Olgu Sunumu / *Case Report*

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BUERGER'S DISEASE AFFECTING THE VISCERAL VASCULATURE – A VERY RARE CLINICAL ENTITY

VİSSERAL DAMARLARI ETKİLEYEN BUERGER HASTALIĞI - NADİR BİR KLİNİK OLGU

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ÖZ

Giriş:*Buerger hastalığı veya Thromboangiitis obliterans, enflamatuar bir vazo-okluzif hastalıktır. Primer olarak küçük ve orta büyüklükteki arterler ile alt ve üst ekstremite venlerini etkiler. Visseral damarların tutulumu nadiren bildirilmiştir.*

Olgu: *33 yaşında erkek hasta kliniğimize karın ağrısı ile başvurdu. Buerger hastalığı tanısı mevcut olan hastanın abdominal aort ve visseral damarlarına yönelik yapılan anjiyografide, superior mezenterik arterde oklüzyon, ince bağırsak terminal dallarında ani oklüzyon ve tirbüşon görünümünde kollateraller tespit edildi. Medikal tedavi uygulanan hastanın semptomları üçüncü ay kontrolünde tamamen kayboldu ve rutin poliklinik kontrolleri ile takibi planlandı.*

Sonuç: *Uyguladığımız medikal tedavi protokolü ile Buerger hastalığının nadir görülen bu formunun tedavisi başarılı bir şekilde gerçekleştirilmiştir. Buerger hastalığının bu zorlu alt grubunu daha iyi anlamak ve tedavisini sağlamak için çok merkezli yüksek hacimli kohort çalışmalarına gereksinim vardır.*

SUMMARY

Introduction:*Buerger's disease or Thromboangiitis obliterans is an inflammatory vaso-oclussive disease.It affects primarily small to medium-sized arteries and veins of both lower and upper extremities.Involvement of the visceral vessels are rarely documented.*

Case: *A 33-year-old male patient was presented to our institution with abdominal pain. He was already diagnosed with Buerger's disease.An angiogram of the abdominal aorta and visceral vessels showed superior mesenteric artery occlusion, abrupt occlusion of small bowel's terminal branches and collaterals with corkscrew appearance . After medical treatment, his symptoms completely disappeared at the third month control and he is scheduled for routine outpatient clinic follow up.*

Result: *We succeeded on management of very rare form of Buerger's disease with our medical management protocol. Further multicenter high volume cohort studies are warranted in order to better understand and provide treatment for this challenging subgroup of Buerger's disease patients.*

INTRODUCTION

Buerger's disease or Thromboangiitis obliterans (TAO) is an inflammatory vaso-oclussive disease that affects primarily small to medium-sized arteries and veins of both lower and upper extremities.It is generally characterized by peripheric arterial occlusions (1).

Angiography is gold standard for the diagnosis. Diagnosis of the disorder is based on Shionoya's clinical diagnostic criteria,which include;a history of smoking, onset before the age of 50, the presence of infrapopliteal arterial occlusions, either upper limb involvement or phlebitis migrans, and the absence of atherosclerotic risk factors other than smoking (2, 3).

Involvement of the visceral vessels in the course of Buerger's disease is rarely documented. In this paper, we present the case of 33-year-old patient with intestinal Buerger's disease.

CASE

A 33-year-old male patient was presented to our institution with abdominal pain which started 2 weeks ago. Physical examination revealed bilateral absent lower extremity pulses below the knee level. The patient had a history of smoking two packs of cigarettes a day for 20 years. Typical corkscrew appearance of bilateral posterior tibial arteries occlusion was present inthe angiography which was performed 1 year ago, and he was already diagnosed with Buerger's disease.

The clinical evaluation revealed no acute pathology confined to the abdomen. Further examination was carried out with contrasted computerized tomography and it showed occlusion of the superior mesenteric artery with increased collateral vessels and thickening of intestinal mucosa at different segments. An angiogram of the abdominal aorta and visceral vesselsshowed superior mesenteric artery occlusion, abrupt occlusion of small bowel's terminal branches and collaterals with corkscrew appearance (Figure 1-3). Depending on the previous diagnosis and the typical appearance of collateral vessels, the patient was considered as visceral involvement of the Buerger's disease.

He had large collaterals and unfavourable run off arterial anatomy; hence surgical reconstruction was not planned and he was considered for medical treatment with 2 periods of iloprost infusion.He stopped smoking, the symptoms relieved with iloprost infusion and he was discharged from hospital with significantly reduced symptoms with cilostazol (100mg twice daily), clopidogrel (75mg once daily) and acetylsalicylic acid (100mg once daily) 15 days after the treatment. His symptoms completely disappeared at the third month control. In the follow up period, the patient was treated especially about smoking cessation and did not start again smoking. The recurrent symptoms did not ocur in the one year follow up and he is scheduled for routine outpatient clinic follow up with medical treatment.

