

Massive Thrombosis after Central Venous Catheterization in a Patient with Previously Undiagnosed Behcet's Disease

Thrombosis is an important complication of central venous catheterization. Among the many intrinsic and extrinsic factors, the patient's medical disease can play a role in thrombogenesis. Behcet's disease (BD), classified as a vasculitis, is a multisystem disease involving the small blood vessels. It is often difficult to recognize and diagnose the disease. A 24-yr-old female patient showed massive central venous thrombosis which caused superior vena cava syndrome after subclavian vein catheterization. Twenty days after catheterization, the patient exhibited swelling of the face, neck, and both upper extremities. Despite thrombectomy and continuous anticoagulation therapy, her facial and upper extremity swelling reappeared and follow-up chest computed tomography (CT) showed the recurrent thrombosis in the same central veins previously affected. A diagnosis of BD was then made. Following steroid therapy, neither clinical symptoms nor CT findings suggestive of central venous thrombosis were observed during the subsequent 6-months of follow-up period. This case emphasizes that central venous catheterization in a patient with BD should be performed with great caution.

Key Words : *Behcet's Syndrome; Catheterization, Central Venous; Superior Vena Cava Syndrome; Venous Thrombosis*

Seong-Hoon Ko^{1*}, Seung-Kwan Kang¹,
Sang-Kyi Lee^{1*}, He-Sun Song^{1†}

Department of Anesthesiology¹, Research Institute of Clinical Medicine¹ and Institute of Cardiovascular Research², Chonbuk National University Medical School & Hospital, Chonju, Korea

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Address for correspondence

Sang-Kyi Lee, M.D.
Department of Anesthesiology, Chonbuk National University Hospital, 634-18 Keumam-dong, Chonju 561-712, Korea
Tel : +82.63-250-1241, Fax : +82.63-250-1240
E-mail : leesk@moak.chonbuk.ac.kr

INTRODUCTION

Central venous catheterization is an essential practice in the modern-day management of critical illnesses. Thrombosis is one of the most frequent and severe complications thereof. However, thrombotic occlusion of the entire superior central venous system is rare. Behcet's disease (BD) is a chronic, heterogeneous, multisystem disorder caused by vasculitis. We describe a case of thrombotic occlusion of the entire superior venous system after subclavian vein catheterization in a 24-yr-old woman with previously undiagnosed BD.

CASE REPORT

A 24-yr-old woman presented herself with enterocutaneous fistula after a simple closure of spontaneously perforated ileum. She has experienced recurrent multiple oral ulcers for the past 2 yr. Otherwise, she has been in good health and has not been taking any medication on a regular basis. Laboratory tests including complete blood cell count, serum electrolytes, and clotting profiles revealed no abnormalities. Fistulectomy and biopsy of perforated ileum were planned. After induction of general anesthesia, a central venous catheter was placed for postoperative total parenteral

nutrition (TPN). A 16-gauge, single lumen, central venous catheter (Intracath[®]; Arrow Inc, U.S.A.) was inserted into the left subclavian vein by infraclavicular approach. Postoperative chest radiograph showed the catheter tip located at level of the body of fourth thoracic spine. The TPN was continued for 10 days, thereafter the catheter was removed. Four days later, the patient was discharged without any remarkable symptoms.

Six days after discharge, the patient complained of swollen face, neck, and both upper extremities. A clinical diagnosis of superior vena cava (SVC) syndrome was made and a computed tomography (CT) of the chest showed complete obstruction of SVC (Fig. 1). Venous angiography showed extensive thrombosis in the internal jugular, external jugular, innominate, and subclavian veins of left side and SVC with collateral circulation along the thoracic wall into the inferior vena cava (Fig. 2). Because her symptoms rapidly deteriorated and the involved areas were so extensive, surgical intervention and anticoagulation therapy were considered.

