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CASE REPORT

**Takashi Kajiya · Ryuichiro Anan · Masahiro Kameko
Naoko Mizukami · Shinichi Minagoe · Shuichi Hamasaki
Ikuro Maruyama · Ryuzo Sakata · Chuwa Tei**

Intracardiac thrombus, superior vena cava syndrome, and pulmonary embolism in a patient with Behçet's disease: a case report and literature review

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Abstract A 26-year-old woman with intermittent fever was admitted to our hospital, and gradually developed facial edema. Examinations including computed tomography, transesophageal echocardiography, digital subtraction angiography, and pulmonary perfusion scintigraphy revealed intracardiac thrombus, superior vena cava syndrome, and pulmonary embolism. Clinical findings and laboratory data led us to make a diagnosis of Behçet's disease. Combination of intracardiac thrombus, superior vena cava syndrome, and pulmonary embolism are rare complications in Behçet's disease. Behçet's disease should be considered in the differential diagnosis of intracardiac mass of the right heart, and early diagnosis and treatment are essential for the management of Behçet's disease especially with large-vessel manifestations. In addition to a case report, we review the literature and report the characteristics of intracardiac thrombus in Behçet's disease.

Key words Behçet's disease · Intracardiac thrombus · Superior vena cava syndrome · Pulmonary embolism

Introduction

Behçet's disease is a systemic inflammatory disorder of unknown cause that is frequent among Japanese and Mediterranean populations.[1,2] Intracardiac thrombus formation is uncommon in Behçet's disease,[3] although large venous or arterial lesion occur in 7%–38% of patients.[1,3] The number of reports on intracardiac thrombus in Behçet's disease is increasing because of the world wide spread of echocardiography. Fifty-four patients with Behçet's disease associated with intracardiac thrombus have been reported in the literature.[3–29]

We describe a patient with Behçet's disease with intracardiac thrombus associated with superior vena cava syndrome and pulmonary embolism. Combinations of intracardiac thrombus, superior vena cava syndrome, and pulmonary embolism are rare complications in Behçet's disease. In addition to a case report, we have reviewed the literature, and report the characteristics of intracardiac thrombus in Behçet's disease.

Case report

A 26-year-old woman was admitted for evaluation of intermittent fever for 6 months with a peak temperature of 39°C. She had a history of recurrent oral aphthous ulcers, epigastric pain, and genital ulcers. On admission, her body temperature was 38°C and the remainder of the physical examination was unremarkable. Laboratory results included a white blood cell count of 14.7×10^9/l, a C-reactive protein concentration of 14.28 mg/dl, and an erythrocyte sedimentation rate of 128 mm/h. No antinuclear antibodies were detected. Microbiologic studies identified no causative organisms. Chest X-ray on admission showed bilateral consolidations without cardiomegaly. Computed tomography of the chest showed multiple nodular densities scattered in bilateral lung fields (Fig. 1). Echocardiography showed a cystic mass in the right atrium that extended to the superior vena cava. Transesophageal echocardiography performed 1 week later showed a new abnormal echo (8 × 15 mm) that was mobile and located at the annulus of the tricuspid valve (Fig. 2). Facial edema developed during the course. Digital subtraction angiography revealed bilateral occlusion of subclavian veins and development of intercostal

T. Kajiya · R. Anan (✉) · M. Kameko · S. Minagoe · S. Hamasaki · C. Tei
Department of Cardiovascular, Respiratory and Metabolic Medicine, Graduate School of Medicine, Kagoshima University, 8-35-1 Sakuragaoka, Kagoshima 890-8520, Japan
Tel. +81-99-275-5318; Fax +81-99-265-8447
e-mail: louanan@m2.kufm.kagoshima-u.ac.jp

N. Mizukami · I. Maruyama
Department of Laboratory and Molecular Medicine, Graduate School of Medicine, Kagoshima University, Kagoshima, Japan

