CASE REPORT
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Superior Vena Cava Syndrome Without Thrombosis Found in Behcet's Disease

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Behcet's disease is a rare multisystemic disorder whose main pathological defectis vasculitis, and superior vena cava (SVC) syndrome without thrombosis is a very rare manifestation of the disease. These authors encountered a case of SVC syndrome without thrombosis caused by Behcet's disease. A 33-year-old man visited the hospital for aggravated dyspnea without any related medical and familial history. He had a three-day history of abrupt swelling of the face, neck, and right arm. He suffered from recurrent oral ulcer, and there were acneiform nodules on his face as well as redness and swelling at the site of the intravenous injection. On the multi-detected computed tomography (CT) chest angiograms (chest angio MDCT), the SVC narrowed without thrombosis. Venogram was carried out, and percutaneous transluminal balloon angioplasty of the SVC stenotic site was performed. The following day, the swelling was found to have subsided. The details of the case are reported herein.

Key Words: Superior vena cava syndrome, Behcet disease

INTRODUCTION

Behcet's disease is a form of vasculitis that may involve mucocutaneous, ocular, cardiovascular, renal, gastrointestinal, pulmonary, urologic, and central nervous systems, as well as the joints, blood vessels, and lungs.

There is no specific pathological diagnostic test for Behcet's disease at present. However, accordingto the International Study Group Guideline,[1] Behcet's disease must have oral aphthous ulcer (any shape, size or number increased at least 3 times in any 12 months), along with 2 out of the next 4 "hallmark" symptoms; genital ulcer (including anal ulcers and spots in the genital region and swollen testicles or epididymitis in men), skin lesion (papulo-pustules, folliculitis, erythema nodosum, acne in post-adolescents regardless of corticosteroids use), eye inflammation (iritis, uveitis, retinal vasculitis, cells in the vitreous), pathergy reaction (papule >2 mm diameters at 24-48 hours or more after needle-prick).

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The main pathologic defect of Behcet's disease is vasculitis such as arterial occlusion, venous occlusions, aneurysms, and varices. Histologic findings include media thickening, elastic fiber splitting, and perivascular round cell infiltration. Venous involvement is usually limited to occlusion, with the varices rarely affected. The affected sites of the venous system are the SVC, inferior vena cava, deep femoral vein, and subclavian vein.

SVC syndrome may be caused by invasion or compression by a pathological process or by thrombosis in the vein itself. Approximately 90% of cases are associated with a malignant tumor that is compressing the SVC. Other cases have a variety of causes, including infectious and catheter-related etiologies and followed by vasculitic syndromes, like Behcet's disease.

We report a case of SVC syndrome without evidence of thrombosis found in Behcet's disease.

CASE

A 33-year-old man visited hospital for aggravating dyspnea without any related medical history and familial history. He had a 3-day history of abrupt swelling of face, neck, and right arm. He experienced right arm swelling several times

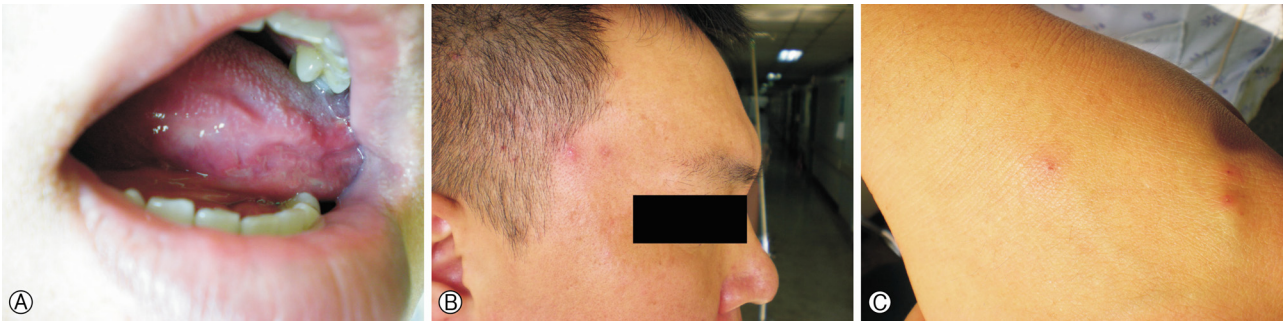

Fig. 1. (A) Oral aphthous ulcer observed on the patient's lower lip. (B) Acneiform nodules observed on the patient's face. (C) Redness and swelling observed at the site of the intravenous injection (positive patergyreaction).

