

# Malignancy Associated Aortitis in a Patient with Fever of Unknown Origin

## Nedeni Bilinmeyen Ateş Etiyolojisinde Malignite İlişkili Aortit Olgusu

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### Abstract

Aortitis, characterized by inflammatory changes in the aortic wall, can present with various manifestations. It should be considered in the differential diagnosis of fever of unknown origin (FUO), especially in patients with aortic aneurysm. Accurate differentiation between infectious and non-infectious aortitis is essential for appropriate treatment. Clinicians must also remain vigilant regarding the potential malignancy risk in patients with vasculitis during initial evaluation and follow-up. This study presents a case of malignancy-associated large vessel vasculitis identified in a patient with FUO.

**Keywords:** Aortitis, large vessel vasculitis, fever of unknown origin

### Öz

Aort duvarındaki enflamatuvar değişikliklerle karakterize olan aortit, kendini çeşitli bulgularla gösterebilir. Özellikle aort anevrizması olan hastalarda, nedeni bilinmeyen ateş (NBA) etiyolojisinde akılda tutulmalıdır. Enfeksiyöz ve enfeksiyöz olmayan aortitin ayırıcı tanısı, hastaların uygun tedavisi için yapılmalıdır. Ayrıca, vaskülit tanısı konmuş hastalarda malignite riski konusunda klinisyenlerin farkındalığı, başlangıç değerlendirmesi ve takip için hayati öneme sahiptir. Bu çalışmada, NBA'sı olan malignite ile ilişkili büyük damar vaskülitini tanıyan konmuş bir hasta sunulmuştur.

**Anahtar Kelimeler:** Aortit, büyük damar vaskülitini, nedeni bilinmeyen ateş

### Introduction

Aortitis, defined by inflammatory alterations in the aorta wall, is an important etiology of fever of unknown origin (FUO) as it can present as fever (1,2). Differential diagnosis between infectious and non-infectious aortitis must be made for appropriate treatment of patients (3). The clinical properties of vasculitis are highly variable, so the clinicians must have a strong suspicion to achieve a reliable and definite diagnosis (4). In addition to complexity of the disease, clinicians should be

aware of increased risk of malignancy in these patients (5). In this study, a patient with malignancy-associated large vessel vasculitis diagnosed with FUO is presented.

### Case Presentation

A 65-year-old male patient exhibited fever on the second postoperative day while hospitalized in the cardiovascular surgery ward. The patient had recently undergone thoracic endovascular aneurysm repair (TEVAR) and aortoiliac bypass surgery due to thoracic aortic aneurysm. He had a history

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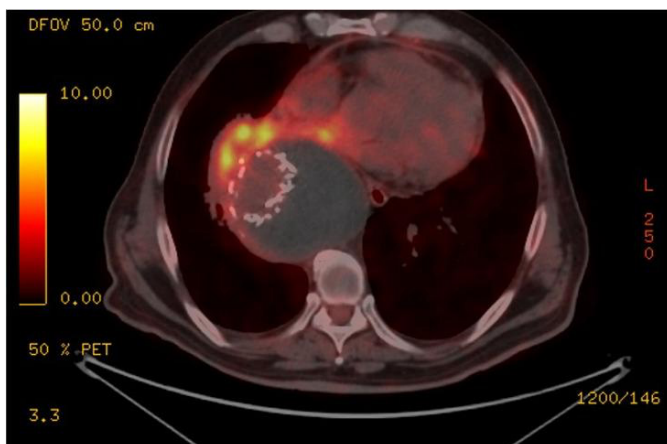
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of two TEVAR performed two years ago for thoracic and abdominal aortic aneurysms. Piperacillin-tazobactam and teicoplanin was empirically initiated for fever on the second postoperative day. He had no additional symptoms other than ongoing fever. Transthoracic echocardiography was normal. Antibiotherapy was discontinued at day 14, due to the absence of obvious focus of infection and culture negativity (blood and urine).