R. Sakata
Department of Cardiovascular Surgery, Graduate School of Medicine, Kagoshima University, Kagoshima, Japan

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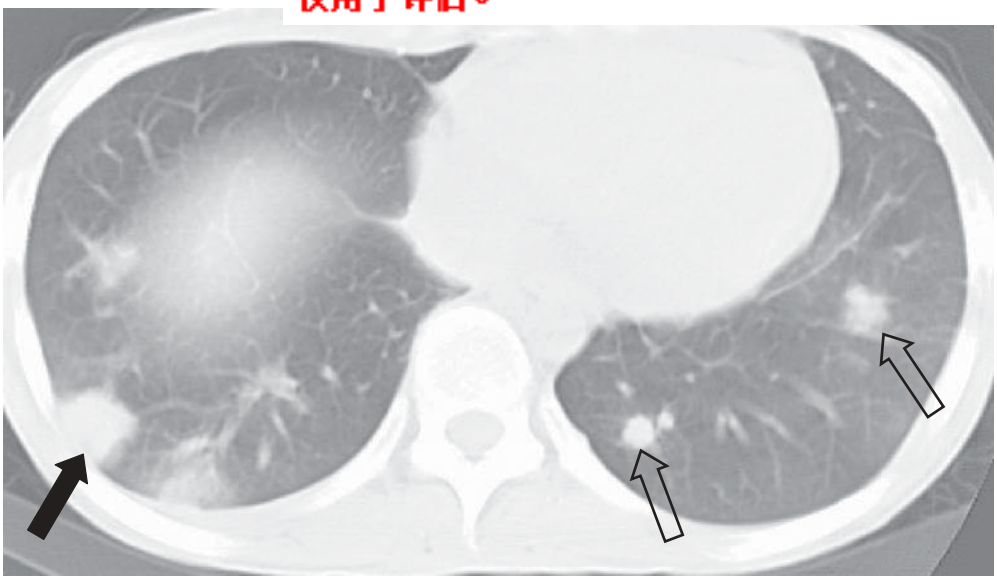

Fig. 1. Computed tomography shows wedge-shaped infiltration in the subpleural right lobe (*closed arrow*) and nodular densities in the left lobe (*open arrows*)

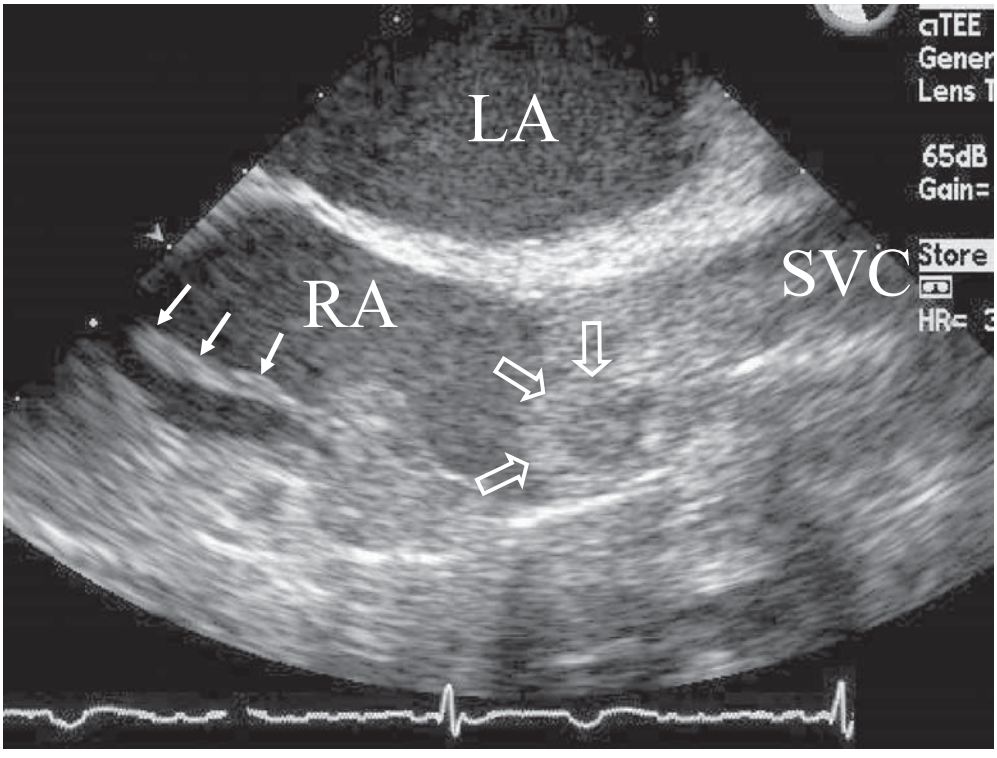

Fig. 2. Transesophageal echocardiography shows a mobile echo located at the annulus of the tricuspid valve (*closed arrows*) and cystic mass in the right atrium that extended to the superior vena cava (*open arrows*). *RA*, right atrium; *LA*, left atrium; *SVC*, superior vena cava