after exercise since last year. He suffered from recurrent oral ulcer over 3 times for a year. We observed oral aphthous ulceration on his lower lip (Fig. 1A), acneiform nodules on his face (Fig. 1B) and redness and swelling at the site of intravenous injection (Fig. 1C) (positive pathergy reaction). His laboratory findings are within normal range; erythrocyte sedimentation rate (ESR) 26 (N: 1-10) mm/hr, high sensitivity c-reactive protein (HS-CRP) 0.88 (N: 0-0.15) mg/dL, anti-nuclear antibody (ANA) titer (-), venereal disease research laboratory (VDRL) test (-), human leukocyte antigens (HLA) B51 (-), anti neutrophil cytoplasmic antibody (ANCA) (-), lupus anticoagulant antibody (LAA) (-), anti-cardiolipin antibody (ACA) (-), Tumor marker [carcinoembryonic antigen (CEA), α-fetoprotein (AFP), carbohydrate antigen 19-9 (CA19-9)].

Under the suspicion of Behcet's disease with SVC thrombosis, chest angio MDCT was performed and revealed focal stenosis of SVC, the site joined by innominate veins, without thrombosis (Fig. 2). We carried out angiography for SVC, which revealed severe luminal narrowing (90%) of SVC. Balloon angioplasty for SVC performed immediately after angiography (Fig. 3). we used 8 mm/4 cm balloon because in the event of a medical emergency with sudden onset of symptoms. we wanted to apply immunosuppressant therapy with cyclophosphamide, but the patient refused immunosuppressant therapy for repulsion.

After 2 months, his right arm and face swelling was recurred. We carried out right subclavian venography and it shows severe (90%) stenosis of SVC. We choose prepared large (18 mm/4 cm) balloon. After balloon angioplasty, it shows residual stenosis of 15% (Fig. 4).

We have applied immunosuppressant therapy with cyclophosphamide after percutaneous transluminal angioplasty and kept going close observation. After angioplasty for SVC, his symptoms was gone. During 5 days, we monitored change of symptoms closely, but he had not any complication.

After 1 year later, Although he did not complain any dyspnea or arm and neck swelling, we scanned chest angio MDCT for follow up and it revealed well-sustained for SVC (Fig. 5).

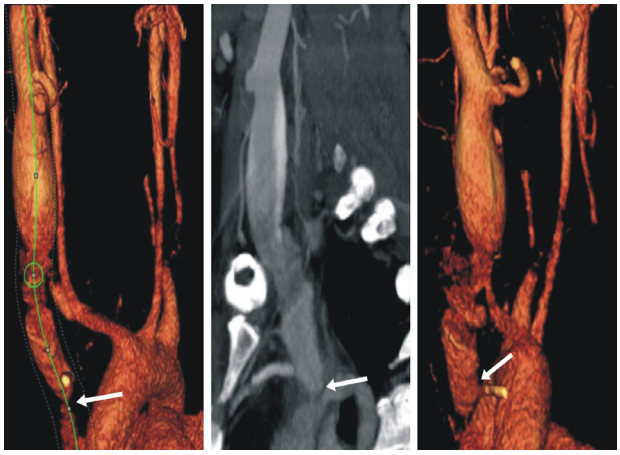

Fig. 2. Angio-CT images reveal focal stenosis of the SVC (arrow), the site joined by innominate veins, without thrombosis.

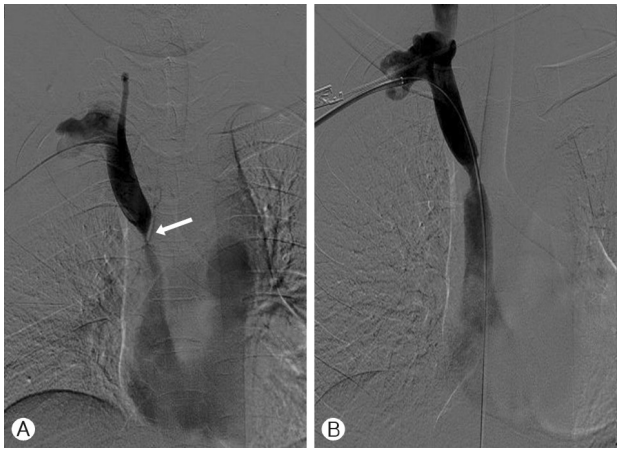

Fig. 3. Angiography findings (initial). (**A**) Angiography shows a focal narrowing of SVC (arrow). (**B**) Dialtation of stenotic segment is noted on post-angioplasty image using an 8 mm/4 cm balloon.

