Interesting Image
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^{18}F-Fluorodeoxyglucose Positron Emission Tomography/Magnetic Resonance Imaging Appearance of Gastrointestinal Behcet's Disease

Gastrointestinal Behçet Hastalığının ^{18}F-Florodeoksiglukoz Pozitron Emisyon Tomografisi/ Manyetik Rezonans Görüntüleme Bulguları

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Abstract

Behcet's syndrome (BS) is a variable vessel vasculitis with multi-organ involvement. Recurrent episodes of oral and genital ulcers, papulopustular and erythema nodosum-like skin lesions, and arthritis are relatively more frequent, whereas uveitis, venous and arterial lesions, nervous system, and gastrointestinal involvement are less common, but are severe manifestations. The frequency of gastrointestinal involvement shows important variation between countries as more common in the Far East and the United States, and much less common in Turkey and the Middle East. The main clinical signs of gastrointestinal Behcet's disease include abdominal pain, diarrhea, blood in the stool, fever, and weight loss. Ulcers seen in the terminal ileum, cecum, and ascending colon are common endoscopic findings. Herein, we presented the positron emission tomography/ magnetic resonance imaging findings of gastrointestinal involvement in BS.

Keywords: Gastrointestinal Behcet's disease, Behcet syndrome, myelodysplastic syndromes, monosomy 7, acute myeloid leukemia, ^{18}F-FDG, PET scan, MRI

Öz

Behçet sendromu (BS) multi-organ tutulumu ile seyreden bir değişken damar vaskülitidir. Tekrarlayan oral ve genital ülserler, papülopüstüler ve eritema nodozum benzeri deri lezyonları ve artrit epizodları nispeten daha sık görülürken, üveit, venöz ve arteriyel lezyonlar, sinir sistemi tutulumu ve gastrointestinal tutulum daha az yaygın olmakla birlikte şiddetli belirtilerdir. Gastrointestinal tutulum sıklığı ülkeler arasında farklılık göstermekte olup, Uzak Doğu ve Amerika Birleşik Devletleri'nde daha sık görülürken, Türkiye ve Orta Doğu'da daha nadir görülmektedir. Gastrointestinal Behçet hastalığının başlıca klinik bulguları karın ağrısı, ishal, kanlı dışkılama, ateş ve kilo kaybıdır. Terminal ileum, çekum ve çıkan kolonda görülen ülserler ise sık görülen endoskopik bulgulardır. Bu olgu sunumunda, gastrointestinal BS'nin, daha önce yayınlanmamış, pozitron emisyon tomografisi/ manyetik rezonans görüntüleme bulgularını göstermeyi hedefliyoruz.

Anahtar kelimeler: Gastrointestinal Behçet hastalığı, Behçet sendromu, myelodisplastik sendrom, monozomi 7, akut myeloid lösemi, ^{18}F-FDG, PET tarama, MRG

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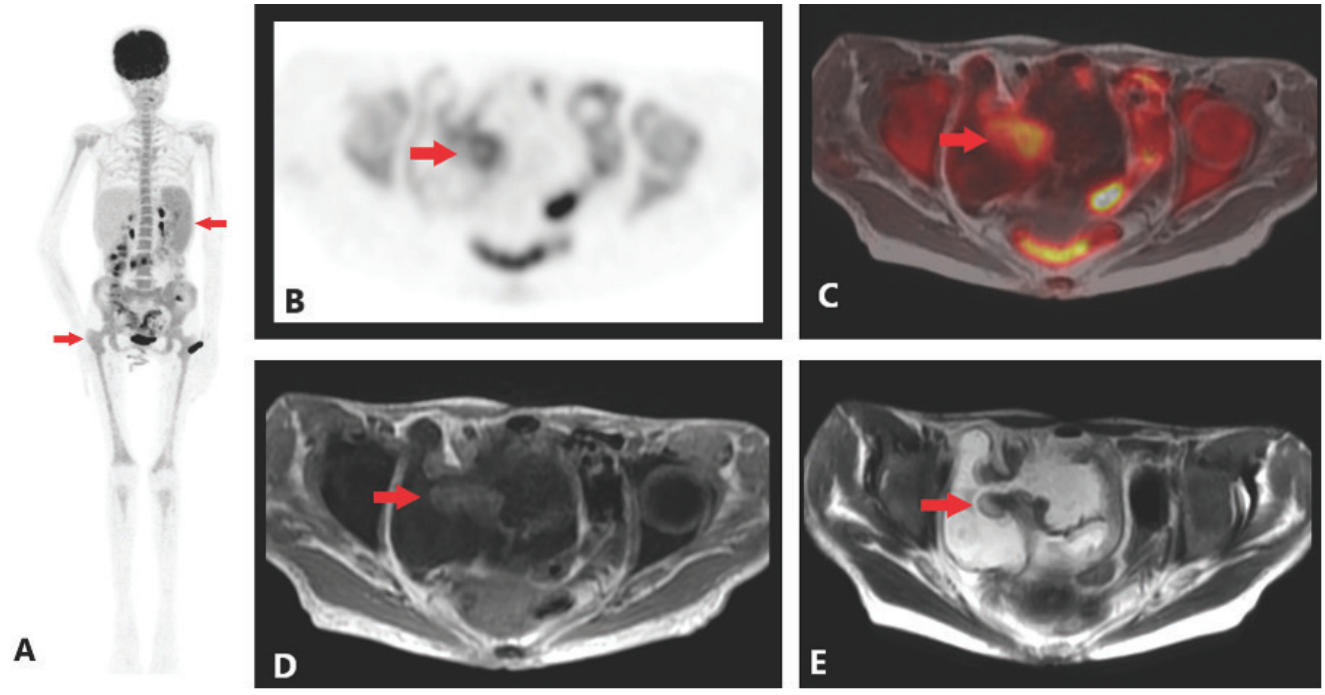

