Paget-von Schrötter syndrome: upper extremity deep vein thrombosis after heavy exercise

Paget-von Schröetter sendromu: Ağır egzersiz sonrası gelişen üst ekstremite derin ven trombozu

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Summary– We present a 52-year-old man with swelling, tightness, heaviness, and redness in the right half of his neck, face, upper arm, and right precordial area after repetitive exercises. No etiological factors such as trauma, venous catheter, malignancy, hematologic disorder history, or any chronic disease were noted. A Doppler ultrasound of the upper extremities was performed, which showed a complete thrombotic occlusion in the right internal jugular vein, partial occlusion in the right brachiocephalic and subclavian veins with thrombosis, and chronic thrombosis occluding the left brachial vein. Complete blood cell count, coagulation profiles, and rheumatologic and tumor markers were normal. The patient reported that he had taken spinning classes for the last 18 months, and for the past 4 days he reported that he had done his spinning exercises harder and more frequently. The swelling and redness regressed partially on the 3rd day of therapy with enoxaparin sodium. In the first month follow-up visit we performed a control Doppler ultrasound and it showed partially thrombosed areas in the right brachiocephalic and internal jugular veins. Also, chronic thrombosis persisted in the left brachial and basilic veins. Computed tomography images showed no evidence for malignancy and some sections showed filling defects in the vena cava superior, which was attributed to thrombotic materials. The patient was successfully treated with anticoagulants for 6 months.

Özet– Bu yazıda, tekrarlayan egzersiz sonrası boynunun, yüzünün sağ yarısında, sağ üst ekstremitede ve sağ prekordiyal bölgede gerginlik, şişkinlik, ağırlık ve kızarıklık olan 52 yaşındaki erkek hasta sunuldu. Travma, venöz kateter ve malign hematolojik veya kronik hastalık öyküsü gibi etyolojik faktörler tanımlanmadı. Üst ekstremitede Doppler ultrasonografisi uygulandı, sağ internal juguler vende tam trombüse bağlı tıkanıklık, sağ brakiyosefalik ve sağ subklaviyan vende kısmi tıkanıklık ve ayrıca sol brakiyal vende kronik trombüs saptandı. Tam kan sayımı, koagülasyon profili, romatolojik ve tümör belirteçleri normaldi. Hastanın son 18 ayda kondisyon bisikleti ile egzersizler yaptığı, son dört günde çok sık ve ağırlaştırılmış şekilde ağır egzersizler yaptığı öğrenildi. Enoksaparin sodyum tedavisinin üçüncü gününde şişlik ve kızarıklıklar kısmi olarak geriledi. Birinci ayda yapılan kontrol Doppler ultrasonografisinde sağ brakiyosefalik ve internal juguler vende kısmi trombüsle kaplı alanlar gözlendi. Ayrıca, sol brakiyal ve bazilik vende kronik trombüsler gözlendi. Bilgisayarlı tomografi görüntülerinde malign hastalık ile uyumlu bulgu gözlenmedi, vena kava süperiyorda trombüse ait dolum defektleri gözlendi. Hasta 6 ay boyunca başarılı bir şekilde antikoagülanlarla tedavi edildi.

Upper-extremity deep vein thrombosis (UEDVT) is an increasingly important clinical entity with potential for considerable morbidity. UEDVT most commonly refers to thrombosis of the axillary and/or subclavian veins and it is classified as either primary or secondary on the basis of pathogenesis.[1] Primary UEDVT is a rare disorder that refers either to effort thrombosis or idiopathic

Abbreviations:

CT	*Computed tomography*
PSS	*Paget-von Schröetter syndrome*
UEDVT	*Upper-extremity deep vein thrombosis*

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UEDVT.[2] Effort thrombosis usually occurs after exercises or sports with repetitive movements, such as wrestling, playing ball, gymnastics, or swimming. Secondary UEDVT often develops in patients with central venous catheters, pacemakers, thrombophilic states, or cancer, and accounts for the majority of cases of UEDVT.[3] Paget-von Schröetter syndrome (PSS) is a subtype of thoracic outlet syndrome that is neither positional nor secondary to indwelling catheters, but rather is related to effort thrombosis. This syndrome was described in 1875 by Paget and in 1884 by von Schröetter and is also known as effort thrombosis.[2]

In this case report, we present a case of PSS caused by repetitive exercises, which was referred to our department.

