CASE REPORT

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A Young Woman Diagnosed with Takayasu Arteritis: A Comprehensive Case Report

Takayasu Arteriti Tanısı Almış Genç Bir Kadın: Kapsamlı Bir Olgu Sunumu

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ABSTRACT

Takayasu arteritis (TAK) is a rare, chronic large-vessel vasculitis that primarily affects the aorta and its major branches. It is more prevalent in Asian populations, with significant geographical variations. The disease predominantly affects women, typically in their second or third decade of life. Diagnosis is challenging due to its non-specific symptoms and lack of definitive laboratory tests, relying mainly on clinical suspicion and imaging. The 2022 American College of Rheumatology/European League Against Rheumatism criteria for TAK diagnosis require patients to be under 60 and have imaging findings consistent with large-vessel vasculitis. Additional clinical and imaging features contribute to a scoring system, with a total of 5 or more confirming the diagnosis. This case report describes a 26-year-old woman with no previous medical conditions, presenting with recurrent fever and weight loss. Clinical examination revealed a difference in pulse and blood pressure between the arms, suggestive of vascular involvement. Laboratory findings included elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) levels, with no other abnormalities. Computed tomography (CT) angiography did not reveal vascular changes, but positron emission tomography (PET)/CT demonstrated mild increased fluorodeoxyglucose uptake in the right subclavian artery, consistent with vasculitis. The patient was diagnosed with TAK and treated with methotrexate and corticosteroids, resulting in significant improvement in inflammatory markers and clinical symptoms. By the sixth month, both ESR and CRP had normalized. This case highlights the importance of evaluating acute phase reactants, comparing pulses and blood pressure in both arms, and considering TAK in young female patients with recurrent systemic symptoms. Imaging plays a central role, with PET/CT offering superior sensitivity in detecting early inflammatory changes compared to CT angiography. The patient's classification as type I TAK, with isolated subclavian artery involvement, aligns with common angiographic patterns seen in Türkiye. Overall, this case underlines the diagnostic value of PET/CT and reinforces awareness of TAK's variable presentation.

Keywords: Takayasu arteritis, vasculitis, subclavian artery, PET/CT

ÖZ

Takayasu arteriti (TAK), başlıca aorta ve aortanın ana dallarını tutan nadir, kronik bir büyük damar vaskülitidir. Asya toplumlarında daha sık görülmekte olup belirgin coğrafi farklılıklar göstermektedir. Hastalık çoğunlukla kadınları etkiler ve genellikle yaşamın ikinci veya üçüncü dekadında ortaya çıkar. Tanı, özgül olmayan semptomlar ve kesin tanı koydurucu laboratuvar testlerinin bulunmaması nedeniyle güçtür; bu nedenle temel olarak klinik şüphe ve görüntüleme bulgularına dayanır. Takayasu arteriti tanısı için 2022 American College of Rheumatology/European League Against Rheumatism kriterlerine göre hastaların 60 yaşın altında olması ve büyük damar vasküliti ile uyumlu görüntüleme bulgularının bulunması gerekmektedir. Ek klinik ve görüntüleme özellikleri puanlama sistemine katkı sağlar ve toplam puanın 5 veya üzerinde olması tanıyı destekler. Bu olgu sunumunda, daha önce bilinen herhangi bir hastalığı olmayan, tekrarlayan ateş ve kilo kaybı ile başvuran 26 yaşında bir kadın hasta sunulmaktadır. Klinik muayenede kollar arasında nabız ve kan basıncı farkı saptanmış olup bu bulgu vasküler tutulumu düşündürmüştür. Laboratuvar bulgularında eritrosit sedimantasyon hızı (ESR) ve C-reaktif protein (CRP) düzeylerinde yükseklik izlenmiş, bunun dışında ek bir anormallik saptanmamıştır. Bilgisayarlı tomografi (BT) anjiyografide vasküler değişiklik izlenmezken, pozitron emisyon tomografisi (PET)/BT'de sağ subklavian arterde vaskülit ile uyumlu hafif artmış florodeoksiglukoz tutulumu gösterilmiştir.

