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A Rare Manifestation of Behçet's Disease; Arterial Involvement

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Abstract:

Behcet's Disease, also known as Behcet's syndrome, is a rare, chronic, autoimmune, auto inflammatory disorder of unknown origin. Its manifestations are thought to be caused by vasculitis resulting in damage to blood vessels throughout the body. The disease is named for the Turkish dermatologist, Dr. Hulusi Behcet, who in 1937 described a triad of oral ulcers, genital ulcers and ocular inflammation. In this case, we describe a case of acute myocardial infarction secondary to coronary artery involvement of Behçet's disease in a patient who was diagnosed with Behçet's disease 15 years ago from aphthous lesions in his mouth and genital area.

Keywords: ***Behçet's disease, myocardial infarction.***

Introduction:

Behçet's disease was first defined in 1937 by a Turkish doctor Hulusi Behçet. It is one of the rare diseases defined by a Turkish doctor in the medical history. The most typical characteristic of Behçet's disease is lession called aphthae recurring in oropharynx and genital regions. The status accompanied by both aphthae and uveitis is defined as Tri Symptom Behçet.

Behcet's disease is defined as a systemic vasculitis that can affect both arteries and venules, and can cause lesions in every affected vessel. Although Behçet's disease has a low rate of cardiac involvement, it increases mortality. Acute myocardial infarction may develop due to coronary artery involvement but is rare. In this case, we describe a case of acute myocardial infarction secondary to coronary artery involvement of Behçet's disease in a patient who was diagnosed with Behçet's disease 15 years ago from aphthous lesions in his mouth and genital area.

Case:

A 55-year-old male patient with a history of aphthous lesions in his mouth and genital area 15 years ago who had a diagnosis of Behçet's disease and had a 0.5 mg 1x1 colchicine treatment and a history of peptic ulcer-associated partial gastrectomy was admitted to emergency clinic with complaint of melenea. Patient's blood pressure was 100/60 mmHg, pulse was 108 / min, hemoglobin was 10 mg / dL. Patient had a history of coronary angiography with normal results due to chest pain 6 years ago. The patient was admitted with a diagnosis of upper gastrointestinal hemorrhage due to positive presentation of the rectal examination. When control hemoglobin values had decreased and melenea continued, the patient underwent transfusion of erythrocyte suspension to correct hemodynamics. Biliary reflux was detected in the upper gastrointestinal endoscopy of the patient who was clinically and hemodynamically stable. Colonoscopy was planned for patient to investigate the gastrointestinal involvement of Behcet's disease. However, acute myocardial infarction findings were detected in the ECG of the patient who had chest pain at night before colonoscopy. Patient's ECG had ST elevations in leads V1-V2-V3-V4, DI, aVL. Coronary angiography was performed to patient with laboratory values of CK: 2108, troponin T: 6.76, CK / MB: 254.2, Hgb: 10.4 mg / dl. In coronary angiography; LMCA was normal; total occlusion before LAD D1, intermediate artery plate, Cx was normal; 70-90% stenosis was detected in the RCA mid region (Figure1,2). Thrombus aspiration was performed from thrombosed lesion in LAD during the angiography, followed by a stent implantation into the lesion (Figure 3). Elective intervention was decided for RCA. In the pre-discharge echocardiography of the patient; ejection fraction (EF) was 30-35%; basal and midanteroseptal region was severe hypokinetic; apicoanterior, apicoseptum and apex were akinetic. Furosemide treatment was continued due to cardiac insufficiency in addition to amiodarone therapy which was started shortly after the onset of atrial fibrillation in cardiology clinic with concurrent therapy of acetylsalicylic acid, clopidogrel, trandolapril, spironolactone, rosuvastatin treatment. Ivabradine treatment was added with the cause of sinus rhythm and tachycardia during discharge.