By left lateral thoracotomy, thrombectomy of SVC and ligation of left innominate vein were performed. Over the next 2 days, the patient's initial symptoms and signs markedly improved. However, one month later, despite continuous anticoagulation therapy, her facial and upper extremity swelling reappeared and follow-up chest CT showed the recurrent

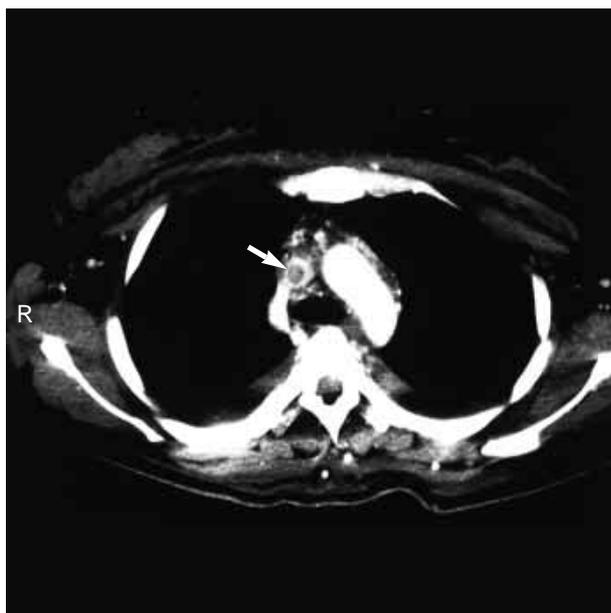


Fig. 1. Enhanced CT scan shows nonenhancing filling defect with superior vena cava representing large thrombus (arrow) with enhancement of the vessel wall.

thrombosis in the same central veins as were affected previously. The abdominal wound showed discharge resulting from recurred enterocutaneous fistula.

To identify the relapsing feature of her vascular and intestinal diseases, she was referred to Department of Internal Medicine for complete medical evaluation. Along with frequent oral ulceration, it was found that she has recently experienced painful erythematous subcutaneous nodules in both lower extremities. Gynecologic examination revealed some tiny ulcers in her labia minor, which had been ignored by the patient, herself. On small bowel series, a moderate-sized, single ulcer was observed and the biopsy findings of the resected ileal tissue were compatible with a systemic inflammatory disease. The Pathergy test, a diagnostic skin test for BD, was positive. A diagnosis of BD was made. Steroid therapy (prednisolone 50 mg daily) was started and the anticoagulation therapy was continued. Over the next 2 months, the patient's symptoms of central venous thrombosis markedly improved and the enterocutaneous fistula resolved completely. Neither clinical symptoms nor CT findings suggestive of central venous thrombosis were observed during the subsequent 6-months of follow-up period.

DISCUSSION

Central venous catheterization plays a major role in critical care medicine. Catheter-related thrombosis (i.e., the presence of a sleeve of fibrin around a catheter or thrombus adherent to a vessel wall) is not uncommon after central venous

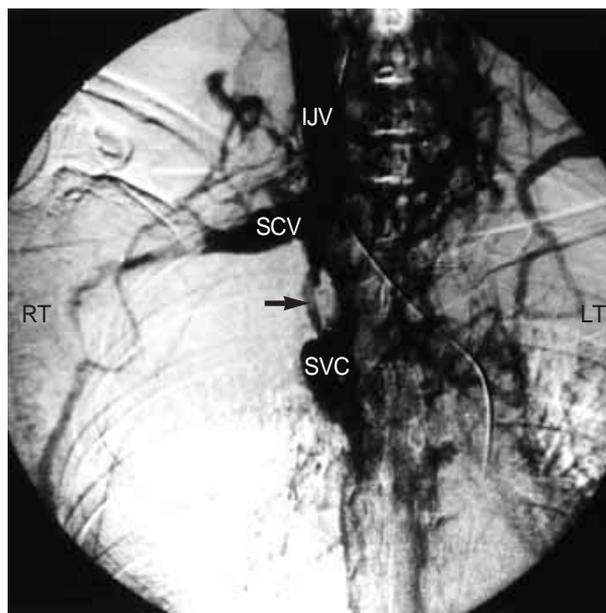


Fig. 2. This venogram shows poor visualization of the large central veins (internal jugular, external jugular, innominate, and subclavian vein of left side) with a large filling defect within the superior vena cava by the thrombus (arrow). The collateral veins were developed along the thoracic wall.