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Hastaya TAK tanısı konulmuş ve metotreksat ile kortikosteroid tedavisi başlanmıştır. Tedavi sonucunda enflamatuvar belirteçlerde ve klinik semptomlarda belirgin iyileşme sağlanmıştır. Altıncı ayda hem ESR hem de CRP düzeyleri normale dönmüştür.

Bu olgu, tekrarlayan sistemik semptomları olan genç kadın hastalarda akut faz reaktanlarının değerlendirilmesinin, her iki kolda nabız ve kan basıncının karşılaştırılmasının ve TAK tanısının akılda tutulmasının önemini vurgulamaktadır. Görüntüleme tanıda merkezi bir role sahiptir; PET/BT, erken enflamatuvar değişikliklerin saptanmasında BT anjiyografiye kıyasla daha yüksek duyarlılık sağlayabilir. Hastanın izole subklavian arter tutulumu ile tip I TAK olarak sınıflandırılması, Türkiye'de görülen yaygın anjiyografik paternlerle uyumludur. Genel olarak bu olgu, PET/BT'nin tanısal değerini ortaya koymakta ve TAK'ın değişken klinik sunumu konusunda farkındalığın önemini desteklemektedir.

Anahtar Kelimeler: Takayasu arteriti, vaskülit, subklavyen arter, PET/BT

INTRODUCTION

Takayasu arteritis (TAK) is an uncommon, persistent inflammatory disease targeting large blood vessels, primarily the aorta and its main branches[1]. Although TAK occurs globally, it is believed to affect the Asian population disproportionately more. In Japan, the highest recorded prevalence has been estimated at 40 cases per million people, whereas in the United States, the lowest documented rate is approximately 0.9 cases per million[2]. In a study conducted in the United Kingdom, the average prevalence of TAK was determined to be 4.7 cases per million population[3]. In a retrospective cohort study comprising 1,011 patients diagnosed with TAK, the mean age at disease onset was 32.5±15.1 years. Of the total study cohort, 763 participants were female, comprising 75.5% of the population and yielding a female-to-male ratio of 3.1:1[4]. In another cohort study involving 852 patients diagnosed with TAK, the median age at disease onset was reported to be 25 years[5]. The female-to-male ratio demonstrates regional variability. In a study conducted in Southern Norway, the female-to-male ratio was reported as 6.8:1, whereas in the United States, this ratio was identified as 4.2:1[6,7]. This condition is broadly recognized as panarteritis, with the earliest signs of inflammation commonly appearing near the vasa vasorum and at the interface between the media and adventitia layers[8].

Vascular remodeling characterized by segmental stenosis and/or aneurysm formation is a hallmark of TAK. Gaining insight into the underlying mechanisms of TAK is essential for advancing accurate diagnostic approaches and effective treatment methods. The disease is believed to have an autoimmune basis, in which dysregulated immune responses lead to chronic vascular inflammation. Because clinical symptoms are often vague and there are no conclusive diagnostic tools available, making an accurate diagnosis is especially difficult[9].

Invasive modalities such as angiography have largely been replaced by non-invasive options like fluorodeoxyglucose-positron emission tomography (FDG-PET) scanning, computed tomography angiography (CTA), magnetic resonance imaging/angiography (MRI/MRA), and ultrasound (US). An optimal imaging modality should facilitate early identification of vascular wall inflammation, offer precise delineation of vascular morphology, correlate reliably with clinical disease activity, exhibit sensitivity to therapeutic interventions and possess prognostic value in anticipating future vascular complications[10]. PET/CT has been recognized for its significant diagnostic value in the early identification of vasculitis and is therefore strongly recommended as an imaging modality by both the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR)[11,12].

