

A Rare Cause of Upper Extremity Thrombosis: Paget-Schroetter Syndrome

Üst Ekstremitte Trombozunun Nadir Bir Nedeni: Paget-Schroetter Sendromu

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ABSTRACT

Upper extremity deep vein thrombosis (UEDVT) is extremely rare in children and adolescents. Paget - Schroetter Syndrome (PSS) is the primary, spontaneous thrombosis of the subclavian venous (SV) axillary tract. Herein, we report on PSS in two children who did regular body building exercises. 16-year-old two male patients admitted with complaints of swelling, coldness and pain on the right upper extremity. Their recent history disclosed that they have been doing advanced body building exercises for the last year. Physical examination revealed a difference of 4 cm and 5 cm in the diameter of each arm, as well as coldness and venous engorgement in the upper extremities. An acute thrombus was present in the proximal right SV lumen and no flow was detected on upper extremity venous doppler ultrasound. Surgical treatment was performed in addition to medical thrombolysis treatment. Early diagnosis and treatment of PSS is important for better outcomes. PSS should be considered in young patients with UEDVT, especially those with a history of vigorous exercise.

Key Words: Exercises, Paget - Schroetter Syndrome, Thrombosis

ÖZ

Üst ekstremitte derin ven trombozu (UEDVT) çocuklarda ve ergenlerde oldukça nadir görülür. Paget-Schroetter sendromu (PSS), subklavyen venöz (SV) aksiller sistemin primer, spontan trombozudur. Burada; düzenli vücut geliştirme egzersizleri yapan PSS tanısı alan iki hastayı bildirmekteyiz. 16 yaşında iki erkek hasta sağ üst ekstremitede şişlik, soğukluk ve ağrı şikayetiyle başvurdu. Yakın zamanda, vücut geliştirme egzersizleri yaptıkları öğrenildi. Fizik muayenede her iki kol çaplarında arasında fark olduğu, üst ekstremitelerde soğukluk ve üst ekstremitte toplar damarlarında belirginleşme dikkat çekmişti. Her iki olguda da üst ekstremitte venöz doppler ultrasonografide sağ SV proksimal lümeninde akut trombüs saptandı. Medikal tromboz tedavisine ek olarak cerrahi tedavi uygulandı. Tromboz sonrası iyileşmenin optimal olabilmesi için PSS'nin erken tanı ve tedavisi önemlidir. Bu nedenle ağır egzersiz öyküsü olan genç erkeklerde üst ekstremitteyi ilgilendiren şişlik, soğukluk, ağrı yakınmalarında PSS düşünülmelidir.

Anahtar Kelimeler: Egzersiz, Paget-Schroetter Sendromu, Tromboz

INTRODUCTION

Upper extremity deep vein thrombosis (UEDVT) is extremely rare in children and adolescents (1). The underlying conditions include mediastinal mass, central venous catheter, or idiopathic (1,2). Paget - Schroetter Syndrome (PSS) is the primary,

spontaneous thrombosis of the subclavian venous (SV) axillary tract (2). Herein, we report PSS in two children who did regular body building exercises and discuss treatment strategies in the light of the current literature. We also aimed to draw attention to PSS which is rare in childhood, but the clinical presentation is easy to define and the management is peculiar.



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CASE REPORT

Case 1: A 16-year-old male patient presented with complaints of swelling, coldness and pain in the right upper extremity. His recent history disclosed that he had been doing advanced body-building exercises for the last year. Physical examination revealed a 4 cm difference in the diameter of his arms, as well as coldness and venous engorgement the right upper extremity (Figure 1). A thrombus extending from the right proximal of the subclavian vein to the axillary vein was detected via doppler ultrasonography. Enoxaparin was initially started subcutaneously 1 mg/kg/ twice daily. Enoxaparin was discontinued after obtaining INR values over 2 were achieved with warfarin treatment. Then, INR values were adjusted to be between 2-3. Genetic risk factors for thrombosis including Factor V Leiden, and Prothrombin G20210A revealed no mutation. Anticoagulant treatment was stopped after 6 months. However, the swelling, numbness and diameter difference in the right arm recurred two months after the warfarin withdrawal. Partial thrombosis was again detected in the right SV. The patient's computed tomography (CT) angiography was compatible with PSS. Oral anticoagulation treatment was restarted. After 5 months there



Figure 1: Swelling of right upper extremity, discoloration due to circulatory disorder (Case 1).

was a 1 cm difference in diameter, mild drowsiness and pain in the right arm of the patient. These findings were attributed to post-thrombotic syndrome. Control doppler ultrasound revealed that the right subclavian venous calibration was minimally reduced compared to the other branches, the vessel surface was irregular due to the thrombus, venous flow was markedly reduced in the compulsive movements of the arm, and several venous collaterals were found in the right axillary region. To the decompress of the PSS, the first right costa resection and scalenectomy were performed with a right transaxillary intervention. The patient was treated with warfarin for 3 months postoperatively. Pain and drowsiness were relieved.

Case 2: Another 16-year-old male patient presented with a complaint of swelling, coldness and pain on the right side of the body. His recent history disclosed that he was doing advanced body building exercises. Physical examination revealed a 5 cm diameter difference, and cyanosis of the right arm. An acute thrombus was present in the proximal lumen of the right SV and venous doppler ultrasonography of the upper extremity showed no flow. The first 10 cm of the proximal portion of the cephalic vein was found to be completely occluded by thrombus. There was no family history of thrombosis. Enoxaparin was initially started subcutaneously 1 mg/kg/ twice daily and switched to warfarin as in Patient 1. Genetic testing revealed a heterozygous MTHFR C677T mutation and a homozygous PAI-1 4G/4G polymorphism. The patient was consulted with thoracic surgery for decompression. For the decompression of the PSS, a right first costa resection and scalenectomy were performed with a right transaxillary intervention. He did well after the operation with relief of symptoms. However, he lost follow-up one month after surgery.

DISCUSSION

Upper extremity DVT due to a primary cause is referred to as PSS that is associated with subclavian and axillary venous compression. In young, healthy patients presenting with UEDVTs, it is important to consider the primary causes (1,2). Early recognition of UEDVT's is important to reduce life-threatening complications such as pulmonary embolism (5). In PSS, thrombus formation is usually triggered by the subclavian venous exertion (2). The subclavian vein passes through the clavicle, first rib and subclavian muscles. Endothelial damage caused by recurrent retroversion, hyperabduction and enlargement movements can activate the coagulation system (2,3). It has usually been reported in young men involved in tennis, wrestling, swimming, billiards, hockey, baseball (3). Both of our patients were healthy young adolescents who were doing body building.

Upper extremity DVT usually presents with severe and sudden pain, swelling, numbness, and discoloration after a heavy

exercise or work. The right upper extremity is more likely to be affected because the majority of patients, like our patients, are right-handed.

Imaging is required to confirm the diagnosis. Doppler ultrasonography has been proposed as the first diagnostic tool because of its high sensitivity and specificity (4). The diagnoses of our patients were quickly confirmed by doppler USG.

Treatment aims to prevent complications and obstructive symptoms (6). The main treatment approaches should be discussed depending on the absence of clinical trials to guide the treatment plan (5). More aggressive treatment approaches including thrombolysis and surgery may be required in addition to anticoagulation to prevent symptoms and post-thrombotic complications (6). In our patients, anticoagulant treatment together with surgery was successful. Therefore, we believe that thrombolysis was not always necessary in the treatment.

In conclusion; PSS should be considered in young patients with upper extremity thrombosis, especially in young men with a history of heavy exercise. Early diagnosis and treatment of PSS is important for better outcomes.

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