

A Case of Behcet's Syndrome with Superior Vena Cava Syndrome

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Behcet's syndrome is a multi-systemic and chronic disorder that affects many organs. It has been suggested that the diagnosis was based on the presence of the 'major' and 'minor' clinical criteria. When thrombophlebitis, arthritis, central nervous system or gastrointestinal lesions are also present, Behcet's syndrome will be thought to be present in the appropriate geographic area.

We report a case of superior vena cava syndrome caused by Behcet's disease in a 40-year-old man with recurrent oral aphthous ulcers and skin rashes on the anterior chest wall. There were multiple thrombosis of the superior vena cava, innominate and subclavian veins. This patient also had a solitary cecal ulcer with an ileocecal fistula and downhill varix. The chest CT, veno-cavography, pulmonary angiography and colon study were taken and follow-up was performed.

Key word : *Behcet's disease, SVC syndrome, Ileocecal ulcer, Down-hill esophageal varix*

INTRODUCTION

Behcet's syndrome was described as a clinical syndrome with aphthous oral ulcers, genital ulceration and eye involvement. As it affects many other organs, Behcet's syndrome is now recognized as a systemic disease¹⁾. The clinical characteristics of Behcet's syndrome are thought to be caused by vasculitis that principally affects veins, venules and capillaries. But there were no pathognomic findings and the diagnosis was based on the presence of the 'major' and 'minor' criteria, e.g. oral ulcers, genital ulcers, ocular lesions, skin lesions, arthropathies, gastrointestinal lesions and cardiovascular lesions²⁾. Although there were no definite symptoms of major criteria, thrombophlebitis, arthritis, central nervous system or gastrointestinal lesions are also present and Behcet's syndrome is thought to be present in the appropriate geographic area³⁾.

We report a patient with Behcet's syndrome having recurrent oral ulceration, cutaneous pustules, as well as

non-deforming arthritis and ileocecal fistula presenting SVC syndrome and down-hill esophageal varix.

CASE REPORT

In November 1995, a 40-year-old man presented with a four-month history of intermittent lower abdominal pain and neck vein engorgement. He had had recurrent oral ulcers for 7 years. His face was plethoric and the external jugular vein was markedly engorged. There were multiple macuberythematosus lesions, about 2-3mm in diameter, on his face, neck and anterior chest. There were distended collateral vessels on the anterior chest wall and abdomen. Chest radiograph revealed a slightly widened superior mediastinum, suggesting a pathologic problem of the superior mediastinum (Fig. 1). Initial computed tomography (CT) scan, following a bolus injection of intravenous contrast medium, demonstrated obstruction of the superior vena cava with thrombosis and pleural effusion of the right hemithorax (Fig. 2). Abdominal sonogram did not show any sign of obstruction of the inferior vena cava or portal vein. Gastroscopy revealed a mild degree of esophageal varix (Form 1, white color, Locus superior, negative red color sign). Pleural tapping was not taken because the patient did not desire admission or further

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ased. He had painful swelling of the right knee joint. The pathergy skin test was negative. Hematological and biochemical tests were as follows: white blood cell count was 7,200/mm³, hemoglobin 12.6g/dL, platelet 320,000/mm³. Erythrocyte sedimentation rate was 39mm/hr and C-reactive protein was negative. The prothrombin time and activated partial thromboplastin time were normal. The rheumatoid factors, antinuclear antibody, antinuclear DNA, anticardiolipin antibody, lupus anticoagulant, antiphospholipid antibody and antineutrophil cytoplasmic auto-antibody were negative. There was no proteinuria and hematuria. Protein C, protein S, factor VIII, fibrinogen and antithrombin III were all normal. Follow-up CT scan demonstrated decreased pleural effusion, but newly developed consolidation at the left lower lobe territory, 3 × 1.5 × 3cm size in diameter, suggested focal pulmonary infarction(Fig. 3). SVC obstruction and many collateral

Fig. 1. Chest radiography shows slightly widened superior mediastinum and laterally shifted dome of the right hemidiaphragm.