According to the TAK classification criteria updated by ACR/EULAR in 2022, being under 60 years of age at the time of diagnosis and having imaging findings consistent with large-vessel vasculitis are mandatory requirements for diagnosis. In patients who meet the initial criteria, additional clinical and imaging features are evaluated and scored. A total score of 5 or more confirms the classification of TAK[13].

CASE REPORT

A 26-year-old woman with no prior medical conditions, no regular medications, and no family history of rheumatic diseases presented to the internal medicine outpatient clinic complaining of fever and weight loss. The patient reported that her fever had been recurring every two-three days for the past four years. She reported an unintentional weight loss over the past six months, decreasing from 55 kg to 50 kg. There was no morning stiffness or Raynaud's phenomenon. The pulse rate was recorded as 62 beats per minute in the right upper extremity and 84 beats per minute in the left. Additionally, the pulse amplitude in the right arm was notably diminished compared to the left. Apart from the observed discrepancy in blood pressure between the arms (116/72 mmHg in the right and 133/84 mmHg in the left), physical examination revealed no other abnormalities. Cardiac auscultation demonstrated normal, rhythmic first (S1) and second (S2) heart sounds, without any audible additional heart sounds, gallops, or murmurs. In laboratory tests, no pathology was detected except for an elevated erythrocyte sedimentation rate (ESR) of 63 mm/h and a C-reactive protein (CRP) level of 151 mg/L. The patient's DNA was analyzed for 18 mutations in the *MEFV* gene, with results reported as negative.

Radiological examinations, including neck ultrasonography, revealed significant intima-media thickening measuring up to 2.8 mm in the bilateral common carotid arteries and the right subclavian artery. Considering TAK, CT angiography of the aorta, its branches, and the upper extremity vessels, as well as a FDG-PET scan, were requested. CT angiography of the neck, thoracic aorta, upper extremities, and abdomen showed no stenosis or vascular wall pathology. However, the PET/CT scan demonstrated diffuse mild increased FDG uptake consistent with vasculitis in an approximately 2.5 cm segment of the right subclavian artery extending from the thyroid to the thoracic inlet (see Figures 1, 2). The patient was diagnosed with TAK. A patient experiencing recurrent febrile episodes every two to three days over the past four years, accompanied by a 5 kg unintentional weight loss within the last six months, was diagnosed with TAK one month following her initial presentation to the rheumatology outpatient clinic. Treatment was initiated with methotrexate 15 mg/week, folic acid 5 mg/week, and oral methylprednisolone 40 mg/day. The patient's inflammatory indicators decreased, and both clinical condition and laboratory results demonstrated signs of recovery. Following the initiation of treatment, a reduction of over 50% in ESR and approximately 80% in CRP levels was achieved within 3 months. By the 6th month, ESR had decreased to 5 mm/h and CRP to 0 mg/L (see Table 1). The patient is currently being followed in a stable clinical condition.

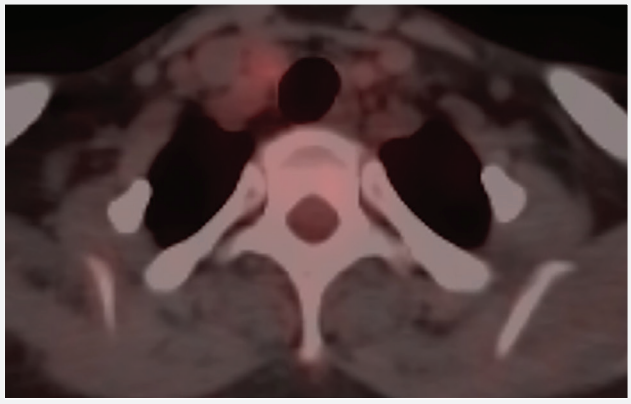

Figure 1. Diffuse, mild increased FDG uptake in the right subclavian artery, axial plane, PET/CT

FDG: Fluorodeoxyglucose, PET/CT: Positron emission tomography/computed tomography

