A huge intramyocardial cystic-like dissection with left ventricle outflow tract obstruction in Behcet’s heart disease

Wai Yen Yim1,*, MD, Yafei Hu2,*, MD, Bing Yang2, MD, Xinyao Cheng1, MD, PhD

1Echocardiography Laboratory, Zhongnan Hospital, Wuhan University, Wuhan, 2Medical Ultrasonic Diagnostic Department, Enshi Center Hospital, Enshi City, Hubei, China

*Both authors contributed equally as first authors in this manuscript.

Correspondence: Dr Xinyao Cheng, Echocardiography Laboratory, Zhongnan Hospital, Wuhan University, 169 Donghu Road, Wuhan 430071, China. xycheng@whu.edu.cn

Singapore Med J 2021, 1–6
https://doi.org/10.11622/smedj.2021079
Published ahead of print: 18 June 2021

Online version can be found at
http://www.smj.org.sg/online-first
Dear Sir,

Behcet’s disease (BD) was originally reported in 1937 by Hulusi Behcet, Professor of Dermatology at the University of Istanbul (1889–1948). It was first described as a triple symptom complex involving the mouth, genitalia and eyes. Multisystemic clinical manifestations of BD have since been reported. When the heart is involved, BD is referred to as Behcet’s heart disease. Davatchi et al reported that only 0.5% of patients with BD in Iran, 4% of patients in China, 3.2% in Germany and 3.1% in Morocco showed cardiac manifestations.\(^1\) It has been reported that all cardiac tissues could be affected in BD; however, the interventricular septum is rarely involved.\(^2\) Herein, we present a case of Behcet’s heart disease with destruction of the interventricular septum (IVS) and multiple compound lesions of the heart.

A 55-year-old Chinese male farmer was admitted with complaints of recurrent fever, syncope and chest discomfort for two years, which had worsened and had been accompanied with dyspnoea for the past four months. He had a permanent pacemaker implanted at another hospital two years ago owing to a third-degree atroventricular (AV) conduction block. He had a 30-year history of BD, which was diagnosed based on his recurrent oral aphthous ulceration, genital aphthous and positive pathergy test.\(^3\) He had superficial gastritis for four years and a ten-year history of smoking and alcohol consumption. Physical examination showed a blood pressure of 105/50 mmHg, body temperature of 36.5°C, heart rate of 74 beats per minute and respiratory rate of 16 times per minute. A rough systolic murmur and a decrescendo diastolic murmur were heard over the precordial area. Laboratory examination revealed a white blood cell count of \(5.8 \times 10^9/L\) (normal range 4.0–10.0 \(\times 10^9/L\)), red blood cell count of \(4.02 \times 10^{12}/L\) (normal range 4.3–5.8 \(\times 10^{12}/L\)), haemoglobin 109.0 (normal range 130–175) g/L, platelet count 190 \(\times 10^9/L\) (normal range 125–350 \(\times 10^9/L\)), haematocrit 34.0% (normal range 40%–
50%). Other biochemical test results showed an erythrocyte sedimentation rate of 23 (normal range 0–15) mm/h, C-reactive protein (CRP) 10.4 (normal range 5–10) mg/L, high-sensitive CRP 15.29 (normal range 0–8.2) mg/L and interleukin-6 6.21 (normal < 7) pg/mL. Repeated blood cultures showed negative results. Transthoracic echocardiography (Fig. 1) revealed a subaortic interventricular septal dissection that appeared as a huge cystic mass projecting to the left ventricular outflow tract (LVOT), which led to a local obstruction. A pathological orifice was observed between the base of the right aortic sinus and the cystic dissection, with blood flow signals passing to and fro through the ruptured orifice during cardiac cycle. The large intramyocardial interventricular septum appeared to be ruptured across the basal and the middle segment where the anterior papillary muscle arises. A strong irregular echogenicity was noted, indicating partial thrombosis inside the cystic mass. Severe aortic, mitral and tricuspid regurgitations coexisted. Both atria and the left ventricle were enlarged. CT angiography (not shown) showed signs of enlarged heart and bilateral pulmonary obsolete tuberculosis-like changes. Based on the medical history and clinical presentation of the patient, he was diagnosed as having Behcet’s heart disease. However, the patient denied surgery owing to the surgical risk and complications involved. He was discharged after his clinical manifestations improved following administration of diuretics and inotropics combined with immunosuppressants.