The patient was evaluated for FUO. There was no specific epidemiological risk factors for infectious diseases. Laboratory parameters indicated the presence of anemia and persistent C-reactive protein elevation not accompanied by leukocytosis and elevated procalcitonin. No pathological findings was detected in endoscopy-colonoscopy performed for the investigation of anemia etiology and malignancy screening. Positron emission tomography/computed tomography (PET/CT) which was performed to screen for malignancy and rheumatological diseases revealed increased metabolic activity in the anterior wall of the aneurysmal segment in the thoracic aorta and in the posterior wall at the level of the proximal iliac bifurcation compatible with aortitis (SUV<sub>max</sub> 16.3) (Figure 1). In addition to PET/CT findings antinuclear antibody positivity was detected among rheumatological tests. The laboratory tests for brucellosis, syphilis and Q fever were negative. When all the clinical, radiological and laboratory findings were evaluated together, the patient was diagnosed with non-infectious inflammatory large vessel vasculitis/aortitis. Methylprednisolone and methotrexate was initiated by the rheumatologist. Infectious aortitis was excluded in the patient who had clinical improvement and fever response to the immunosuppressive treatment. Due to the diagnosis of vasculitis, whole body CT was performed to screen for malignancy. A nodular solid lesion with 28 mm diameter was detected in the right kidney and biopsy taken



**Figure 1:** The abnormal metabolic activities in the anterior wall of the aorta in PET/CT

PET/CT: Positron emission tomography/computed tomography imaging

from the lesion revealed papillary renal cell carcinoma type 1. The radiofrequency ablation of the lesion was performed. The patient was discharged with recovery and followed up with immunosuppressive treatment for vasculitis.

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

## Discussion

Aortitis is often an unanticipated diagnosis in patients being investigated for an unexplained fever, chest or abdominal pain depending on the region of aortic involvement (6). As the symptoms are nonspecific a high level of clinical suspicion is necessary for an accurate diagnosis of aortitis (7). Aortitis may encompass a variety of conditions with different prognostic features (1,2). Carrer et al. (8) showed that infectious aortitis had higher mortality rates compared to non-infectious aortitis. Differential diagnosis between infectious and non-infectious aortitis must be made, because there are significant differences in the treatment strategies (3). Corticosteroids and immunosuppressive agents for disease activity control is the mainstay of the treatment of non-infectious aortitis (3). Here we presented a case diagnosed with malignancy-associated non-infectious aortitis successfully treated with methylprednisolone and methotrexate.

The clinical spectrum of vasculitis can range from localized involvement to multiple organ manifestations with high morbidity and mortality. In addition to the complexity of the disease, clinicians should be aware of the increased risk of malignancy in these patients (5). Not only malignancy risk can be increased in some types of vasculitis but also vasculitis can be seen as a sign of malignancy (5). Screening for malignancy in patients with vasculitis has an important impact on patient survival as it improves the diagnosis and therapy. In this case, the patient was diagnosed with renal cell carcinoma associated vasculitis and had the opportunity to promptly obtain appropriate intervention for malignancy.

It is important for cardiovascular surgeons to be aware of the inflammatory etiology of aortic aneurysms. As surgical or percutaneous interventions including stenting and ballooning have a high risk of recurrence, vasculitic manifestations in other parts of the aorta and requirement for re-operations in vasculitis (3). Elective reconstructive surgery and endovascular interventions are most likely to succeed when performed in stable remission phases under immunosuppressive treatment (3,4). In case of urgent surgical indications like arterial vessel dissection, large aneurysm, or critical vascular ischemia, the patients should be referred to a vascular team (4). Using perioperative corticosteroids should be considered and postoperative disease activity should be controlled with close monitoring. The risk for

possible complications like anastomotic site aneurysms should be kept in mind of surgeons (3).

## Conclusion

As a conclusion, the patient was diagnosed with malignancy associated large vessel vasculitis/aortitis with PET/CT as the etiology of FUO. The inflammatory changes in aortitis often manifest with fever, and the timing and outcomes of medical and surgical treatment depend on the timely and accurate diagnosis. These patients should be evaluated by a multidisciplinary team consisting of cardiovascular surgeon, infectious disease specialist and rheumatologist.

## Ethics

**Informed Consent:** Written informed consent was obtained from the patient for publication of this case report and accompanying images.

## Footnotes

### Authorship Contributions

Surgical and Medical Practices: E.M.S., G.B., E.G., G.Ç., İ.A., A.A., Concept: E.M.S., G.B., E.G., G.Ç., İ.A., M.C.S., A.G., E.Ö., A.A., Design: E.M.S., G.B., E.G., G.Ç., İ.A., A.A., Data Collection and/or Processing: E.M.S., G.B., E.G., G.Ç., İ.A., M.C.S., A.G., E.Ö., A.A.,

Analysis and/or Interpretation: E.M.S., G.B., E.G., G.Ç., İ.A., M.C.S., A.G., E.Ö., A.A., Literature Search: E.M.S., G.B., E.G., G.Ç., İ.A., A.A., Writing: E.M.S., G.B., E.G., G.Ç., İ.A., M.C.S., A.G., E.Ö., A.A.

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