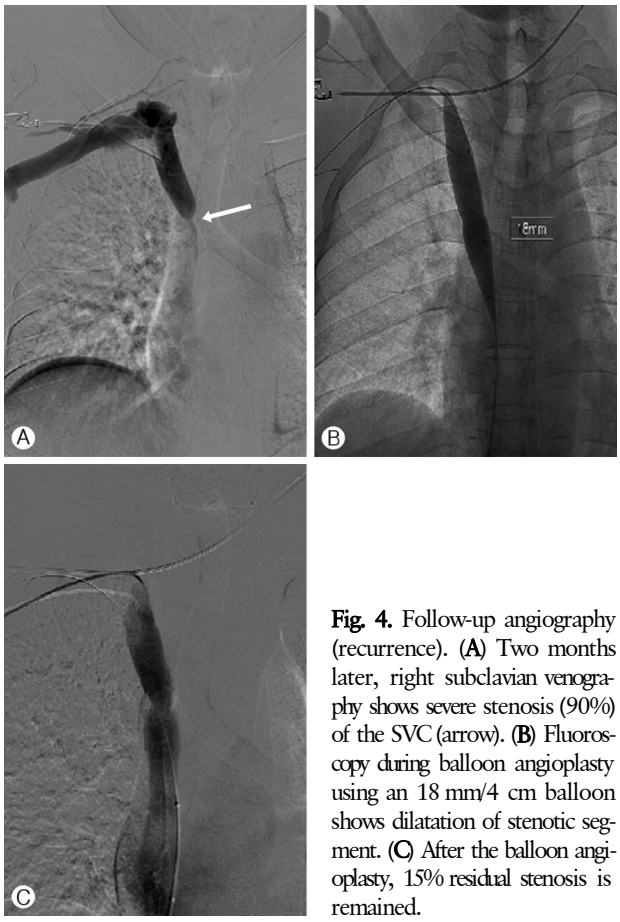

Fig. 4. Follow-up angiography (recurrence). (**A**) Two months later, right subclavian venography shows severe stenosis (90%) of the SVC (arrow). (**B**) Fluoroscopy during balloon angioplasty using an 18 mm/4 cm balloon shows dilatation of stenotic segment. (**C**) After the balloon angioplasty, 15% residual stenosis is remained.

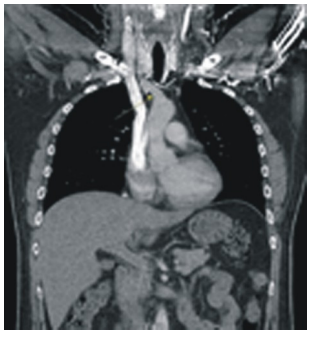

Fig. 5. CT image at follow-up. 1 year later, the chest angio MDCT showed a well-sustained SVC (arrow).

DISCUSSION

SVC syndrome is a devastating complication of obstructive lesions compromising the SVC and its branches.[7] SVC syndrome is a relatively common complication of lung cancer or lymphoma, and may appear as initial manifestation of these diseases. However, benign causes also exist, and physicians should not automatically assume that SVC syndrome is due to cancer.[2] As a manifestation of Behcet's disease, SVC syndrome is rare and generally develops after diagnosis of the disease.[3]

The main pathologic defect of Behcet's disease is vasculitis that is an inflammatory disorderof blood vessels leading to compromised lumen, which ultimately leads to ischemia of the region involved.[4] A leading cause of death in Behcet's disease is vasculitis with an approximate prevalence of 25%, and isseen more frequently in male than in female.[5]

We would like to emphasize that this case implies the critical importance of history taking and physical examination. Although SVC stenosis was suspected because of suddenly developing swelling in the right arm and facialedema and subsequently SVC stenosis was diagnosed with chest angio MDCT, detailed history taking and physical examination enabled us to diagnose this case as Behcet's disease in accordance with the diagnostic criteria of International Study Group for Behcet's disease.

Any treatments for SVC syndrome must be tailored to the underlying etiology and work quickly enough to relieve its severity or prevent life-threatening conditions.[6] Since SVC stenosis can be regarded as a medical emergency, SVC stenosis without thrombosis was diagnosed based upon the findingson venograms, followed by performing immediately balloon angioplasty. As a result, the patient's symptoms couldbe improved dramatically.

In modern medicine, with increasing acceptance of percutaneous and minimally invasive therapies, treatment of SVC syndrome with balloon dilatation and stenting has also become more popular.[8] Nevertheless, as we failed to obtain any appropriate large stents or balloons for SVC dilatation in the event of a medical emergency with sudden onset of symptoms, the small (8 mm/4 cm) balloon angioplasty only was available at that time. At follow-ups, it was observed that the relevant symptoms appears in the patient, we choose prepared large (18 mm/4 cm) balloon and immunosuppressant therapy because it is well-known that stenting of vein can cause instent restenosis.

We report our experience in SVC syndrome without thrombosis caused by Behcet's disease which has been treated by percutaneous transluminal angioplasty. When examining patients with SVC stenosis, it is required to take account of various diseases like Behcet's disease and to make a prompt decision on interventional therapies such as balloon angioplasty or stent insertion that it should be regarded as a medical emergency.

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