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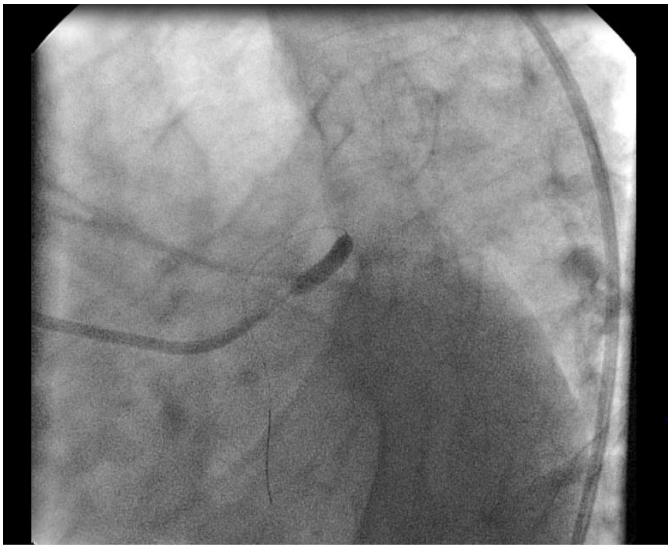

Figure 1: LEFT CAUDAL VIEW OF LAD PTCA APPLICATION

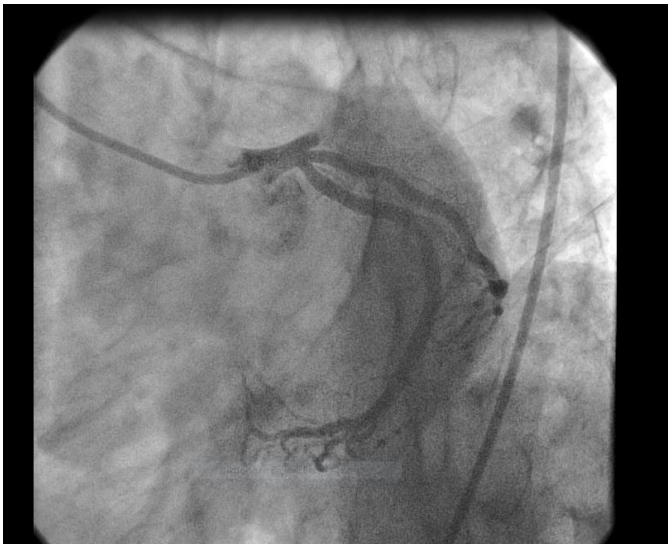

Figure 2: LEFT CAUDAL VIEW OF LAD TOTAL LESION

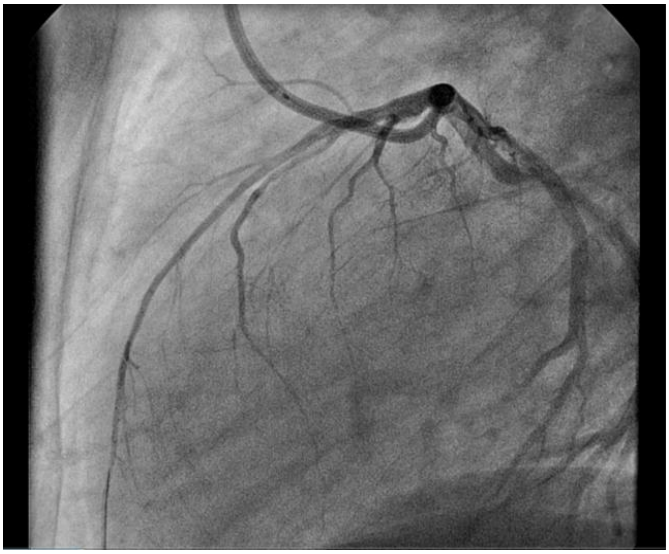

Figure 3: LEFT LATERAL VIEW AFTER STENT IMPLANTATION

Discussion:

Behçet's disease is a chronic disease with attacks. It is a multisystemic vasculitis with oral aphthae, genital ulcers, cutaneous, ocular, gastrointestinal, articular, pulmonary, vascular, neurological symptoms and findings.[1,2] Dr. Hulusi Behçet described this disease in 1937 as three symptomatic diseases (Tri Symptom Behçet) with oral and genital ulcers together with hypopyon uveitis.[3]