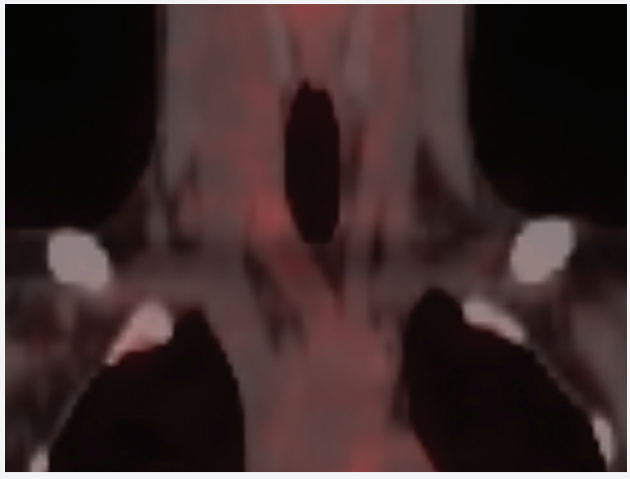

Figure 2. Diffuse, mild increased FDG uptake in the right subclavian artery, coronal plane, PE/CT

FDG: Fluorodeoxyglucose, PET/CT: Positron emission tomography/computed tomography

DISCUSSION

This case is presented to emphasize the importance of evaluating acute phase reactants in the laboratory, measuring blood pressure and pulses in both arms during physical examination, and considering TAK in the differential diagnosis, especially in young female patients with recurrent fever attacks and weight loss. In cases with strong suspicion of TAK, angiographic imaging should be performed, and PET/CT evaluation considered when clinically indicated.

TAK was initially documented in 1908 by Mikito Takayasu, an ophthalmology professor at Kanazawa University in Japan[14]. The disease is seen predominantly in women (84.6%) with a mean age of onset of 32.3 (±10.4) years[13]. TAK is significantly more common in women than men in Türkiye as well (F/M ratio: 8.2/1), and the average age of onset is 40.1 (±14) years[15]. This case is consistent with the literature in terms of age and gender.

TAK shows geographical variations in distribution. In Asian countries, both the incidence and prevalence are relatively higher. The condition affects between 3.3 and 40 individuals per million, with yearly new cases estimated to fall between 0.34 and 2.4 per million people[16]. It predominantly occurs in regions such as Japan, Southeast Asia, India, and Mexico[17]. A study carried out in Türkiye determined the yearly occurrence rate of TAK to be 1.11 per million individuals, with a 95% confidence interval ranging from 0.54 to 1.67[18].

Table 1. The patient's laboratory values at the initial presentation, 3-month, and 6-month follow-up examinations

Presentation	ESR (mm/h)	CRP (mg/L)	WBC (x10^3/µL)	HGB (g/dL)
Initial presentation	63	151	8.9	10.3
3-month	26	28	5.06	11.1
6-month	5	0	9.23	13.1

ESR: Erythrocyte sedimentation rate, CRP: C-reactive protein, WBC: White blood cell, HGB: Hemoglobin

In published studies on TAK, systemic manifestations like fever (34.7%) and fatigue (12.1%) are the most frequently reported early symptoms, with neck pain (9.7%) following as a less common initial sign[19]. This case also presented to the hospital with fever, which is the most common and non-specific symptom of TAK. Weight loss (9.3%) is an relatively uncommon finding in TAK[20]. In this case, however, weight loss was present. Other complaints observed in patients include syncope (6.98%) and night sweats (4.65%), which were not observed in this case[20]. In a study conducted on patients with TAK, the frequency of having a blood pressure difference between the two arms was found to be 53.8%[21]. In this particular instance, a discrepancy in blood pressure readings between the two arms was also observed.