Fig. 2. Initial contrast enhanced CT scan shows filling defect in the superior vena cava suggesting thrombosis (long arrow) and rich collateral veins in the mediastinum (arrow head) and chest wall. Pleural effusion is seen in the right hemithorax.

diagnostic studies to be done.

In February 1996, he was admitted to the hospital. In the ensuing twelve weeks, he felt less plethoric and had mildly collapsed neck vein engorgement. On examination, he had several painful aphthous oral ulcers, 2-8mm in diameter, with a central yellowish necrotic base on the tongue. Neck vein engorgement was somewhat decre-

Fig. 3. Follow-up CT after 3 months from initial CT scan shows newly developed consolidation in the left posterior basal segment (long arrow). The pleural effusion in the right hemithorax is decreased.

vesels still persisted. Venocavogram was obtained after injection of 40ml contrast materials (Ultravist, Schering, Germany) through both basilic veins. The venocavogram showed complete occlusion of the SVC, right subclavian vein and partial occlusion of the left subclavian vein. Subsequent pulmonary angiography was normal. The colon study and colonoscopy revealed solitary cecal ulcer with an irregular mucosal change of the ileocecal area and ileocecal fistula (Fig. 5).

A final diagnosis of Behcet's syndrome with superior vena cava syndrome and ileocecal fistula was made and we started medical treatment with high dose sulfasalazine

Fig. 4. Superior veno-cavogram shows complete occlusion of both superior subclavian veins(long arrow) and rich collateral veins in the chest wall and mediastinum. The thoracic duct and azygose and hemiazygose veins are opacified by reverse flow of contrast medium toward the subphrenic veins.

Fig. 5. Double contrast colon study shows ileocecal fistula and irregular shaped cecal ulceration.

and azathioprine. He showed mild decompression of neck vein engorgement and disappearance of right abdominal pain, but still plethoric.

DISCUSSION

Behcet's disease is a multi-systemic disorder having orogenital ulcers, uveitis, cutaneous, arthritic, neurological,

vascular and intestinal manifestations. This syndrome affects mainly men aged 20-30 years. Because Behcet's disease has various clinical manifestations, it is difficult to diagnose^{1, 2)}. Diagnosis of Behcet's disease is based only on clinical findings, because there are no pathognomonic laboratory or histopathological diagnostic criteria for this syndrome. The histological hallmark of Behcet's disease is a nonspecific vasculitis of the arteries, veins, arterioles, venules, and capillaries of both systemic circulations⁴⁾.

The pulmonary vasculitis characteristic of Behcet's syndrome is usually necrotizing, and histopathologic findings indicate perivascular infiltration by lymphocytes. The syndrome characteristically affects the large vessels(mainly the veins such as SVC and innominate vein)^{5, 6)}. In this case, obstruction of the SVC and subclavian veins were confirmed by the venocavogram. Right pleural effusion, which was not confirmed by pleural tapping, was thought to be transudate resulting from increased high venous pressure due to obstruction of the azygose vein and vena cava. Downhill esophageal varix with no evidence of portal hypertension is thought to arise by the same mechanism. Air space consolidation, which was seen on the CT scan, was thought to be an infarction or infiltration. But this was not identified by angiography. The previous post-mortem study reported that the pulmonary infarct, with no evidence of peripheral venous thrombosis, resulted from an in situ thrombosis rather than an embolism. In situ venous thrombosis secondary to vasculitis probably plays an important role in the development of the major vessel venous thrombosis seen in Behcet's syndrome⁵⁾.

The gastrointestinal manifestation of this syndrome resembles that in Crohn's disease, but granulomas are not found and free perforation is common⁷⁾. Microscopically, vasculitis, often of the leucocytoclastic variety, is a common multi-systemic nature of Behcet's disease usually allowing its recognition. In this case, there was a fistula between the cecum and terminal ileum. Endoscopic biopsy at the cecum shows massive infiltration of inflammatory cells without granuloma and crypt abscess.

