
Cardiac mass in Behçet's disease

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Received on December 2, 2011; accepted in revised form on February 17, 2012.

Clin Exp Rheumatol 2012; 30 (Suppl. 72): S27-S31.

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Key words: Behçet's disease, cardiac mass

ABSTRACT

Cardiac mass is a rare manifestation of Behçet's disease (BD). Intracardiac thrombosis, endomyocardiofibrosis, endocardial fibroelastosis, inflammatory mass and cystic change have been reported as different entities of cardiac mass in BD. Here we presented 6 cases of this rare manifestation of BD. The clinical and pathological features were reviewed.

Introduction

Behçet's disease (BD) is a systemic vasculitis characterised by recurrent oral aphthae and any of several systemic manifestations including genital aphthae, ocular disease, skin lesions, neurologic disease, vascular disease, or arthritis. Among the systemic vasculitis, BD is remarkable for its ability to involve blood vessels of all sizes – small, medium, and large – on both the arterial and venous sides of the circulation (1). Cardiac mass is a rare manifestation of BD. Intracardiac thrombosis (2-18), endomyocardiofibrosis (11, 19-22), endocardial fibroelastosis (23), inflammatory mass (24), and cystic change (25) have been reported as different entities of cardiac mass in BD. However, most of these reports were case reports or systemic review, and cumulative view of this manifestation of BD is not available. Here we provide 6 cases of BD with cardiac masses. The clinical and pathological characteristic of these patients were reviewed.

Methods

A retrospective review of the medical records of in patients was conducted at Peking Union Medical College Hospital over a 20-year period. The medical records were accessed on all patients with the diagnosis of Behçet's disease and disease of cardiovascular system. Review of the records yielded 6 cases of BD with cardiac mass. Patient demographics and data were obtained from chart review. The pathologic data was obtained by review of the pathology

reports. Follow-up data were obtained from the medical records, contacting the treating physicians, or by discussions with the patients.

Results

Clinical features

A review of the medical records of in patients of Peking Union Medical College Hospital through 1990 to February 2012 revealed 642 patients diagnosed with Behçet's disease. Cardiac mass lesions were found in 6 patients, accounting for approximately 1% of the BD population. Clinical features of the 6 patients are summarised in Table I and II. Patients included 5 males and 1 female, with a male predominance (83%). The mean age by the time of cardiac mass lesion was 16±14 years old (ranging from 19 to 40 yrs). All patients fulfilled the ISG (International Study Group) diagnostic criteria for BD created in 1990 (26), that is, the presence of oral aphthous ulcer in addition to any two of four additional features: genital aphthous ulcer, ocular involvement, positive pathergy test, or skin lesions (erythema nodosum, papulopustular lesions, acneiform nodules, pseudofolliculitis). All patients had thrombosis of sites other than heart. Pulmonary artery was the mostly involved vessel, and 5 patients (83%) had pulmonary thrombosis. Other sites of thrombosis included ocular artery (patient 2), superior/inferior vena cava (patients 3, 4 and 6) and aorta (patient 5). While thrombosis accounted for majority of the vascular disease, 1 patient had aneurismal changes of aorta (patient 5) (Table I). Disease duration before discovery of cardiac mass varied from 1 month to 10 years, with a mean disease duration of 5.4 years. Most patients had fever and weight loss as their presenting symptom, and had various degrees of dyspnea and cough, which might be contributed to concomitant pulmonary infarction. 3 patients were diagnosed with Behçet's disease prior to presentation and were on oral corticosteroids (Table II).

Competing interests: none declared.

Table I. Principle clinical features of Behçet's disease cases complicated with cardiac mass*.

