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Case Report / Olgu Sunumu

Arterial and venous thrombosis due to primary thrombocytosis after splenectomy

Splenektomi sonrası primer trombositoz nedeniyle gelişen arteriyel ve venöz tromboz

Okay Güven Karaca,[1] Mehmet Taşar,[2] Mehmet Kalender,[1] Ata Niyazi Ecevit,[1] Mine Tavlı Yılmaz[3]

Institution where the research was done:
Department of Cardiovascular Surgery, Konya Training and Research Hospital, Konya, Turkey

Author Affiliations:
[1]Department of Cardiovascular Surgery, Konya Training and Research Hospital, Konya, Turkey
[2]Department of Cardiovascular Surgery, Medical Faculty of Ankara University, Ankara, Turkey
[3]Department of Cardiovascular Surgery, Tire State Hospital, İzmir, Turkey

Patients with myeloproliferative disorders such as primary thrombocytosis are prone to vascular thrombotic events. Splenectomy is also a risk factor for thrombocytosis. Such significant thrombosis may cause serious extremity or cardiac problems. It is beneficial to use prophylactic platelet anti-aggregating medication such as aspirin, cytoreductive agents such as hydroxyurea or anticoagulation with close monitoring in these patients that have additional risk factors. In this article, we present a case for whom medical therapy was more important and life-saving than surgery for peripheral vascular thrombosis after splenectomy operation.

Keywords: Myeloproliferative disorder; peripheral arterial disease; splenectomy.

Primer trombositoz gibi miyeloproliferatif hastalığı olan kişiler vasküler trombotik olaylara yatkınlık gösterirler. Splenektomi de trombositoz gelişimi açısından bir risk faktörüdür. Bu tür anlamlı tromboz, ciddi ekstremite ve kalp sorunlarına yol açabilmektedir. Ek risk faktörüne sahip bu hastalarda aspirin gibi profilaktik anti-agreganlar, hidroksiüre gibi sitoredüktif ajanlar veya yakın monitörize edilmesi gereken antikoagülan tedavi rejimlerinin kullanımı faydalı olmaktadır. Bu yazıda, splenektomi ameliyatı sonrası periferik vasküler trombozun tedavisi için medikal tedavinin cerrahi tedaviden çok daha önemli ve hayat kurtarıcı olduğu bir olgu sunuldu.

Anahtar sözcükler: Miyeloproliferatif hastalık; periferik arter hastalığı; splenektomi.

The normal platelet count is 150-450 K/μL, and thrombocytosis is diagnosed when the platelet count is higher than 500 K/μL.[1] When myeloproliferative disorders occur or after a splenectomy, the platelet count can be extremely high, and when thrombocytosis is present, it can lead to a stroke, myocardial infarction, extremity ischemia, venous thrombosis, or embolic events that can be catastrophic.

Portal vein thrombosis (PVT) is a serious problem related to thrombocytosis, and Doppler ultrasonography (USG) performed before or after the splenectomy may be beneficial for detecting this condition so that treatment with prophylactic agents can begin.[2]

Myeloproliferative disorders have to be detected before splenectomies because they both can lead to serious cardiac or other thrombotic complications. Hence, it is important to monitor the platelet counts closely, and if necessary, anti-aggregating agents and anticoagulation medications can be utilized for both prophylaxis and treatment.

Surgery is also a risk factor for these patients. After arterial or venous complications, an early

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Correspondence: Mehmet Taşar, M.D. Konya Eğitim ve Araştırma Hastanesi, Kalp ve Damar Cerrahisi Kliniği, 42090 Konya,, Turkey.
Tel: +90 505 - 442 47 80 e-mail: mehmet.tasar@hotmail.com

vascular evaluation is essential, and if an intervention is required, it should be done immediately to rescue the extremities or organs. The success of the surgery is related to the platelet count at that time, so using the proper prophylaxis and medications becomes even more critical for these patients.