CASE REPORT

A 52-year-old male was admitted with swelling, tightness, heaviness, and redness in the right half of his neck, face, upper arm, and right precordial area (Fig. 1). On clinical questioning, no etiological factors like trauma, venous catheter, malignancy, or hematologic disorder history were noted. The patient did not report any chronic diseases, such as hematological disorders, except for hypertension and hyperlipidemia. The patient's family history was negative for coagulopathies, venous thrombosis, and pulmonary embolism. In addition to his description, physical examination revealed small collateral vessels on his right subclavicular area. There were no signs of jugular vein distension or dilated peripheral veins around his right arm. Additionally, neither his subclavicular nor supraclavicular fossas were distinguished clearly because of the swelling, nor was tenderness was noted over the supraclavicular fossa. His Wright, Adson, and Halsted tests were all negative. His extremities were compared, and there was no directly observed or relative atrophy of any group of muscles. Tinel and Phalen signs were absent, and his chest radiogram showed no clinical pathology. A Doppler ultrasound of the upper extremities showed complete thrombotic occlusion in the right internal jugular vein, partial occlusion in the right brachiocephalic and subclavian veins with thrombosis, and chronic thrombosis occluding the left brachial vein (Fig. 2).

Complete blood cell count and echocardiographic tests were all within the normal ranges. The coagulation profiles including protein C, protein S, homocysteine, and antithrombin III were also within normal ranges. Heterozygote Metilen Tetrahydrofolate Reductase (MTHFR) Enzyme Mutation and Heterozygote F XIII V34L mutation which are not prothrombotic markers were detected. Folic acid and homocysteine ranges were normal. Rheumatologic

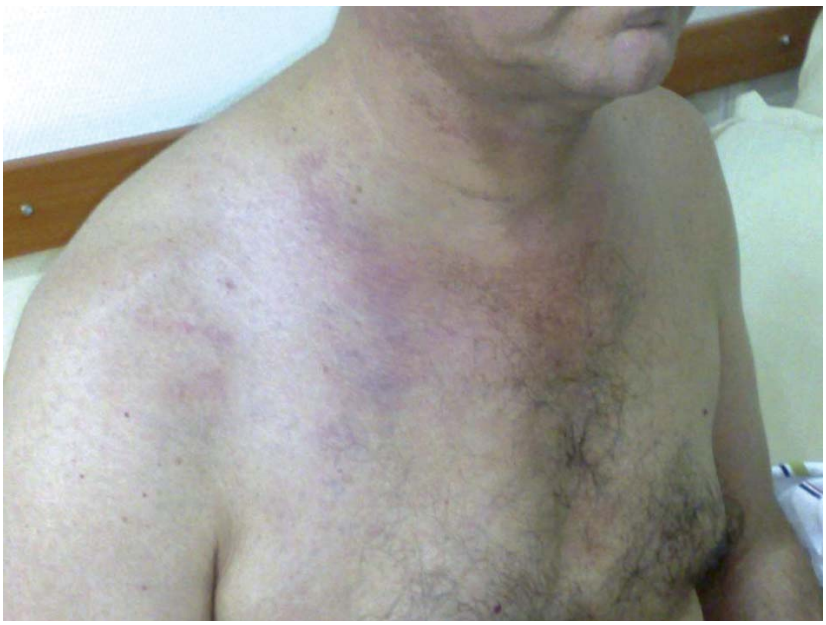

Figure 1. Redness in the right half of the patient's neck, face, upper arm, and right precordial area.

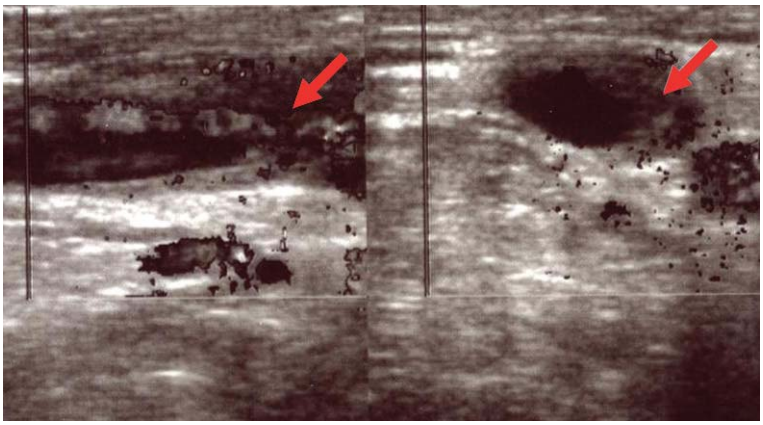

Figure 2. Complete thrombotic occlusion in the right internal jugular vein (arrows).

markers and questioning of the patient for rheumatologic diseases were both negative, especially for anti-phospholipid antibody syndrome and Behcet's disease. Tests for malignancy showed that tumor markers and the patient's clinical history were negative. First, the upper extremity deep vein thrombosis was considered for prediagnosis, and then the right upper extremity was elevated, and enoxaparin sodium was administered to the patient in suitable doses. During the service follow-up a detailed trauma history was taken. We noted that the patient had done spinning exercises for the last 18 months, and for the past 4 days he reported that he had done these spinning exercises harder and more frequently. Taking all results into account, the patient was diagnosed with PSS.