Pt. no.	Sex	Age (yr)	Diagnostic signs					Other signs of BD				
			OU	GU	PAT	OL	SL	HLA-B5	Vasculitis	Thrombosis	Other	
1	M	37	+	+	+	–	+	–	–	PA	–	
2	M	35	+	–	+	–	+	–	–	OA	–	
3	M	19	+	+	–	+	–	–	–	PA, IVC	MR, TR, Pleural effusion	
4	M	21	+	+	n/a	–	+	n/a	–	PA, SVC, IVC	EP	
5	F	20	+	–	+	–	+	–	AA pseudo-aneurysm (supra-renal)	PA, AA,	MR, TR, Angina, Pleural effusion	
6	M	40	+	+	+	–	+	–	–	PA, SVC	TS, Pleural effusion, Pericardial effusion,	

*AA: aortic artery; BD: Behçet's disease; EP: encephalopathy; IVC: inferior vena cava; GU: genital ulceration; MR: mitral valve regurgitation; n/a: not available; OA: ocular artery; OU: oral ulceration; OL: ocular lesion; Pt: patient; PA: pulmonary artery; PAT: pathergy test; SL: skin lesion; SVC: superior vena cava; TR: tricuspid valve regurgitation; TS: tricuspid valve stenosis.

Table II. Clinical features by the time of cardiac mass*.

Pt. no.	Disease duration	Presenting symptoms						Prior treatment
		Fever	Cough	Dyspnea	Haemoptysis	Weight loss	Other	
1	10 years	+	+	–	–	–	–	–
2	1 month	+	–	+	–	+	TIA	–
3	4 years	+	–	–	–	+	–	Prednisone + CTX
4	2 years	+	+	+	+	+	–	Prednisone
5	6 years	–	–	+	–	+	Chest pain, Abdominal pain	Prednisone
6	5 years	+	–	+	–	–	–	–

*TIA, transient ischaemic attack.

Table III. Features of cardiac masses*.

Pt. no.	Diagnostic procedure	Presumed diagnosis	Comobid condition	Position	Size	Surgery	Gross morphology	Pathology
1	Echo, MRI	IE	–	RV, Apex	20×14mm	+	jelly-like	Acute and chronic inflammation with fibrous exudates and necrosis.
2	Echo, MRI	IE	–	LV, Apex RV, AS RA, FW	54×32mm 16×14mm 29×20mm	+	Yellow vegetation, with 2 cystic changes filled with inflammatory exudates (LV mass)	Acute and chronic inflammation with fibrous exudates, necrosis, granulation, and attachment of thrombosis (LV mass)
3	Echo, MRI	–	IE	RA	41×26mm	–		
4	Echo	–	congenital HOCM, CHF	RA, IVC RA, SVC RV, FW	8×14mm 9×9mm 8×6mm	– – –		
5	Echo	–	–	LV, VS	37×7mm	–		
6	Echo	–	CHF	Pericardial Cavity	n/a	–		

*AS: atrial septum; CHF: congestive heart failure; HOCM: hypertrophic obstructive cardiomyopathy; Echo: echocardiogram; FW: free wall; IE: infectious endocarditis; IVC: inferior vena cava; LV: left ventricle; n/a: not available; RA: right atrium; RV: right ventricle; SVC: superior vena cava; MRI: magnetic resonance imaging; VS: ventricular septum.

Features of cardiac masses

These are summarised in Table III. In all 6 patients, diagnosis of cardiac mass was made by echocardiography, and in some patients, confirmed by magnetic

resonance imaging (Fig. 1). In 2 patients, infectious endocarditis (IE) was the initial suspicion, and was initially treated with antibiotics (patients 1 and 2). In 1 patient, the right atrial mass

was complicated with IE (patient 3). 1 patient had multiple mass lesions in the pericardial cavity (patient 6). The other 5 patients had intra-cardiac mass lesions, and 2 of them had multiple le-

