LETTER TO THE EDITOR
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Erdheim Chester Disease Presenting with Bradicardia

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Author Contributions

A 65-year-old male patient applied to emergency department with complaints of headache and nausea. The patient's medical history included hypertension and chronic kidney disease. Electrocardiography revealed bradicardia and mobitz type 2 block. On physical examination, blood pressure was 157/85 mmHg, pulse rate was 43/min and oxygen saturation was 97% in room air. Blood tests revealed mild leukocytosis with white blood cell count of $12.62x10^3/\mu L$ (reference range: 3,9 – 10,9), creatinin of 1,66 mg/dL (reference range 0,70 – 1,20) and increased C-reactive protein of 46,2 mg/L (reference range: 0-5 mg/L). Aminotransferase levels were within reference range. The patient hospitalized for implantation of a permanent pace-maker. Pulse rate was normalized immediately after the procedure (72/min). During hospitalization, a non-contrast abdomen computed tomography performed due to patient's complain of back pain revealed para-aortic and perirenal soft tissue densities. Gerota's and Zukerkandl's fascias were thickened and renal calyces were dilated indicating possible uretheral involvement (Figure1A). Lower extremity x-ray showed osteosclerosis of distal femurs (Figure 1B). Echocardiography revealed biatrial dilatation and left ventricle hypertrophy. There was mild mitral and tricuspid valve insufficiency. Ejection fraction was %62. The patient consulted to interventional radiology for tissue diagnosis. Upon examination of the imaging findings before the procedure, biopsy of the perirenal soft tissue deemed more suitable. Pathology report confirmed CD68+, CD1a - foamy histiocytes. Further investigation with positron emission tomography showed FDG avid pseudotumor on right atrial wall (Figure 1C).
Erdheim Chester disease is a non-Langerhans cell histiocytosis characterized by mutation of the BRAF gene [1]. It is mostly diagnosed in the 5th-7th decades of life. It primarily affects the long bones of lower extremity but may also involve retroperitoneum, orbita, paranasal sinus, central nervous system(CNS), lung and heart. The diagnosis is challenging due to diverse presentation and rarity of the disease [2]. Involvement of the CNS presenting with diabetes insipidus, exophtalmus and cerebellar ataxia is seen in about half of the patients. Computed tomography may show hairy kidney sign implying perirenal fat stranding as a typical radiologic feature of the disease [3]. Paraaortic soft tissue densities and hydronephrosis secondary to uretheral compression resemble Ormond's disease. Hypertension due to renal artery compression may be present. Cardiac involvement is not rare in Erdheim-Chester disease affecting %40 of the patients [4]. Cardiac magnetic resonance imaging is the method of choice since it has the best soft tissue resolution. Cardiovascular manifestations include right atrial pseudotumor, high-degree conduction disorders, pericarditis and coronary artery infiltration [5]. All patients with ECD should be investigated for cardiac involvement to appropriately manage patients.
Interferon alfa has been used extensively in the treatment of ECD with response rates of %50-80. In recent years, targeted therapies including BRAF inhibitors (vemurafenib, encorafenib) are approved by the US Food and Drug Adminintration for BRAF-V600 mutant ECD. Surgery is not curative given the systemic nature of the disease but may be employed in selected cases which requires emergent palliation.

Fatih Hakan Tufanoğlu: Wrote the paper, Conceived and designed the analysis, Collected the data, Performed the analysis

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Conflict of interest
The Authors declare that there is no conflict of interest.

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