Although the etiopathogenesis of Behçet's disease is not fully known, the most hypothesized hypothesis is an uncontrolled immune response triggered by autoantigens such as antigen and / or heat shock proteins of viral, bacterial, etc. in people susceptible to genetically disorders.[4]

Epidemiological studies on Behçet's disease which is most commonly seen in Turkey are few.[5.6] It is seen mainly in Mediterranean countries, Central Asia, Far East countries. Because of its geographical distribution, it is also called "Silk Road Disease".[7] In this distribution, it is suggested that HLA-B51 expression and environmental factors may also be effective from genetic factors.[8]

The average onset age of the disease is around 28 years. The average onset age was 23.3 in Turkish patients, 26 in Germany and 35.7 in Japan. It is rare in children and after 50 years, but may start at any age.[9]

The results regarding the distribution of men and women vary. In addition to publications reporting that male cases are more frequent in the Mediterranean countries, there are also publications reporting that there is no difference between male and female rates. Pustular lesions, ocular findings, vascular lesions are more severe in male patients whereas genital aphthae and eritema nodosum are more frequent in female patients. In male patients, systemic evidence of early onset, HLA B51 positivity is reported as a poor prognostic indicator.[10]

Behcet's disease affects the gastrointestinal system by 10-50%. Gastrointestinal system involvement; 50-60% in Japanese patients, 38-50% in English, 1/3 'in Taiwanese patients and 1/4 of Spanish patients; In Lebanon, Saudi Arabia and Turkey, gastrointestinal involvement is rarely seen (0-5%).[11]

In the gastrointestinal system, at the small veins of the intestinal wall, and often at the venules, it manifests itself as an ulcer due to the formation of vasculitis. Mucosal ulcers are most common in the ileocecal region, although they can hold any region from the mouth to the anus. The ulcers may cause spontaneous perforations, as well as abdominal pain, diarrhea, retrosternal burning secondary to esophagitis, perianal fistula. The presence of ulcers is an indication of

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intestinal involvement.[7,8] Colonoscopic examinations can easily identify the ulcers located in the colonic region. However, as perforation can easily develop during colonoscopy, excessive air flow should be avoided. Because of easy development of perforation; if a biopsy is to be taken, biopsy is recommended after air is aspirated from the colon.[12]

For Behcet's disease; venous involvement is more common, although systemic vasculitis affects both arteries and venules. Patients are more likely to be presented with superficial thrombophlebitis, deep vein thrombosis.[13] Vena cava superior, vena cava inferior, dural sinus and Budd-Chiari syndrome can also be seen.[14]

Arterial involvement is less than venous involvement but causes more morbidity. Mostly pulmonary artery, femoral artery, popliteal, subclavian and carotis arteries are affected. It causes aneurysm, arterial occlusion in arterias.[15,16]

Pulmonary artery aneurysm due to Behcet's disease causes hemoptysis most commonly.[17] Pulmonary artery aneurysm is an important factor affecting mortality.[14]

Cardiac involvement is rare. However, pericarditis, coronary venous involvement, intracardiac thrombosis, endomyocardial fibrosis, and coronary artery involvement are rarely reported.[18]

Myocardial infarction can develop as secondary to vasculitis and coronary angiography of these patients can be detected normally.[19]

In our case; patient was admitted because of melenea and the upper gastrointestinal system bleeding was considered firstly for the patient. Upper gastrointestinal bleeding was initially thought to be due to an anterior peptic ulcer in the patient, and not considered due to Behcet's disease. However, no peptic ulcer was detected in the upper gastrointestinal endoscopy of the patient. We can exemplify our case as an example of ulcers in Behçet's disease that can be seen in the entire gastrointestinal tract.

In patient's follow up, chest pain and myocardial infarction developed. Coroner angiography of the patient which was done 6 years ago was normal. This can be seen in Behçet's disease eventhough it's rare.