There were some case reports about Behcet's syndrome with SVC syndrome in Korea⁸⁾, but there was no evidence of gastrointestinal involvement. This case has typical solitary ulcer with ileal fistula near the ileocecal valve. Also, there was downhill inverted superior esophageal varix. The downhill superior esophageal varix had been described in thoracic goiter, lung cancer, mediastinal fibrosis, tumors, superior vena cava ligation,

subclavian vein thrombosis and congestive heart failure. There were few cases of downhill superior esophageal varix due to Behcet's disease in the literature⁹⁾.

Treatment of Behcet's syndrome is symptomatic and empirical. Mucous membrane involvement may respond to topical corticosteroids. The arthritis responds to rest and analgesics. Thrombophlebitis is treated with antiplatelet agents. But uveitis, central nervous system involvement and pulmonary disease usually require corticosteroid and immunosuppressives. Other medications which have been tried include azathioprine, chlorambucil, colchicine, levamisole, cyclosporine and fibrinolytic-enhancing drugs, such as stanozolol and phenformin¹⁰⁾.

However, there is little data on the use of the drugs specifically for the pulmonary disease of Behcet's syndrome. Therapy for gastrointestinal Behcet's disease is identical to that of inflammatory bowel disease, especially high dose sulfasalazine¹¹⁾.

REFERENCES

1. Conn DL, Hunder GG, O'Duffy JD. *Behcet's disease*. In : Kelley WN, Harris ED, Ruddy S, Sledge CB. eds. *Textbook of Rheumatology*. 4th ed. Philadelphia :WB Saunders Co. 1993 ; 1097- 1099.
2. International study group for Behcet's disease: *criteria for diagnosis of Behcet's disease Lancet* 1990; 335:1078-1080.
3. Morita H, Kawai S, Kobori O, Ohya N, Sumiya M, Dohi T. *Incomplete form of Behcet's colitis in Japan: Is it a distinct entity. Am J Gastroenterol* 1995; 90 :523-524
4. Moutsopoulos HM. Behcet's syndrome. In : Iselbacher KJ, Braunwald E, Wilson JD, Martin JB, Fauci AS, Kasper DL. eds. *Harrison's Principles of Internal Medicine*. 13th ed. New York : McGraw-Hill Inc. 1994; 1669-1670.
5. Tunaci A, Berkmen YM, Gokmen E. *Thoracic involvement in Behcet's disease: Pathologic, clinical and imaging features. ARJ* 1995; 164 :51-56.
6. Ahn JM, Im JG, Ryoo JW, Kim SJ, Do YS, Choi YW, Oh YW, Yeon KM, Han MC. *Thoracic manifestation of Behcet syndrome: radiographic and CT findings in nine patients. Radiology* 1995; 194:199-203.
7. Kasahara Y, Tanaka S, Nishino M, Umemura H, Shiraha S, Kuyama T. *Intestinal involvement in Behcet's disease: Review of 136 surgical cases in the Japanese literature Dis Colon Rectum* 1981; 24:103- 106.
8. Hee Won Min, Seung Yun Cho, Sang Il Chun, Dong Sik Bang. *A case of Behcet's syndrome with the superior vena cava syndrome. Kor J Int Med* 1986: 30:401-406(in Korean)
9. Ghayad E, Ghayad A. *Inverted esophageal varices with superior vena cava obstruction secondary to Behcet's disease. In; O'Duffy JD, Kokmen E. eds. Behcet's disease. Basic and clinical aspects. 1st. New York, Marcel Dekker Inc, 1993; 2 15-2 16.*
10. Yazici H, Pazarli H, Barnes CG, Tuzun Y, Ozyazgan Y, Silman A, Serdaroglu S, Oguz V, Yurdakul S, Lovatt GE, Yazici B, Somani S, Muftuoglu A. *A controlled trial of azathioprine in Behcet's syndrome. N Engl J Med* 1990; 322:281-285.
11. Matsukawa M. *Clinical Course and treatment for the intestinal Behcet and so called "simple ulcer". Stomach and Intestine* 1992; 303-311(in Japanese).