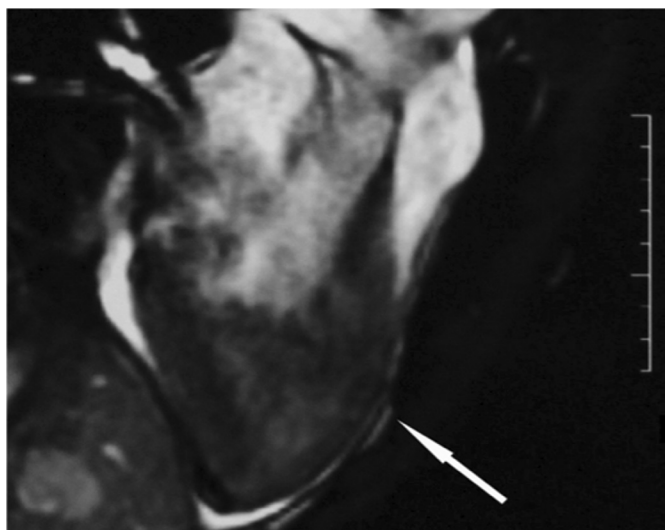


Fig. 1. MRI imaging of cardiac mass in a BD patient. The arrow shows a mass lesion adhering to the apex of the left ventricle.

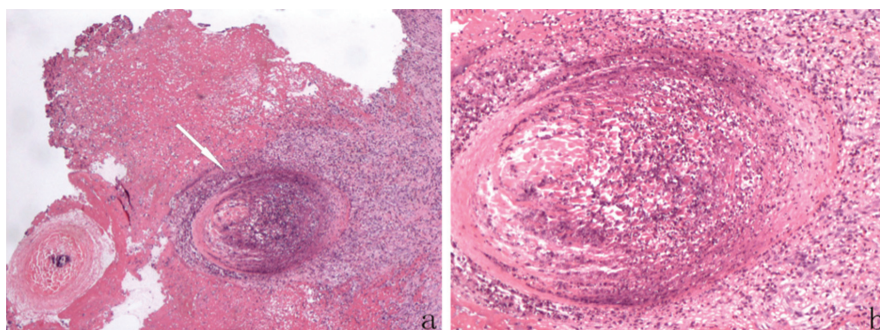


Fig. 2. Pathology of the cardiac mass in Fig. 1, revealing a inflammatory mass with thrombosis formation. (a) The arrow shows extensive inflammatory infiltration of a small vessels (H&E, $\times 60$). (b) Chronic and acute inflammation coexisted in the lesion. Degeneration of myocardium could be observed. No granuloma was detected (H&E, $\times 150$).

sions (patient 2 and 4). In total 9 intra-cardiac masses were revealed in the 5 patients. Of these masses, 2 were located in the left ventricle, 3 in the right ventricle, and 4 in the right atrium, indicating a right heart predominance (78%). The lesions varied in size, might be attached to apex, septum or free wall, and could extend to adjacent cavities. Two patients had surgical resection of the cardiac masses. The pathology revealed inflammatory mass and inflammatory cystic change with thrombosis formation, respectively (Fig. 2).

Cardiologic sequel and outcome

Heart failure was not a prominent manifestation, even in patient 2 who had a huge left ventricular mass. However, 2 patients did develop heart failure, including patient 6 who had pericardial masses, pericardial effusion, and cardiac tamponade, and patient 4 who had concomitant congenital hypertrophic

obstructive cardiomyopathy (HOCM). Valvular involvement was not observed. Among the 2 patients who had surgical removal of cardiac mass but without following immunosuppressive therapy, 1 recurred with right atrial masses (patient 2), and 1 had no recurrence of cardiac mass at a short-term follow-up of 5 months (patient 1). All patients were finally treated with corticosteroid, immunosuppressive agents and anticoagulation, and all achieved partial or complete resolution of cardiac masses. Complete resolution of the cardiac masses were achieved at a short-term follow-up of 1 year in half of the patients (patient 3, 5 and 6). No recurrence has occurred while the patients have been under observation.