Figure 1. Behcet's syndrome (BS) is a variable vessel vasculitis with multi-organ involvement. The pathogenesis is unclear; however, genetic factors, as well as environmental factors, are thought to play a role. Human leukocyte antigen B51 allele in major histocompatibility complex locus is the best-known genetic risk factor for BS (1). It is a multi-system disease with recurrent episodes of oral and genital ulcers, papulopustular and nodular skin lesions, arthritis, uveitis, venous and arterial thromboses, arterial aneurysm, and nervous system involvement (1). According to different studies, gastrointestinal involvement occurs 4.5-6 years after the onset of oral ulcers. The frequency of gastrointestinal involvement was reported as high as 50% in the Far East, whereas approximately 1-1.5% in Turkey (2,3). The ileocolonic site is the most frequent disease localization in the gastrointestinal system (4). BS diagnosis is based on clinical manifestations, as there are no pathognomonic laboratory tests (1). However, endoscopic confirmation of typical ulcers is necessary for gastrointestinal involvement diagnosis in a patient with BS, since relying solely on clinical manifestations, such as abdominal pain and diarrhea, may be misleading. Inflammatory bowel diseases, especially Crohn's disease, and tuberculosis should be ruled out for the differential diagnosis of gastrointestinal involvement of BS (5). Medical treatment including 5-aminosalicylic acid, corticosteroids, immunosuppressive drugs, such as azathioprine, and monoclonal tumor necrosis factor-alpha inhibitors are commonly used, and surgery is generally reserved for patients who present with emergencies, such as major bleeding or perforation (6).

A 25-year-old female patient who was treated for BS, familial Mediterranean fever and myelodysplastic syndrome (MDS), and secondary acute myeloid leukemia (sAML) was referred to our center for [18]fluorine-fluorodeoxyglucose ([18]F-FDG) positron emission tomography (PET) imaging. She was diagnosed with gastrointestinal involvement of BS 3 years ago, after a colonoscopic examination for abdominal pain, weight loss, diarrhea, and mucus in the stool. Her colonoscopy revealed multiple colonic and ileocecal ulcers. Her abdominal pain, diarrhea, and elevated acute phase reactants continued despite treatment with colchicine, high dose prednisolone, thalidomide, and monoclonal TNF inhibitors. Abdominal imaging studies, which were performed due to fever, in addition to these findings, revealed a collection between the bladder and intestine. She was operated and a part of her ileum, as well as the bladder, was removed. Pathologic examination showed active chronic enteritis with multiple ulcers that extend to the subserosa in the small intestine. Additionally, lymphoplasmacytic cell infiltration, vascular proliferation, fresh thrombi, and medial calcifications in some vessels were observed in the submucosa, muscularis propria, and subserosa. The patient then developed pancytopenia and was diagnosed as MDS with positive monosomy 7 and negative trisomy 8. During the follow-up, she progressed to sAML. The association of MDS and AML with gastrointestinal involvement is an interesting and well-documented entity (7,8). [18]F-FDG PET/magnetic resonance imaging (MRI) was performed. [18]F-FDG PET maximum intensity projection (A) image revealed diffuse hypermetabolism on the bone marrow and hypermetabolism associated with splenomegaly, which was related to AML-M4. Axial PET (B), fused PET/T1-weighted (T1W) MRI (C), T1W MRI (D), and T2-weighted MRI (E) images revealed segmental dilatation and hypermetabolic wall thickening around the ileocecal valve (maximum standard uptake value: 6.23), which followed gastrointestinal involvement of BS. To our knowledge, this is the first case of intestinal BS with [18]F-FDG PET/MRI.

Ethics

Informed Consent: Was obtained from the patient.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Concept: M.S.S., R.L.U.B., S.A., Design: M.S.S., R.L.U.B., K.S., Data Collection or Processing: B.İ., A.K., S.A., Literature Search: B.İ., A.K., S.A., Writing: B.İ., A.K., R.L.U.B., A.E.E., A.İ.H., G.H.

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