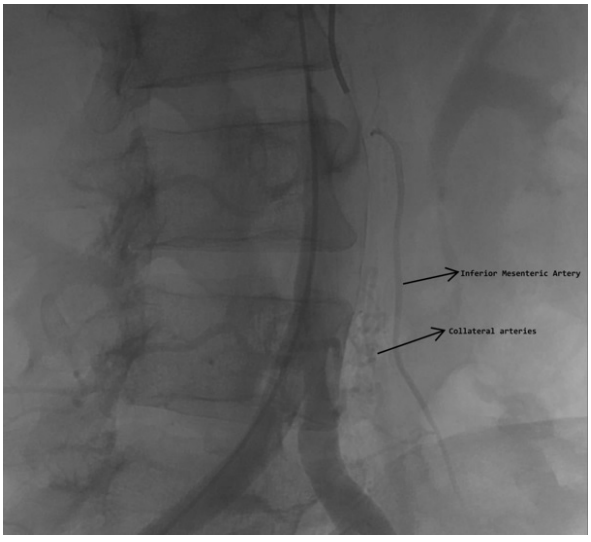

Figure 1. Digital substraction angiography showing inferior mesenteric artery and collaterals.

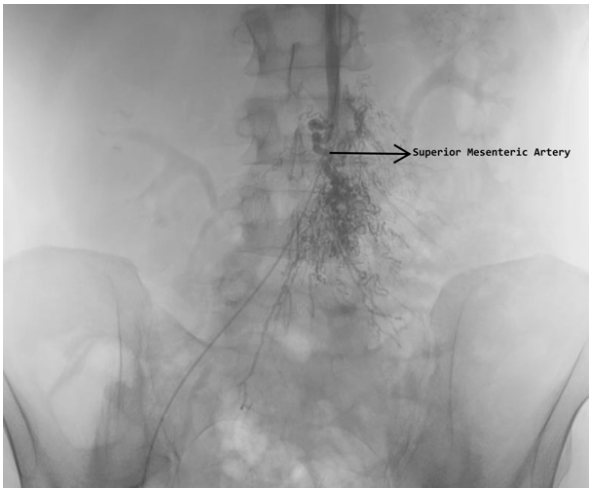

Figure 2. Digital substraction angiography showing superior mesenteric artery and collaterals.

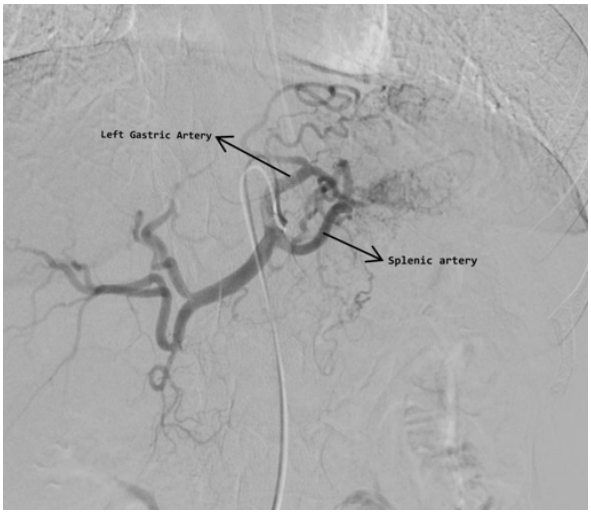

Figure 3. Digital substraction angiography showing splenic, left gastric arteries and collaterals.

DISCUSSION

Although the involvement of intestinal vessels in Buerger's disease is rare, it may be asymptomatic or symptomatic. Symptomatic cases can either be acute or chronic.Acute cases are generally treated with bowel resection while chronic cases are mostly treated with bypass surgery (4). According to Takehisa Iwa's study, there are only five rare cases of chronic Buerger's disease whom underwent arterial reconstruction surgery with successful short term outcome (4).

Visceral arteries involvement may be revealed by angiography or aortography and looks like the arteries of the extremities. Because of it is overlooked since far, some cases with vascular involvement of multiple organs has been noted in autopsies of patients with Buerger disease (5). Although in the description of Buerger disease, involvement of the small- and medium-sized arteries of the extremities is only described as diagnostic criteria; Leo Buerger was firstly detected the visceral involvement in thromboangiitis obliterans. But this presentation of TAO was known as unusual or progressive form of TAO (5).

There is a low chance of improving the condition with surgery,especially in patients with severe disease. Therefore pharmacological agents are used such as cilostazol, clopidogrel, and pentoxifylline, or medicine derivatives of prostacyclin and prostaglandin. They redirect blood flow and improve the circulation mostly in affected areas and relieve rest pain (3, 6). There are some studies suggest that smoking cessation and systemic medical treatment also lead to the healing of the patients with visceral Buerger and reduce the mortality rate (5). In our case, we treated the patient with iloprost, which is a stable prostacyclin analog, that has been studied for the treatment of thromboangiitis obliterans related ischemia.Iloprost can be given by inhalation, oral administration or intravenous infusion (6). We administrated iloprost by intravenous infusion.

In our case, the patient was already diagnosed with Buerger's disease. He had all criteria of the pathology such as; a history of smoking, symptoms before the age of 50, the presence of infrapopliteal arterial occlusions, phlebitis migrans and did not have any atherosclerotic risk factors other than smoking. The angiography revealed the typical collaterals with corkscrew appearance as it seems in the arteries of the extremities. Hence, he was considered as visceral involvement of TAO. The patient was not considered for surgical treatmentdue to the large collaterals surrounding affected vessels and unfavourable run off arterial anatomy and we preffered medical management.Patient's symptoms relieved with ilomedin infusion and completely disappeared with oral cilostazol, clopidogrel and aspirin combination and especially when he stopped smoking.

In conclusion, in this report we present a very rare form of Buerger's disease, visceral Buerger together with our succesful medical management protocol. Further multicenter high volume cohort studies are warranted in order to better understand and provide treatment for this challenging subgroup of Buerger's disease patients.

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