IVS dissection is a rare abnormality that may result from an aneurysm of one of the aortic sinuses, bacterial endocarditis, surgical therapeutic manipulation, trauma or congenital abnormal myocardial development. It also can occur in patients with ischaemic myocardial infarction or BD. IVS dissection most commonly begins in the right sinus of Valsalva, which ruptures into the basal septal segment. The middle part of the septum is also occasionally involved. The dissection can communicate with the aortic cavity through a perforation at the aortic base or with the left ventricular cavity through the ruptured site of the endocardium. As this abnormality is at the centre of the heart, which is critical to the
underlying normal anatomical structure and functions, the patient usually has a progressive course and poor prognosis.

The current patient was confirmed as having BD based on the International Criteria for Bechet’s Disease.\(^{(13)}\) To the best of our knowledge, this is the first case of BD in which IVS dissection involved the anterior papillary muscle. The patient had a huge intramyocardial dissection covering the field of the basal and middle segments of the septum that was well detected in long-axis, short-axis and apical four-chamber views of transthoracic echocardiography (Fig. 1). The huge cystic-like dissection mass projected into the LVOT and caused local hemodynamic obstruction, with a systolic velocity of 284 cm/s at the aortic orifice. There was a ruptured hole at the base of the RSV, with large amount of blood regurgitating into the cystic-like dissection through the opening during early diastole (Fig. 1). Severe aortic, mitral and tricuspid regurgitations occurred, which were closely related to the large area of destruction at the septum and involved the anterior papillary muscle as well as aortic perforation. Behcet’s heart disease showing AV conduction block has previously been reported.\(^{(14)}\) Our patient had a permanent pacemaker implanted owing to third-degree AV block two years prior to admission. This indicated that the inflammation activity in BD can cause cardiac conduction tissue lesions, which could also be associated with the damaged basal septum. During diagnosis, we found that echocardiography can clearly reveal the cardiac structural changes and hemodynamic derangements, whereas CT angiography could only demonstrate the contour of the enlarged heart. Hence, echocardiography would be the preferred technique to investigate the intracardiac structures.

In conclusion, we encountered a rare case of Behcet’s heart disease. The patient had IVS dissection involving the anterior papillary muscle with left ventricular outflow tract obstruction accompanied by right aortic cusp perforation and multiple valve insufficiency as well as a third-degree AV conduction block. Destruction of the central cardiac structure could
severely affect various cardiac functions; hence, early diagnosis and intervention is extremely important in Behcet’s heart disease.

Yours sincerely,

Wai Yen Yim¹,*, Yafei Hu²,*, Bing Yang², Xinyao Cheng¹

¹Echocardiography Laboratory, Zhongnan Hospital, Wuhan University, Wuhan, ²Medical Ultrasonic Diagnostic Department, Enshi Center Hospital, Enshi City, Hubei, China

*Both authors contributed equally as first authors in this manuscript. xycheng@whu.edu.cn

REFERENCES


FIGURE

Fig. 1 Echocardiographs show the cystic dissection mass projecting to the left ventricle outflow tract (LVOT) and leading to local obstruction. (a) LVOT is 3.8 mm wide at systole in the long-axis view of the left heart; (b) LVOT is 1.3 mm wide at diastole; (c) LVOT is filled with the systolic blood flow signal; (d) AR: aortic regurgitation; (e) CMO: pathological orifice at the right aortic sinus base connecting the aortic cavity and cystic dissection; (f) CMOF: blood signal through the orifice into the dissection mass at early diastole; (g) Short axis view at mitral level; (h) Short axis view at papillary muscles; (i) Four-chamber view shows both mitral and tricuspid moderate regurgitations; and (j) The maximum velocity at the aortic orifice was reached at 284 cm/s. M: cystic dissection mass; MR: mitral regurgitation; AMV: anterior mitral valve; CMO: cystic mass orifice; CMOF: cystic mass orifice flow; PMV: posterior mitral valve; AP: anterior papillary muscles; LV: left ventricle; RV: right ventricle; RA: right atrium; LA: left atrium