Comments on the risk of pulmonary embolism in Behçet's syndrome

Serdal Ugurlu1; Yusuf Yazici2; Gulen Hatemi1; Sebahattin Yurdakul1*

1Division of Rheumatology, Department of Internal Medicine, Cerrahpasa Medical Faculty, University of Istanbul, Istanbul, Turkey
2Department of Rheumatology, NYU Hospital for Joint Diseases, New York University, New York, NY, USA

Dear Editor,

Zöller et al. recently reported on the risk of pulmonary embolism in patients with 33 different autoimmune diseases from Sweden.1 In this study they obtained data from the MigMed2 database, constructed from several national Swedish data registers. They studied the risk of venous thromboembolism in patients without previous hospital admission for venous thromboembolism. In addition, these patients had a history of admission for pulmonary embolism with a primary or secondary diagnosis of an autoimmune disorder between Jan 1, 1964, and Dec 31, 2008. The risk of pulmonary embolism was classified into five categories; patients who had <1 year, 1-5, 5-10, >10 years of follow up and all combined. The risk of pulmonary embolism was reported as 6.38 (95% CI: 6.19-6.57) for 33 autoimmune disorders in the first year after hospital admission. On the other hand, the overall follow-up time risk was determined as 1.59 (95% CI: 1.56-1.61). The highest risk for pulmonary embolism was reported in patients with polymyositis and dermatomyositis; 16.4 (95% CI: 11.57–22.69). They showed that the risk of pulmonary embolism during the first year after admission for BS was 9.03 (95% CI: 6.51-12.22), but the overall follow-up time risk was less at 1.68 (95% CI: 1.45-1.93).

Several issues need further discussion: It was suggested in the introduction of the manuscript that BS was associated with a high risk of venous thromboembolism with reference to a review by Yazici Y et al.3 Although venous thrombosis is present, thromboembolism does not occur in BS probably because of the strong adherence of thrombi to the diseased veins.3 Moreover, pulmonary embolism was rarely found in a large Japanese autopsy registry.4 In another study, pulmonary embolism was not observed in 47 patients with pulmonary artery involvement of BS.5 Pulmonary artery involvement included either pulmonary artery aneurysms or ‘in situ’ pulmonary artery thrombosis which developed as a complication of underlying vasculitis. Furthermore, it is to be noted that, immunosuppressive agents are essential in treating venous thrombosis and preventing relapses in BS. As reported in retrospective studies,
solo anti-coagulation is ineffective in treating venous disease
of BS.\textsuperscript{6,7} Vascular involvement occurs in up to 40% of BS
patients, and the lower extremity deep vein thrombosis is the
most common type with a frequency of 60-80%.\textsuperscript{9} Thus a SIR
of 1.68 in such a population would indeed point out to the
scarcity of pulmonary embolism in BS patients. Finally, BS is
most active during the third decade of life, burning out after the
fifth decade. The tendency for the disease to abate with the
passage of time has been reported by several centers
around the globe.\textsuperscript{8-12} We had observed that mucocutaneous
and joint lesions decreased in frequency after 20 years of dis-
ease onset and in at least 60% of the patients the disease had
been observed to fade away.\textsuperscript{9} Thus it is unlikely that an
increased risk of pulmonary embolism in men between the
ages of 70-79 and in women between 50-69 could be related
to BS itself. This could be attributed to another cause rather
than BS and older patients with pulmonary embolism should be
investigated further and anti-coagulated if needed.

References
1. Zöller B, Li X, Sundquist J, Sundquist K. Risk of pulmonary
embolism in patients with autoimmune disorders: a nationwide fol-
3. Hamuryudan V, Melikoglu M. Vascular involvement in Behçet’s
York: Springer; 2010. p. 115-134.
T. Pathologic features of Behçet’s syndrome: a review of Japanese
5. Seyahi E, Melikoglu M, Akman C, et al. Pulmonary artery involve-
ment and associated lung disease in Behçet disease: a series of 47
6. Ahn JK, Lee YS, Jeon CH, Koh EM, Cha HS. Treatment of
venous thrombosis associated with Behçet’s disease: immunosup-
pressive therapy alone versus immunosuppressive therapy plus
sants reduce venous thrombosis relapse in Behçet’s disease. Arthritis
Rheum 2012;64:2753-60.
8. Kural-Seyahi E, Fresko I, Seyahi N, et al. The long-term mortali-
ty and morbidity of Behçet syndrome: a 24-month outcome survey
of 387 patients followed at a dedicated center. Medicine (Baltimore)
2003;82:60-76.
10. Mamo JG, Baghdassarian A. Behcet’s disease; a report of 28 cases.