catheterization. The reported incidence of a "fibrin sleeve" ranges from 42% to 100% (1-3). The formation, growth, and dissolution of venous thrombi represent a balance between various thrombogenic stimuli and several protective mechanisms. Factors contributing to the development of thrombosis are duration of catheterization, physical properties of the catheter, and nature of infused fluid. Factors predisposing to extension of thrombus are infection, intrinsic abnormalities of coagulation, and relative intravascular stasis (4). However, despite the apparent high incidence and many predisposing factors of local thrombosis around catheter tips, symptomatic massive extension of thrombus as in this patient is very rare.

BD is a systemic inflammatory illness characterized by exacerbations and remissions of unpredictable duration. The clinical manifestations of BD are recurrent oral and genital ulcers, uveitis, skin lesions, arthritis, thrombophlebitis, and various neurological syndromes (5). The common histopathological process in all clinical manifestations appears to be a vasculitis. The diagnosis of BD is based only on clinical grounds as there are no pathognomonic laboratory or histopathological features. The clinical symptoms and laboratory finding of this patient (recurrent oral ulceration, erythema nodosum-like skin lesions, and positive Pathergy test) satisfied the Diagnostic Criteria for BD Proposed by the International Study Group for BD (6).

Even though thrombosis is a minor component of BD, Kabbaj et al. (7) reported spontaneous venous thrombosis in the patients with BD. It is not well known which mecha-

nism of coagulopathy is involved in thrombogenicity in BD. One study (8) showed that there is no specific coagulation and fibrinolytic changes in the patients with BD. However, multisystemic inflammatory reaction of BD may contribute to the formation and extension of thrombosis. Although there has been no report about catheter-related thrombosis in BD, we can assume that BD aggravated the thrombosis in this patient. The bases for this assumption are as follows. Firstly, we strictly kept to the general rules for central venous catheterization such as aseptic cannulation and dressing. Secondly, the thrombosis was quite massive and extensive in its nature, which is rare among the cases of the usual catheter-related thrombosis. Thirdly, she was never given the hyperosmolar TPN solution that is one of the contributing factors to the development of thrombosis. Fourthly, the thrombosis recurred in a short time in spite of thrombectomy and systemic anticoagulation therapy. Lastly and most decisively, the repetitive thrombosis was completely resolved after a short administration of steroid. Because spontaneous thrombosis is one of the components of BD and the patient's symptoms manifested 10 days after catheter withdrawal, one may think that the thrombosis in this patient was not related to central venous catheterization. However, the thrombosis occurred only in the superior venous system of the left side that was the catheterization site. The right side had no thrombosis on the venography. When the catheter is inserted, a clot may form at the puncture site and also close to the catheter tip, and the entire catheter length may become sheathed in a fibrin sleeve. The thrombus may grow, and clinical symptoms can result from occlusion of the vessel by the thrombi. Therefore, clinical symptoms result from total occlusion of the superior venous system occurred 10 days after catheter withdrawal in this patient.

Anticoagulation therapy and thrombectomy have been the mainstay of therapy preventing clot propagation and relieving the symptoms in a patient with catheter-related thrombosis. Because the exact pathophysiology of BD is still unknown, treatment is aimed to individual symptoms as they occur. Medication for thrombosis of BD is given to

reduce inflammation or to regulate the immune system because the thrombosis does not respond to usual anticoagulation or thrombectomy. This patient responded well to the combination of anticoagulation and steroid therapy.

This case showed that central venous catheterization may result in severe thrombosis in a patient with systemic vasculitis such as BD, emphasizing the need for close evaluation in patients managed with central venous catheter. We suggest that central vein catheterization in a patient with BD should be used with great caution.

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