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LETTER TO THE EDITOR

Intracardiac thrombosis in Behçet disease: Predictable complications associated with the risk of sudden death

Dear Editor,

Although vascular manifestations are the main predictors of mortality in Behçet disease (BD),[1] it is important to note that these patients tended to deny any history of genital ulcers, joint pain, and skin rash. Thus, the important issue about vasculo-Behçet is which biomarkers can predict cardiac complications. We describe a case of recurrent erythema nodosum (EN) which developed large intracardiac thrombi consistent with vasculo-Behçet.

A 32-year-old man presented with a 2-month history of painful indurated erythema on his legs (Figure 1A,B) and episodic fever. Physical and laboratory examination were unremarkable except for slight CRP elevation (1.5 mg/dL). Histological examination of the erythema was consistent with EN (Figure 1C). He did not fulfill the International Criteria for BD.[2] Although he was under an excellent disease control with colchicine, CRP levels increased progressively without any clinical symptoms of BD (Figure 1A). Seven months later, he presented to the cardiovascular department with dyspnea and abnormal electrocardiogram findings. Three-dimensional computed tomography showed a large thrombus in the right ventricle (Figure 1D). Cardiac catheterization confirmed right ventricular thrombi. He was started on oral prednisolone and warfarin. His presentation could then be considered consistent with vasculo-Behçet. One month later, the thrombi had disappeared.

A review of case reports suggests that cardiac complications in BD may remain entirely asymptomatic until patients present with an acute cardiac event.[1,3,4] Of note, many patients presented with

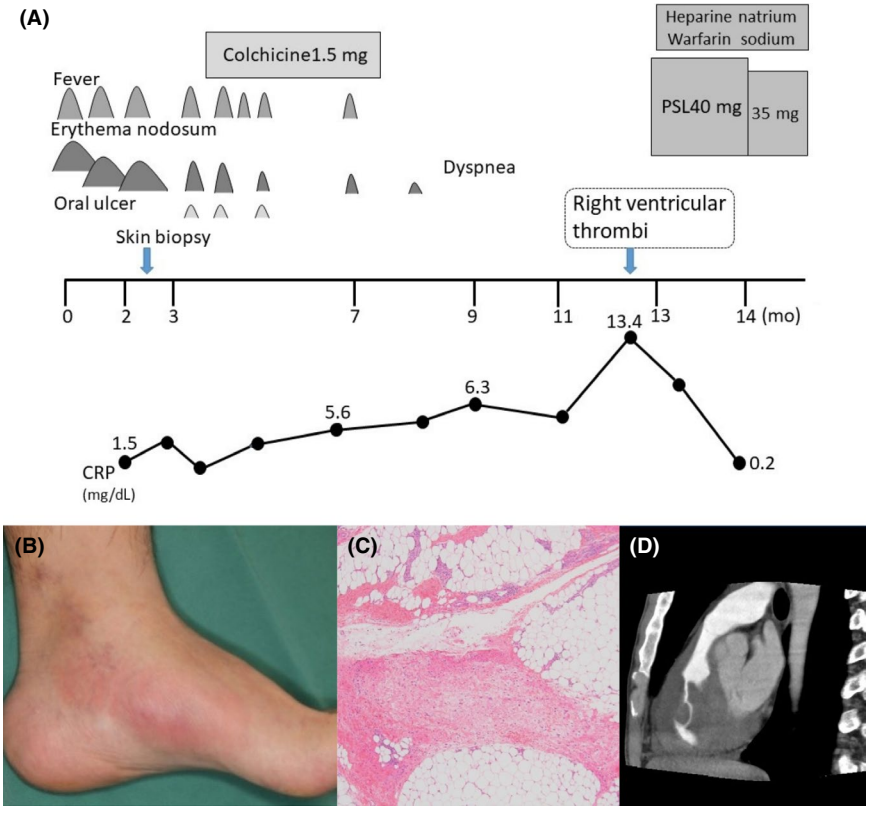

FIGURE 1 A, Clinical and laboratory course. B, Painful indurated erythema on leg. C, Aggregates of lymphocytes and neutrophils around vessels of the plexus (stain type: hematoxylin and eosin, magnification level:×100). D, A large thrombus in the ventricle

intracardiac thrombi as the first vascular event.[1,4] In addition, most vasculo-Behçet patients cannot fulfill the criteria for BD because of the absence of ocular involvement.[1] Consistent with this view, the present case was never formally diagnosed as having BD. Thus, a history of clinical illness compatible with BD should be sought in the patients presenting with cardiac complications. It should be also noted that development of intracardiac thrombi may be recognized only years later at the time of sudden death.

In the present case, only serum CRP levels continued to increase despite resolution of other clinical symptoms until the onset of dyspnea. This suggests that CRP levels can be used as useful biomarker for identifying patients with suspected BD at a risk of subsequently developing intracardiac thrombi. Even when clinical symptoms consistent with BD are absent, monitoring of CRP levels can help clinicians stratify patient's risk of cardiovascular complications. CRP levels associated with intracardiac thrombi may point out the inflammatory nature of thrombi in BD. Gerg et al. reported that thrombosis in BD differs from other forms of thrombosis in that thrombi stick firmly to the vessel wall and that treatment for thrombosis in BD should target underlying vasculitis.[5] Indeed, in our case, intracardiac thrombi resolved with immunosuppressive agents.

In conclusion, BD should be considered in the differential diagnosis when patients with a history of repeated fever, EN-like lesions, and high CRP levels but without eye involvement develop vascular manifestations. Prompt recognition of vasculo-Behçet presenting with intracardiac thrombi as the initial symptom of BD and rapid treatment with oral prednisolone and anticoagulation would reduce the risk of sudden death.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

Noriko Sato MD
Yoshiko Mizukawa MD, PhD
Tetsuo Shiohara MD, PhD

Department of Dermatology, Kyorin University School of Medicine, Mitaka, Japan

Correspondence
Noriko Sato, Department of Dermatology, Kyorin University School of Medicine, Shinkawa, 6–20–2, Mitaka, Tokyo 181–8611, Japan.
Email: nrk.cc.0605-ao@hotmail.co.jp

ORCID

Noriko Sato https://orcid.org/0000-0003-2942-3851
Tetsuo Shiohara https://orcid.org/0000-0003-1637-6312

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