veins as collateral veins, leading to a diagnosis of superior vena cava syndrome (Fig. 3). Pulmonary perfusion scintigraphy demonstrated bilateral pulmonary perfusion defects and a diagnosis of pulmonary thromboembolism was made (Fig. 4). Differential diagnoses of the masses were intracardiac thrombus, vegetation, and cardiac tumor. Surgery was selected to remove the masses for fear of massive pulmonary embolism leading to death.

Yellowish white, rough-surfaced masses were removed surgically from the right atrium and superior vena cava. Pathologic diagnosis for the masses was thrombus with inflammatory granulation. No other thrombi were found in the inferior vena cava, superficial veins, or deep veins.

We investigated possible causes of the patient's thrombotic tendency. She gave no history of arrhythmia, pregnancy or medications which might have thrombotic effect. Lupus anticoagulant, protein C, protein S, and antithrombin III were normal. We detected the presence of human leukocyte antigen B51 and a serum IgD concentration of 7.1 mg/dl (normal range, 0.3–4.0). The clinical picture, including recurrent oral ulceration, recurrent genital ulceration, gastrointestinal complaints, arthritis, presence of

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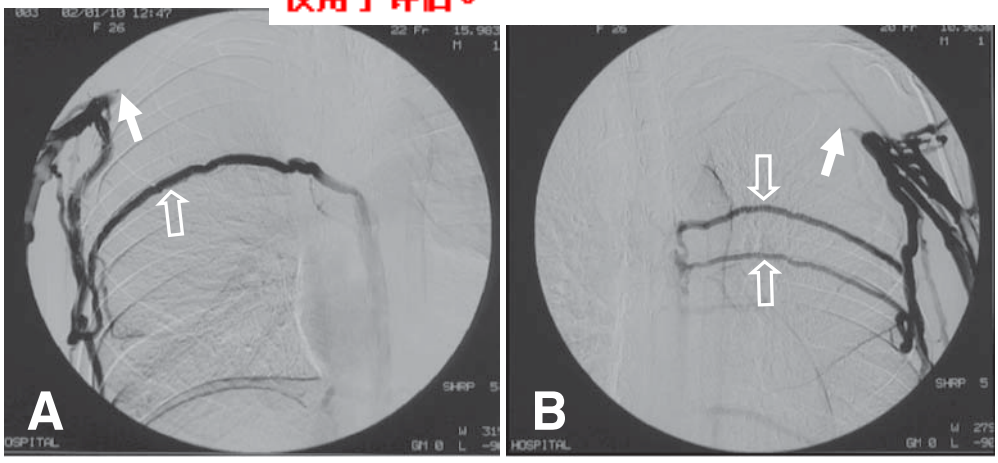

Fig. 3A,B. Digital subtraction angiography shows bilateral occlusion of subclavian veins (*closed arrows*) and development of intercostal veins (*open arrows*). **A** Infusion from right median cubital vein. **B** Infusion from left median cubital vein

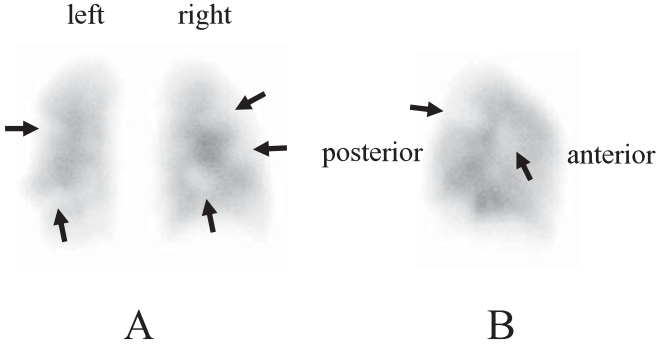

Fig. 4A,B. Pulmonary perfusion scintigraphy shows bilateral perfusion defects (*arrows*). **A** Posterior view. **B** Right lateral view

human leukocyte antigen B-51, and elevated serum IgD, led us to the diagnosis of Behçet's disease.[1,2] After starting a combination of prednisolone, colchicine, and warfarin, her facial edema and high fever relieved and the C-reactive protein reduced to within normal range.

