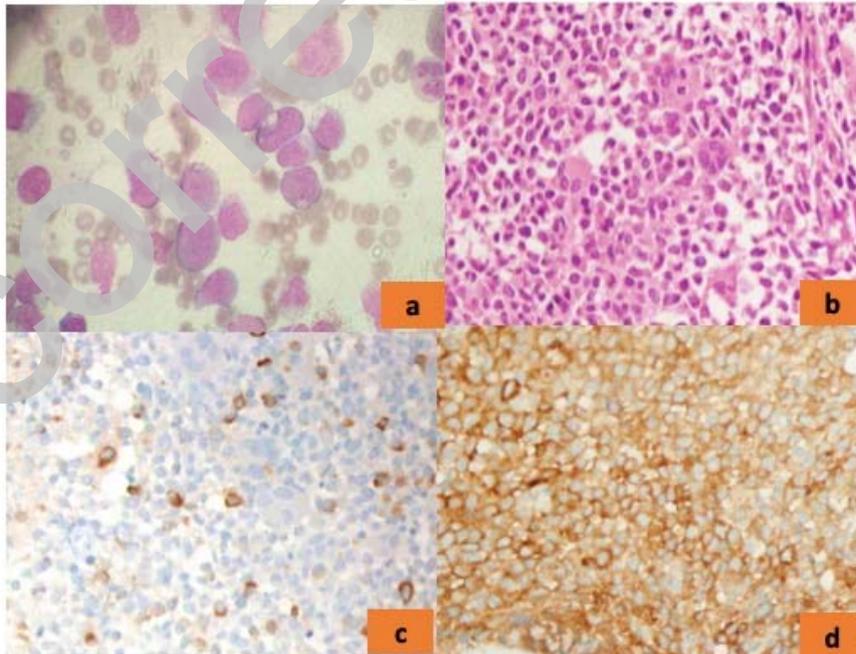
**Informed Consent:** Informed consent was obtained from the individual participant included in the study.

### **References**

1. Arber DA, Orazi A, Hasserjian R, et al. The 2016 revision to the World Health Organization classification of myeloid neoplasms and acute leukemia. *Blood*. 2016;127(20):2391-2405. doi:10.1182/blood-2016-03-643544
2. Roupie AL, de Boysson H, Thietart S, et al. Giant-cell arteritis associated with myelodysplastic syndrome: French multicenter case control study and literature review. *Autoimmun Rev*. 2020;19(2):102446. doi:10.1016/j.autrev.2019.102446



**Figure 1.** PET-CT showed aortic arch involvement and diffuse bone marrow FDG uptake.



**Figure 2.** Bone marrow aspiration shows monocytic cells over 20% and monoblasts 7%, dysplasia in all cell lineages. (a, Giemsa x1000). Bone marrow biopsy shows, hyperplasia, dysplasia in all cell lineages

(b, Hematoxylin&Eosine x400), immunohistochemically MPO is positive very few cells (c,x400). CD33 shows that, monocytic cells are increased (d, x400).

Uncorrected proof