Discussion

Cardiac manifestations were found in 1–5% of the BD patients in clinical series (27, 28) and in 16.5% of cases in

a Japanese systematic necropsic study (29). They consist of cardiomegaly, endocarditis, pericarditis and less commonly, myocardial infarction and myocarditis (27). Cardiac mass is a rare entity of cardiac manifestation of BD, and were reported in cases. Our review of literature revealed that most of the cardiac masses of BD were thrombi, with or without underlying myocardio-pathy or endocardiopathy (2–18), and in rare cases, were reported as endomyocardiofibrosis (11, 19–22), endocardial fibroelastosis (23), inflammatory mass (24), and cystic change (25), and these pathological changes may be complicated with thrombosis.

Intra-cardiac thrombosis

This is the most frequently reported cause of cardiac masses in BD. Since its first description in 1977 (30), fewer than 40 cases have been reported (2–18). It has been noticed that cardiac thrombi of BD has prominent right heart predominance. Left ventricular thrombi were reported in 3 cases (19, 31, 32) and left atrial thrombus was found only in 1 case (33). Right heart thrombi may predispose the patient to pulmonary embolic events. A systemic review by Mogulkoc *et al.* revealed that up to one-third of BD patients with intra-cardiac thrombi have pulmonary infarction (2). The pathological bases of intra-cardiac thrombosis might be underlying myocarditis, endocarditis, endocardial fibrosis, or pancarditis. However, thrombi were found on normal endocardium in some cases (2).

Cardiac inflammatory mass

This was a less common manifestation of cardiac mass in BD, compared to intra-cardiac thrombosis. In our case series, 1 patient (patient 1) was diagnosed with inflammatory mass of the right ventricle according to surgical pathology. Lee *et al.* reported 4 cases of cardiac inflammatory mass in BD (24). All the masses were located in the right side of the heart, mostly in the right ventricle. The pathology showed endocardial fibrin deposition, dense inflammatory infiltration, neovascularisation, and fibrosis. Acute endothelialitis was also present in the newly formed vessels.

Table IV. Cardiologic sequel and outcome*.

Pt. no.	Valve Involvement	Heart Failure	Treatment	Outcome
1	–	–	Surgery + warfarin	No recurrence at 5 months, no recurrence on IST at 15 months
2	–	–	LV mass: surgery + aspirin	Recurrence with right heart masses at 2 months
	–	–	RA mass: warfarin + prednisone + CTX	Partial resolution at 1 year
3	–	–	Prednisone + CTX + warfarin	Partial resolution at 6 months, complete resolution at 1 year
4	–	+	Prednisone + CTX + warfarin	Partial resolution at 3 months, symptom free at 5 years
5	–	–	Prednisone + CTX + aspirin + warfarin	Complete resolution at 6 months, no recurrence at 2 years
6	–	+	Prednisone + CTX + warfarin	Complete resolution at 6 months, no recurrence at 2 years

*CTX: cyclophosphamide; IST: immunosuppressive therapy; LV: left ventricle; MTX: methotrexate; MV: mitral valve; RA: right atrium.

Though rare, cardiac inflammatory mass might not be that uncommon as reported. Most of the intra-cardiac masses were reported as thrombi when no pathology was available, or when concomitant thrombosis was present. Moreover, since the treatment usually included both corticosteroids and anticoagulation, it is difficult to determine which one played a chief role in the remission of the cardiac masses. So the incidence of cardiac inflammatory mass in BD might be underestimated.

Intracardiac cystic change

This was found in 1 patient (patient 2) in our series. Steward *et al.* reported another case of inflammatory cyst formation in the right atrium that was filled with an acute inflammatory exudates and fibrin, which was the first and only reported case of cardiac cystic change of BD (25). Both patients were not complicated with pulmonary infarction. The incidence of cystic change might also be underestimated, for the same reason as that of cardiac inflammatory mass in BD.