For 3 years after the surgery, recurrence of intracardiac thrombus has not been identified. However, the patient has been treated for intestinal Behçet's disease, which was identified after the surgery.

Discussion

We have reported a patient with Behçet's disease associated with intracardiac thrombus, superior vena cava syndrome, and pulmonary embolism. Intracardiac thrombus formation is uncommon in Behçet's disease.[1,3] In a MEDLINE search covering articles (including English abstracts) from 1966 to September 2006 using the keywords "Behçet's disease," "thrombus," and "cardiac" or "intracardiac," we have found 54 cases (Table 1).[3–29] Forty-eight of the 54 patients (89%) are male, and the mean age is 27.3 ± 8.4 years old (range, 12–51 years old).

Our patient was associated with intracardiac thrombus in the right side of the heart. Including our case, 53 of the 55 patients (96%) are associated with thrombus in the right side of the heart or both side of the heart, and 2 of the 55 patients (4%) are associated with thrombus in the left side of the heart only.[3–29] Most of the intracardiac thrombus in Behçet's disease are seen in the right side of the heart.

Only six of the 55 patients (11%) with intracardiac thrombus are associated with superior vena cava syndrome. Association of superior vena cava syndrome with intracardiac thrombus is not common in Behçet's disease.

Thirty-seven of 55 patients (67%) with intracardiac thrombus are also associated with pulmonary thromboembolism. Incidence of pulmonary embolism is high in Behçet's disease in the presence of intracardiac thrombus.

Twenty-eight of 55 patients (51%) are associated with venous thrombosis. We have examined the source of intracardiac thrombus by repeated Doppler ultrasonography of the lower extremities, which did not detect any venous thrombus. Deep vein thrombus formation is found in about half of the patients with intracardiac thrombus in Behçet's disease.

Association of intracardiac thrombus, superior vena cava syndrome, and pulmonary embolism in patients with Behçet's disease seems to be rare. There have been only four cases including our case in the literature with this association (cases 31, 46, 50, and 55 in the Table 1). Of these 4 cases, 2 were associated with venous thrombosis and 2 were not. In patients with Behçet's disease with intracardiac thrombus, pulmonary embolism should be considered even when venous thrombosis cannot be detected. The mobile thrombus in the right atrium in our patient supports that the source of the pulmonary embolus could be intracardiac thrombus.

The mechanism of intracardiac thrombus formation in Behçet's disease has not been fully clarified. In patients with Behçet's disease, deep vein thrombosis has been documented in about half of the patients with intracardiac thrombus (Table 1). We speculate that endomyocardial fibrosis in the right side of the heart is one of the causes of intracardiac thrombus formation in patients with Behçet's disease without deep vein thrombosis.[1,3]

Sometimes cardiovascular and pulmonary involvements are seen before making diagnosis of Behçet's disease. Those manifestations are sometimes life-threatening and failure

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Table 1. Cases of Behçet's disease with intracardiac thrombus ar

