



Case Report

Aortitis presenting as acute myeloid leukemia - A case report

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ABSTRACT

Introduction: Aortitis can be an initial presentation of many diseases, but most often include large cell vasculitis.

Case presentation: We present a case of a 53 year old female who presented with a myriad of symptoms including abdominal pain, nausea, fever, and headaches. CT scan of the chest showed an inflamed aorta with an initial concern for a large cell vasculitis, but found later to be from her underlying acute myeloid leukemia (AML).

Discussion/conclusion: Our case emphasizes the importance of aortitis being the initial presentation of a multitude of diseases including malignancy. It remains important, especially for rheumatologists, to consider blood cancers when presented with aortitis.

Background/introduction: Aortitis can be an initial presentation of many diseases, but most often include large cell vasculitis. We describe a case of aortitis being the initial presentation of AML.

Methods: SCARE 2020 Guidelines.

1. Case presentation

A 53 year old female with a history of hyperlipidemia, anxiety, depression, chronic tobacco abuse, and history of one miscarriage presented to the ED for two weeks of feeling ill. She was in her usual state of health until two weeks prior to admission when she began to have fever, headaches, nausea, and abdominal pain. Associated symptoms included diarrhea, dyspnea on exertion and generalized body aches. She denied any past surgical history, family history, or drug allergies. Her symptoms were progressively getting worse. She denied any recent travel or any new medication intake prior to the onset of the symptoms. Review of systems was negative for joint swelling, joint pain, hematuria, cough, or skin rash.

On admission, she was afebrile with a normal blood pressure and heart rate. Physical exam was positive for pale skin and a diffusely tender abdomen. Basic metabolic panel showed normal creatinine and electrolytes. Complete blood count revealed a Hgb of 6.8 g/dL and platelet count $32,000 \times 10^9/L$. She received 1 unit of packed RBC's, and repeat Hgb was 8.3 after the transfusion. Other pertinent labs include LDH 324 units/L, CRP 133 mg/dL, and absolute reticulocyte count

$<0.010 \times 10^9/L$. The urinalysis was negative for blood or protein, and the hepatic function panel was within normal limits. The ANA and ANCA panel was negative. Infectious work up, including blood cultures, EBV, Parvovirus, and CMV, were negative. CTA chest showed central lobular emphysema without evidence of pneumonia and minimal pleural and parenchymal scarring CT chest, CT abdomen and pelvis revealed fat stranding and mild inflammatory changes centered around the infrarenal abdominal aorta, concerning for aortitis (Fig. 1).

Given the inflamed aorta, concern for large cell vasculitis grew. Rheumatology was consulted, and recommended more labs including complement 3 and 4, rheumatoid factor, anti-smooth muscle antibody, hepatitis panel, anti beta 2 glycoprotein antibody, anticardiolipin antibody and cryoglobulin were negative. With the history of miscarriage, an anti-phospholipid syndrome panel and lupus inhibitor was ordered, which were positive. Haptoglobin levels were found to be 327 mg/dL. Concern for underlying malignancy grew, and a bone marrow aspirate was done by a hematologist. Aspirate showed hypercellular bone marrow (100%) with dysgranulopoiesis, dysmegakaryopoiesis, and decreased erythroid precursors, consistent with acute myeloid leukemia. Further lab testing revealed monosomal karyotype and TP53 mutation

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Fig. 1. Inflammation of the aorta noted in the CT abdomen and pelvis.

(approximately 25% blasts) and plan was to start outpatient chemotherapy. Patient was discharged home with rheumatology, primary care, and oncology follow up. On clinic follow ups, she is doing well tolerating chemotherapy.

2. Discussion

Aortitis is a broad term describing inflammation of the aorta. Common causes include large cell vasculitis, diseases predominantly involving the aorta and its major branches. Major examples include giant cell arteritis and takayasu arteritis, both of which can be life threatening. The hallmark of giant cell arteritis is inflammation originating from the aorta. The majority of symptoms of temporal arteritis result from the involvement of the cranial branches of the aorta although the involvement of other large vessels such as thoracic/abdominal aorta and its branches is not uncommon [1]. Takayasu arteritis also involves the aorta, specifically adventitial and periadventitial fibrosis [2]. However, aortitis has been reported in cases of hematologic malignancies.

AML is the most common leukemia among the adult population and accounts for 80% of cases. Common presentation includes recurrent infections, easy bleeding and anemia, but symptoms can be non-specific. In our patient, the initial presentation was consistent with a malignancy but was found to have mild inflammatory changes around the aorta consistent with aortitis. While uncommon, aortitis has been reported as an association with hematologic malignancy. From *Spooner, 2018*, a 60 year old male presented with aortitis found to be associated with myelodysplastic syndrome, likely representing chronic myeloid leukemia (CML). In this case, the patient was treated initially with intravenous steroids believing he had large cell vasculitis [3]. From *Sasinowka 2017*, a 68 year old female with a known diagnosis of CML presented for evaluation of an abnormal chest computed radiography, showing inflammation around the thoracic and abdominal aorta. For this patient, they attributed this finding to a paraneoplastic autoimmune phenomenon, and started the patient on intravenous steroids for treatment [4]. Another case of a 44 year old female diagnosed with acute promyelocytic leukemia was found to have retroperitoneal soft tissue thickening around the abdominal aorta with aortic wall related to associated *paraneoplastic aortitis* [5].

Our patient also had a history of miscarriage, uncommon from the two previous reports, and further lab work up confirmed a positive antiphospholipid. It is unclear if the anti-phospholipid picture is related to

the aortitis, but there have been previous reports of the two coexisting together. From *Gonzalez-Garcia 2020*, a 64-year old male presented with severe thrombosis affecting the spleen and kidney, found to have aortitis. He had a history of systemic lupus erythematosus and antiphospholipid syndrome (APS); his clinical course improved with anticoagulation and plasma exchange [6]. The Sapporo criteria require one clinical criterion (vascular thrombosis or pregnancy mortality before 10 weeks) and one laboratory criterion (one of lupus anticoagulant, anticardiolipin antibody, or anti-β2-glycoprotein antibody are positive twice at least 12 weeks apart) for diagnosis of APS [7]. Our patient did not have any concomitant thromboses nor met the criteria for APS, therefore APS was unlikely to be a contributing factor to the aortitis. Finally, this work has been reported in accordance with SCARE [8].

3. Conclusion

Aortitis is a predominant initial presentation in many large cell vasculitis, prompting urgent rheumatologic evaluation. However, it can be the presenting sign in a multitude of diseases, including malignancies. It remains important for clinicians, especially rheumatologists, to consider blood cancers as a differential for aortitis.

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Consent from the patient

Consent has been taken from the patient.

Consent

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Author contributions

Branden Ireifej, Majd Freijat: Main author, data formulation.
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None declared.

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