Endomyocardiofibrosis

Endomyocardiofibrosis is another entity of cardiac mass in BD (11, 19–22), though very rare. In pathology, endomyocardiofibrosis appeared as a dense fibrous tissue with neovessels and various degree of endocardial inflammation

consisting of granulocytes and mononuclear cells infiltrate that could extend to the myocardium. The right ventricle is the most affected chamber, followed by the left. Almost all were associated with thrombosis formation, as well as valvular dysfunction, which were related to extension of endomyocardial fibrosis to leaflets and papillary muscles. Heart failure was common, which might be due to concomitant valvular disease. Endomyocardiofibrosis was considered to be a sequelae of endomyocarditis, endocarditis, and thrombosis (19):

Endocardial fibroelastosis (EFE)

This is a condition characterised by the thickening of the endocardium due to the proliferation of fibrous and elastic tissue. It is most commonly seen in young children and rarely in adults, and is often associated with congenital heart anomalies, infection, or gene mutation. One case of EFE was reported with BD (23). The immunological process causing endocardial injury was considered to be potential cause of EFE.

Echocardiographic appearances of cardiac mass of BD were easily confused with cardiac tumour. The patient history should be carefully reviewed to avoid unnecessary surgery. Rarely could BD coexist with cardiac tumour (34), and further pathological examination is needed for diagnosis in such cases. The clinical presentation of cardiac inflam-

matory mass in BD could be confounded with infectious endocarditis, especially when there is coexisting fever. However, in most patients, the pathologic features of the cardiac BD suggested a chronic progressive disease, with coexisting acute and chronic inflammation, while infective endocarditis consists of acute or subacute inflammation. The presence of heavy neutrophilic infiltration and/or microabscess with negative blood culture also suggest the possibility of cardiac BD. Characteristic endothelial loss with fibrin deposition is also helpful for the differential diagnosis (24).

As in our case series and as reported in literature, there is a clear right heart predominance of cardiac masses in BD, the cause of which is unclear. However, the relatively low pressure in the right heart might facilitate the growth of right heart mass.

In our case series, 5 of the 6 patients were complicated with pulmonary infarction. It was considered that because of the sticking nature of the cardiac thrombi in BD, the pulmonary infarctions were probably caused by in-situ thrombi, rather than embolisms from the cardiac masses (35). However, considering the varying nature of the cardiac mass lesions in BD, there are still propensities for the right heart masses to break down and cause pulmonary embolisation in certain patients. For example, the inflammatory right heart mass we observed in patient 1 was jelly-like and very easy to break down, and might have caused pulmonary infarction in the patient.

Heart failure is not a prominent complication of cardiac mass of BD in our series. However, this might depend on the pathological nature of the cardiac mass. It seems that patients with endomyocardiofibrosis were more predisposed to heart failure, which might be attributed to the intense valvular involvement in this situation.

All the patients had excellent outcome with corticosteroids, immunosuppressive agents, and anticoagulation. Surgical removal was not preferred because of the high rate of surgical complications during active disease. However, surgery should be considered when the cardiac mass is complicated with heart failure

or valvular disease, and concomitant immunosuppressive therapy may reduce surgical complications (2, 19, 24). Anticoagulant therapy is not recommended in BD patients, for the high haemorrhagic risk and small benefit combined with anticoagulant therapy in these patients (35). However, in our series, anticoagulation was well-tolerated in all patients, and there was no haemorrhagic complication. Considering the severity of cardiac mass, we recommend anticoagulation in these patients, but arterial aneurysm should be excluded before starting anticoagulation in order to reduce the risk of haemorrhage.

Conclusion

Intra-cardiac thrombi account for the majority of intra-cardiac masses of BD. However, endomyocardiofibrosis, endocardial fibroelastosis, inflammatory mass and cystic change are other entities of cardiac mass lesion in BD, and the incidence might have been under estimated. Right heart masses are more common, and might predispose patients to pulmonary embolism. Heart failure is more prominent when with valvular involvement. Most patients achieve good response with corticosteroids and anticoagulant therapy. For patients with valvular disease, heart failure, or suspected neoplasm, surgery should be considered.

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