CASE REPORT

A 76-year-old woman with a previous history of primary thrombocytosis had undergone a splenectomy for subscapular hematoma and rupture after a traumatic event at another hospital, and she came to our facility after the operation. On the postoperative second day, the patient began to complain of bilateral lower extremity pain, but she was taking daily doses of acetylsalicylic acid (aspirin) for the primary thrombocytosis, and there was no vascular event in her medical history. The patient's extremities were pale, and both lower extremities were cold and cyanotic. In addition, the bilateral femoral artery pulses were palpable, but the popliteal and distal artery pulses were absent. Lower extremity arterial Doppler USG then revealed a bilateral superficial femoral artery occlusion proximal to the popliteal artery, and lower extremity arterial magnetic resonance angiography (MRA) also identified a bilateral superficial femoral artery occlusion (Figure 1). All of the laboratory tests were normal except for the platelet count (1,200 K/μL). We performed an emergency bilateral femoral embolectomy until the intraoperative popliteal pulses were palpable. Unfortunately, during the postoperative first hour, the popliteal distal pulses were lost, and the patient then underwent a popliteal artery embolectomy. However, both the popliteal and distal pulses were absent postoperatively. During her intensive care unit (ICU) stay, the patient received an intravenous systemic heparin infusion (15 U/kg/hour), but the distal perfusion could not be restored. On the postoperative second day, the patient's liver enzymes increased [alanine transaminase (ALT): 875 units per liter (U/L) and aspartate transaminase (AST): 1256 U/L), and renal failure occurred (urea: 174 mg/dl; creatinine: 3.45 mg/dl), so a bilateral above-the-knee amputation was performed. At the 18th hour after the amputation, the patient had abdominal tenderness, and Doppler USG detected PVT. The classic heparin treatment was replaced by low-molecular-weight heparin (LMWH), and on the postoperative 10th day, the patient's liver and renal functions were restored. She was then discharged on the postoperative 14th day.

DISCUSSION

Trombocytosis may occur due to myeloproliferative disorders or splenectomies, and it is a well known risk factor for vascular events.[3,4] Therefore, it is essential to detect and treat it before complications occur. Prophylactic and therapeutic agents are well defined for this group of patients.[5,6] Most often, when vascular complications occur, vascular surgical or interventional procedures are inevitable, and, unfortunately, they most often have poor outcomes. This is probably caused by the deterioration of the vascular structures due to asymptomatic recurrent thrombosis. In our patient, vascular surgery was attempted, but we ultimately had to perform a bilateral leg amputation.

Portal vein thrombosis is a rare but significant complication associated with splenectomies performed for hematological diseases, with myeloproliferative diseases being the most common hematological disorder that causes this condition. These are followed by hemolytic anemia, hereditary spherocytosis, and thalassemia major and idiopathic thrombocytopenic purpurae. Prolonged unexpected febrile and abdominal tenderness in patients who have undergone

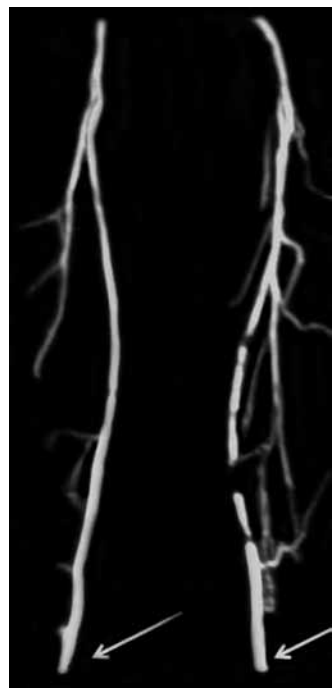

Figure 1. Preoperative magnetic resonance angiogram image of the lower extremity arterial system showing bilateral superficial femoral artery thrombosis.

a splenectomy must be promptly evaluated for the development of PVT, and Doppler USG may then be performed to diagnose this condition. Furthermore, prophylactic antiplatelet and antithrombotic therapy should be considered after splenectomies to help prevent PVT.[2]

Asplenia also has been known to predispose patients to infection, and infectious complications associated with the postsplenectomy state are well documented.[7] Thus, a discussion should be take place with the patient regarding special perioperative management. Our patient had a vascular operation and bilateral amputation. We used prophylactic antibiotherapy for all of her operations, and she did not experience any infectious complications.

The risk to the patient from the increased platelet count should be assessed first. Patients with thrombocytosis who have had thrombotic events and possess cardiovascular risk factors should be treated with hydroxyurea, interferon alpha, ticlopidine, enoxaparin, and anagrelide, a newer platelet-lowering agent that has been approved for patients with essential thrombocytosis[5,6] either before or after vascular surgical procedures performed for thrombocytosis. The risk of bleeding associated with the use of aspirin should be kept in mind for patients with thrombocytosis, and patients taking hydroxyurea should be monitored for leukemic transformation. Another treatment option for rapidly reducing the platelet count in life-saving clinical situations is plasmapheresis, which involves the removal, treatment, and return of plasma from the circulating blood.

Partial splenectomies and splenic autotransplantation techniques may also protect against complications, but whether or not they do this job as well as a total splenectomy remains controversial. Currently, prophylaxis remains the treatment of choice to prevent complications after a splenectomy.

Thrombocytosis is a serious risk factor associated with vascular and cardiac events after a splenectomy performed for myeloproliferative disorders. Therefore, either before or after surgical interventions, prophylactic medical therapy with anti-aggregating and anticoagulating agents is essential for these patients.

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