Imaging plays an important role in the diagnosis and follow-up of TAK. CT angiography is the preferred initial imaging modality due to its availability, better image resolution compared to MR angiography, and lower cost. It serves an important function in pinpointing the regions involved and evaluating the intensity of TAK. CT angiography can reveal changes such as luminal narrowing, dilatation, vessel wall thickening, calcification, and contrast enhancement[22]. PET/CT represents an alternative imaging modality for the evaluation of TAK. As a non-invasive method, PET/CT is becoming an increasingly valuable tool in the diagnostic evaluation of TAK. PET/CT can detect increased metabolic activity in the arterial wall and provide information about vascular inflammation[11]. The capacity of FDG-PET imaging to identify inflammatory activity during the early, pre-edematous phase—prior to the onset of overt vascular structural alterations—represents a clinically valuable advantage. Nonetheless, its broader implementation is constrained by limited accessibility and substantial financial burden[23]. In the assessment of TAK activity, PET/CT parameters including maximum standardized uptake value (SUV_{max}), vascular SUV_{max}-to-liver mean SUV ratio (SUV_{ratio}), and PET vascular activity score (PETVAS) are utilized. As reported in a study, the sensitivity and specificity of SUV_{max}, SUV_{ratio}, and PETVAS for evaluating TAK activity using PET/CT were reported as 100% and 100%, 100% and 90.9%, and 88.9% and 90.9%, respectively[24]. In the case which presented here, although no findings suggestive of TAK were detected on CT angiography, arterial involvement was identified on PET/CT. In the comparative study conducted by Quinn et al.[25] evaluating MRA and FDG-PET in patients with TAK, FDG-PET–derived assessments of disease activity demonstrated a correlation with clinical status. When benchmarked against MRA findings, FDG-PET exhibited a sensitivity of 71% and a specificity of 52% in detecting active disease. Therefore, MRA was not performed in this patient. Digital subtraction angiography (DSA) remains the definitive diagnostic technique for TAK, as it enables detailed imaging of the coronary vessels and facilitates evaluation of pressure within the aorta[26]. Given the vascular involvement identified on PET/CT in this case, it was deemed that a DSA examination might not be required.

The Numano classification, introduced in 1996, remains the most widely utilized angiographic system for categorizing TAK. Based on this classification system, TAK is categorized angiographically according to the specific vessels involved. Type 1 includes the aortic arch branches, such as the brachiocephalic trunk, carotid arteries, and subclavian arteries. Type 2a encompasses the ascending aorta, the aortic arch, and its branches. Type 2b includes all the features of Type 2a with additional involvement of the thoracic descending aorta. Type 3 refers to disease affecting the thoracic descending aorta, abdominal aorta, and/or renal arteries. Type 4 is limited to the abdominal aorta and/or renal arteries. Type 5 presents a combination of vascular involvement seen in both Type 2b and Type 4[27]. In a study evaluating patients with TAK in Türkiye angiographically, the most common types were found to be Type V (51%) and Type I (32%). In the same study, the most frequently involved arteries were observed to be the subclavian (76%), carotid (52%), and renal (28%) arteries, respectively[15]. Current case's PET/CT examination revealed involvement of the right subclavian artery and the patient was classified as Type 1 TAK.

CONCLUSION

In this study, a rare case of TAK was presented. This case is consistent with the literature due to being young and female, presenting to the hospital with fever, having elevated inflammatory markers in laboratory tests, exhibiting a blood pressure difference between the two arms and involvement of the subclavian artery. The detection of vascular involvement on PET/CT rather than on CT angiography supports the increasing importance of PET/CT in the diagnosis of TAK.

Ethics

Informed Consent: The patient provided written informed consent for the publication of this case report, including all associated clinical details and images.

Footnotes

Authorship Contributions

Concept: Ö.A.S., D.B.G., B.S.S., R.M., Design: Ö.A.S., D.B.G., B.S.S., R.M., Data Collection or Processing: Ö.A.S., D.B.G., R.M., Analysis or Interpretation: Ö.A.S., D.B.G., B.S.S., R.M., Literature Search: Ö.A.S., D.B.G., B.S.S., R.M., Writing: Ö.A.S.

Conflict of Interest: No conflict of interest was declared by the authors.

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