We have also seen in our case that it causes more serious morbidity in arterial involvement, especially in male gender. Despite the early intervention of the patient, the ejection fraction had decreased to 30-35% .In Behcet's disease, coronary artery involvement is rare, but it shows the serious morbidity.

References:

[1] Tsuyoshi S, Mitsuhiro T, Noboru S et al: Behcet disease. N Engl J Med, 1999; 341(17):1284-1291.
[2] Yazıcı H: Behcet's Disease. Klippel *JH*. Eds Rheumatology, *2nd* Ed. London: Mosby. 1998; 26: 1-6.
[3] Dilşen N: Behçet Hastalığı tarihçesi. Aktüel Tıp Dergisi 1997; 2: 62-65.
[4] Akman A, Alpsoy E: Behçet Hastalığı: Etyopatogenezde güncel bilgiler. Türkderm 2009; 43 Özel Sayı 2:32-8.
[5] Doğanavşargil E, Keser G: Behçet Hastalığı. Klinik Romatoloji (Derleyenler) Gümüşdiş G, Doğanavşargil E, Ege Üniveristesi Yayınları, Deniz matbaası, 1999;423-439.
[6] Tüzün Y, Yurdakul S, Mat C et al: Epidemiology of Behcet's syndrome in Turkey. Int J Dermatol 1996; 35:618-20.
[7] James DG: Silk route disease. Postgrad Med J 1986; 62:151-3.
[8] Verity DH, Marr JE, Ohno S et al: Behcet's disease, the Silk Road and HLAB51: historical and geographical perspectives. Tissue Antigens 1999; 54:213-220.
[9] Altenburg A, Papoutsis N, Orawa H et al: Epidemiology and Clinical Manifestations of Adamantiades-Behcet disease in Germany current pathogenetic concepts and therapeutic possibilities J DtschDermatolGes2006; 4:49-64.
[10] Saylan T, Marc, Fresko İ, Melikoğlu M: Behcet's Disease In The Middle East Clinic in Dermatology 1999; 17: 209-222.
[11] Yurdakul S, Tüzüner N, Yurdakul I et al: Gastrointestinal involvement in Behcet's syndrome: a controlled study. Ann Rheum Dis 1996; 55:208-10.
[12] Örmeci N: Behçet Hastalığında Gastrointestinal Sistem. Ankara Üniversitesi Tıp Fakültesi Mecmuası 2011, 64(Ek 3): 59-62.
[13] Kuzu MA, Ozaslan C, Koksoy C, Gurler A &Tuzuner A. Vascular involvement in Behcet's disease: 8-year audit. World Journal of Surgery1994; 18:948 –953. Discussion 953– 954.
[14] Hamuryudan V, Yurdakul S, Moral F, et al. Pulmonary Arterial Aneurysms in Behcet's syndrome: a report of 24 cases. British Journal of Rheumatology 1994; 33:48–51.
[15] Sakane T, Takeno M, Suzuki N &Inaba G. Behcet's disease. New England Journal of Medicine 1999; 341:284–1291.
[16] Kural-Seyahi E, Fresko I, Seyahi N, et al. The long-termmortality and morbidity of Behcetsyndrome: a 2-decade outcome survey of

DOI: 10.23958/ijirms/vol02-i07/13

387 patients followed at a dedicated center. Medicine (Baltimore) 2003; 82.

[17] Mader R, Ziv M, Adawi M &Lavi I. Thrombophilic factor sand their relation to thromboembolic and other clinical manifestations in Behcet's disease. Journal of Rheumatology 1999; 26:2404–2408.

[18] Espinosa G, Font J, Tassies D, et al. Vascular involvement in Behcet's disease: relation with thrombophilic factors, coagulation activation, and thrombomodulin. American Journal of Medicine 2002; 112:37–43.

[19] Calamia K, Schimer M, Melikoğlu M: Major Vessel Involvement in Behcet disease. Current Opinion in Rheuma 2005; 17:1-8.

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