Case	Age	Sex	Location of intracardiac thrombus	SVC syndrome	Pulmonary thromboembolism	Venous thrombosis	References
1	29	M	RV		+	+	Davies[a]
2	45	M	RA, RV			+	Buge et al.[a]
3	35	M	RV		+		Candan et al.[a]
4	15	M	RV				Augarten et al.[a]
5	29	M	RV		+	+	Lie[a]
6	18	M	RA, RV, LV			+	Vanhaleweyk et al.[a]
7	23	M	RV		+		El-Ramahi et al.[a]
8	18	M	RV, LV		+	+	El-Ramahi et al.[a]
9	30	M	RV		+	+	El-Ramahi et al.[a]
10	33	F	RV		+		Pottiez and Francois[a]
11	17	M	RV			+	Koc et al.[a]
12	29	M	RA, SVC	+		+	Sayin et al.[a]
13	51	M	LA				Madanat et al.[a]
14	29	M	RV		+	+	Mendes et al.[a]
15	32	M	RA			+	Islim et al.[a]
16	12	M	RA, PA		+		Nakata et al.[a]
17	27	M	RV		+		Soulami et al.[a]
18	32	F	LV				Huong et al.[a]
19	27	M	RV	+		+	Huong et al.[a]
20	48	M	RV		+	+	Huong et al.[a]
21	26	M	RA, RV		+		Rougin et al.[a]
22	28	M	RA, RV			+	Kirali et al.[a]
23	28	M	RA, RV				Harmouche et al.[a]
24	14	M	RV		+		Duchene et al.[a]
25	19	M	RA, RV		+	+	Mogulkoc et al.[3]
26	25	M	RV		+	+	Yasuo et al.[4]
27	19	M	RA, RV, PA		+	+	Gurgun et al.[5]
28	28	M	RA, RV, IVC				Basaran et al.[6]
29	16	M	RA, RV, IVC		+		Vaya et al.[7]
30	39	M	RA, RV		+		Dincer et al.[8]
31	27	M	RA, RV, SVC	+	+	+	Ozatli et al.[9]
32	33	M	RA, RV		+	+	Baykan et al.[10]
33	19	M	RA, RV, PA		+		Yoshida et al.[11]
34	27	M	RA		+		Cemri et al.[12]
35	29	F	RA, PA		+	+	Altunkeser et al.[13]
36	38	M	N/A[b]		+	+	Hassikou et al.[14]
37	29	M	RA, RV, IVC		+		Houman et al.[15]
38	25	M	RA, RV				Tursen et al.[16]
39	19	M	RV, PA		+	+	Duzgun et al.[17]
40	23	M	RA, RV, PA, IVC		+	+	Goktekin et al.[18]
41	26	M	RA, RV			+	Ozdemir et al.[19]
42	25	M	RV, PA		+		Kaya et al.[20]
43	22	F	RA, RV		+		Noureddine et al.[21]
44	29	M	RA		+	+	Fekih et al.[22]
45	33	M	RV, PA		+		Ben Ghorbel et al.[23]
46	29	M	RA, SVC	+	+	+	Ben Ghorbel et al.[23]
47	25	M	RA, PA				Ben Ghorbel et al.[23]
48	40	F	RV			+	Ozer et al.[24]
49	31	F	RV			+	Darie et al.[25]
50	20	M	RA, RV, SVC	+	+		Hammami et al.[26]
51	29	M	RA, RV, PA, IVC		+		Hammami et al.[26]
52	36	M	RV, PA		+		Atalay et al.[27]
53	46	M	RA				Kaneko et al.[28]
54	20	M	RV, PA		+	+	Ernam et al.[29]
55	26	F	RA, SVC	+	+		Kajiya et al.[c]

IVC, inferior vena cava; LA, left atrium; LV, left ventricle; N/A, not available; PA, pulmonary artery; RA, right atrium; RV, right ventricle; SVC, superior vena cava; +, presence of the findings

[a] Reviewed by Mogulkoc et al.[3]

[b] Hassikou et al. did not describe the location

[c] Present case

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to diagnose a patient with Behçet's disease could be critical for the patient's outcome. In our patient, preoperative differential diagnoses included intracardiac thrombus, vegetation, and cardiac tumor, since the masses contained partially cystic areas and progressed rapidly together with nonspecific clinical features of pyrexia and malaise. It is sometimes hard to differentiate echocardiographically intracardiac thrombi, vegetations, and tumors.[30,31] The distinction is important, since treatments and prognoses differ.

In our case, digital subtraction angiography was effective for making a diagnosis of superior vena cava syndrome. Insertion of an arterial or venous catheter may induce either a thrombosis or pseudoaneurysm formation at the puncture site. Therefore, computed tomography or magnetic resonance angiography may be better methods for analyzing the status of Behçet's disease.[32–35]

For treatment, anticoagulant and thrombolysis are considered for intracardiac thrombus.[36] However, because recurrences of intracardiac thrombus are reported in cases that were treated by anticoagulant only,[8,10,15] systemic glucocorticoid and/or immunosuppressant are highly recommended before using anticoagulant in cases of Behçet's disease. Surgery should be considered in cases with contraindications to thrombolysis or if thrombolysis is ineffective.[36]

In conclusion, we have reported a patient with Behçet's disease associated with intracardiac thrombus, superior vena cava syndrome, and pulmonary embolism. Intracardiac thrombus associated with Behçet's disease should be considered in the differential diagnosis of intracardiac mass of the right heart, and early diagnosis and treatment are essential for the management of Behçet's disease especially with large-vessel manifestations.

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