The swelling and redness regressed partially on the 3rd day of therapy. After 9 days, the patient was discharged and warfarin was continued for 6 months. In the first month follow-up visit we performed a control Doppler ultrasound and it showed partially thrombosed areas in the right brachiocephalic and internal jugular veins. Also, chronic thrombosis persisted in the left brachial and basilic veins. To exclude any hidden malignancy, we performed a chest, neck, and abdominal computed tomography (CT). These CT images showed that there was no evidence for malignancy, and some sections showed filling defects in the vena cava superior, which were attributed to thrombotic materials (Fig. 3). Therefore, the patient was treated with anticoagulants for 6 months.

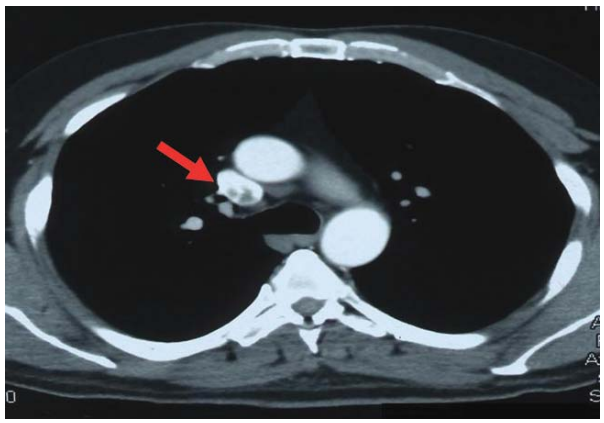

Figure 3. Complete thrombotic occlusion in the right internal jugular vein (arrows).

DISCUSSION

Primary UEDVT is a rare disorder and is also called effort thrombosis (PSS) or idiopathic UEDVT. PSS accounts for 10-20% of spontaneous UEDVT.[4] In this syndrome, patients are usually young and healthy, and spontaneous UEDVT develops most commonly in the dominant arm of patients.[2] Heavy exertion such as retroversion, hyperabduction, and extension of the arm can lead to microtrauma of the endothelium with the activation of the coagulation cascade.[5] Anatomical abnormalities involving the thoracic outlet have a role in the pathogenesis of effort thrombosis, however no anatomical abnormalities were detected with chest X-ray or CT in our patient.

Doppler ultrasound is the initial imaging modality for diagnosis because it is cheap, non-invasive, and highly sensitive and specific for UEDVT.[3] In our

Table 1. Treatment methods of Paget-von Schröetter syndrome

Anticoagulation with low-molecular weight heparin, unfractionated heparin and warfarin
Extremity elevation
Local catheter-directed thrombolysis
Thoracic outlet decompression with surgery and physical therapy
Vena cava filter
Venous stenting
Thrombectomy
Angioplasty

case, we preferred the Doppler ultrasound for the diagnosis.

In recent literature, a higher frequency of prothrombin gene mutation, factor V Leiden, and other thrombophilic states were reported in patients with primary UEDVT.[6] Cassada et al.[7] reported that approximately two-thirds of patients with PSS had concurrent thrombophilia. However, other researchers have refuted this connection by demonstrating that the frequency of inherited thrombophilias in patients with effort thrombosis was comparable to that of the general population.[8]

The role of inherited and acquired thrombophilic disorders in the development and progression of effort thrombosis is unclear. In this case we observed heterozygote factor XIII V34L and heterozygote MTHFR C677T gene mutations. The heterozygote F XIII V34L mutation is not a prothrombotic situation. It is common in white populations and both heterozygote and homozygote mutations have protective effects against venous thrombosis.[9] The heterozygote MTHFR gene mutation does not appear to be a risk factor for venous thrombosis, especially in patients who have normal homocysteine and folate ranges. Excess homocysteine in the plasma, however, is a risk factor, and is the target of therapeutic intervention.[10]

Current management of PSS varies widely, and there are many treatment strategies in clinical practice. Extremity elevation and anticoagulation alone are not effective treatments. Recent data have showed that local catheter directed thrombolysis, venoplasty, venous bypass, stents, and thrombectomy can be used as the management strategies (Table 1). Anticoagulation therapy should be continued at least 6 months after any treatment method.[2,3]

In conclusion, this case report suggests that heavy and repetitive exercise can activate the thrombotic cascade in healthy patients.

Conflict-of-interest issues regarding the authorship or article: None declared

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Key words: Adult; anticoagulants/therapeutic use; exercise; Paget-Schroetter syndrome; venous thrombosis/drug therapy/etiology.

Anahtar sözcükler: Erişkin; antikoagülanlar/terapötik kullanım; egzersiz; Paget-Schroetter sendromu; venöz tromboz/ilaç tedavisi/etyoloji.