Varicose Vein Surgery in Behçet’s Disease

Hulusi Behçet (1889-1948), the first professor in Turkey, recognized “the disease” which is currently known as “Behçet disease”, “Behçet Syndrome”, “Behçet’s triad”, “Tri-symptom Behçet” or “Morbus Behçet” in 1924.¹

Behçet’s disease (BD) is a type of vasculitis which affects skin-mucosa, ocular, musculoskeletal, gastrointestinal, neurologic and vessel systems.¹ This multisystem disease has highly seen in the Middle and Far East, which has nearly 0.4% prevalence in Turkey. The onset of the disease mainly occurs in the third decade and both gender are affected, whereas BD progress a more severe course in men, especially in ocular involvement, folliculitis, skin pathergy test positivity and vascular involvement (vasculo-Behçet).²

“Inflammation is one of the pathologic hallmarks of Behçet’s disease, which affects vessels of all sizes, known as vasculo-Behçet”.³ Vasculo-Behçet’s disease is observed in 14.7-27.7% of Behçet’s disease patients.⁴

One of the major manifestations of vasculo-Behçet is venous involvement. Venous vessels are affected more frequently than arterial system. Vasculo-Behçet with venous involvement, with or without venous thrombosis, may cause erythema, induration, leg pain, varicose veins, edema, skin hyperpigmentation, venous ulcer(s) in the lower leg, with substantial impact on quality of life.

Lower extremity venous thrombosis (LEVT) is the most type of venous disease.⁴ Venous thrombus formation in vasculo-Behçet patients is both extensive and adherent, hence, thromboembolism due to thrombus mass is rarely seen.⁵ The pathogenesis of thrombosis is due to inflammation of the venous wall rather than procoagulant status of blood.⁶

LEVT is relatively an early onset which appears within the first few years of disease, which involves highly femoral and popliteal veins. Chronic LVET leads to post-thrombotic syndrome due to venous hypertension which develops up to 50% after proximal LVET.⁷ Leg edema, with or without lymph content, as well as venous ulcers are predicted severe complications.
Unfortunately, the drawbacks of vasculo-Behçet are not limited to problems mentioned above. The main challenge is the systemic disease itself, which proceed with an undulating course with precipitating and remission periods.

From clinical observation as well as what is known of the pathology that take place in the development of isolated varicose veins, it is predictable that radical surgical intervention of varicosities in Behçet disease is only for “temporary and/or limited value”. This data may be an “urban legend”, with a consideration in light of the high rate or recurrence. So, the management of varicosis in Behçet’s disease was limited to conservative approaches.

There are still no standard guidelines and/or algorithm of prevention and/or management of venous disease in patients with BD.

This so-called “temporary value” was shifted to “when needed” in our daily clinical practice. When surgery is needed in patients with BD, systemic immunosuppressive medication should be administered under the supervision of rheumatic physicians. Basic recommendation is the evaluation of erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) levels to estimate the status of rheumatic activity.

In the present work, our experience as well as foresight in the surgical treatment of BD patients with lower leg venous involvement are briefly presented. Isolated varicosis in BD patients (i.e. varicose veins without venous thrombosis) without surgical treatment had a high morbidity rate due to complications as venous thrombosis. If isolated varicosis was unrecognized or left untreated, it often progressed and developed the signs and symptoms of venous thrombosis and leg ulcers.

Venous thrombosis occurs via three mechanisms ie, Virchow’s triad. The mechanisms are decreased flow rate of blood, damage to the blood vessel wall and an increased tendency of the blood to clot. Venous surgery leads to optimized blood flow to reduce venous thromboembolism, which may be fit for Behçet’s disease patients. As presented, enhancements of both blood velocity and volume flow are key factors in the prevention of venous stasis and ultimately deep vein thrombosis.

As presented, the pathogenesis of thrombosis is due to inflammation of the venous wall rather than procoagulant status of blood, so if one decrease the surface of venous wall in Behçet’s disease by venous surgery, the inflammation of vein wall which leads to venous thrombosis might decrease.

Finally, from clinical observation in 28 patients as well as our experience in Behçet’s disease and venous surgery, it is predictable that superficial vein surgery in patients with superficial venous insufficiency without venous thrombosis might decrease expected venous LEVT as well as leg ulcers in Behçet disease.