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Central Venous Port-A Catheterization-induced Superior Vena Cava Syndrome in a Patient with an Adenocarcinoma of the Rectum

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The superior vena cava (SVC) syndrome is an emergency and potentially fatal event. Malignancy is the most common etiology of the syndrome and catheter-induced thrombotic SVC syndrome is rare. We report a 45-year-old woman who presented with SVC syndrome induced by a Port-A catheter. She had a history of adenocarcinoma of the rectum with resection of the tumor and had completed adjuvant chemotherapy via a Port-A catheter 7 months previously. Computed tomography of the patient's chest revealed a low-attenuation thrombus around the Port-A catheter in the SVC and left brachiocephalic vein. The clinical symptoms improved gradually after sequential management, including anticoagulant therapy, thrombolytic therapy and removal of the catheter.

Key Words: Superior vena cava syndrome, thrombosis, thrombolytic therapy

INTRODUCTION

Superior vena cava (SVC) syndrome is an emergency and potentially fatal condition that is caused by obstruction of the SVC and severe impairment of venous return from the head, neck and upper extremities. Malignancy is the most common cause of the syndrome. The incidence of SVC syndrome arising from benign etiologies such as intravascular devices is rare. We report a patient who presented with SVC syndrome caused by thrombosis related to a Port-A catheter placement, which resolved after emergency therapy.

CASE REPORT

A 45-year-old woman had a history of adenocarcinoma of the rectum, (T3 N2 M0, stage IIIc), found one year previously. Laparoscopic abdominoperineal resection and colostomy were done. A Port-A catheter was implanted into the patient's left subclavian vein for adjuvant chemotherapy with a regimen of leucovorin and fluorouracil, in September 5, 2005. She was free of tumor recurrence after 6 month-long adjuvant chemotherapy and was followed for 7 months. After 13 months, she experienced facial fullness (especially around the eyes), an explosive sensation in her head and shortness of breath while walking. When lowering her head, she felt discomfort, with dyspnea, engorgement of the neck and cyanosis of the lips. Itching sensations and erythematous changes in both eyelids developed gradually. A computed tomography (CT) scan of the patient's chest with contrast revealed a low-attenuation thrombus around the Port-A catheter (maximal axial diameter 1.7 cm; 3-4 cm long) within the SVC and left brachiocephalic vein (BCV; Fig. 1A). The coagulation profiles revealed no hypercoagulable state (Table 1). Thrombolytic therapy with urokinase (240,000 units per day for 3 days) was started. The clinical signs of SVC syndrome disappeared promptly. A cardiovascular surgeon removed the Port-A catheter on the fourth day. Low molecular weight heparin (1 mg/kg every 12 h) was given on days 4 and 5 and oral warfarin (5 mg per day) was prescribed beginning on day 5. The patient had no bleeding or any other complications related to thrombolytic or anticoagulation therapy. Three months later, a repeated CT scan of the patient's chest revealed no thrombus within the SVC or left BCV (Fig. 1B).

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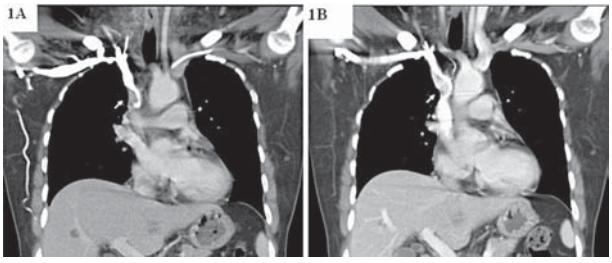

Fig. 1 Computed tomography of chest revealed a segmental thrombus (arrowhead) within the superior vena cava (SVC) with near total occlusion of vascular flow (A). The thrombus was resolved within SVC with patent vascular flow after thrombolytic therapy (B).

Table 1 Coagulation profiles of the patient

Laboratory test	Result	Normal range
Protein S	111.2%	60-145%
Protein C	111%	70-140%
Anti-thrombin III	109.0%	70-120%
Prothrombin time	11.4 seconds	10-14 seconds
Partial thromboplastin time	32.4 second	23.9-35.5 second
Fibrin degradation products	<10 μ g/ml	<10 μ g/m
D-dimer	470 ng/ml	<500 ng/ml
Anti-cardiolipid antibody	<1.00 u/ml	0-15 u/ml
Lupus anticoagulant	1.23	<1.2

DISCUSSION

The SVC syndrome is a structural obstructive emergency that can induce life-threatening respiratory failure and even cardiac arrest. The symptoms usually present with neck and facial swelling, dyspnea, cough, hoarseness, headaches, nasal congestion, hemoptysis, pain, dizziness, syncope and lethargy. Physical examinations reveal dilated neck veins and an increased collateral circulation over the anterior chest wall. More severe clinical presentations include proptosis, laryngeal edema, obtundation and even cyanosis[1].

The incidence of catheter-related venous thrombosis is 4-40% of patients according to clinic assessment or autopsy[2-4]. Thromboses within the SVC and BCV inducing the SVC syndrome are rare[4]. Todd et al. reported that venous thromboembolic disease of the BCV and SVC in hospitalized adults was diagnosed in 23 of 34,567 patients (0.06%)[5]. Diminished or absent opacification of central venous structures with prominent collateral venous circulation is required for the diagnosis of SVC syndrome. Contrast CT scans provide the most reliable view of the mediastinal anatomy and ability to detect a nonthromboembolic etiology[6,7].

Predisposing factors in the development of thrombosis include hypercoagulability with decreased antithrombin III levels, protein C deficiency and hemostatic disorders, mediastinal tumors with possible venous flow abnormalities and suboptimal catheter routines[8]. Tumor type is a risk factor for the development of thrombosis; thus, adenocarcinoma of the lung has a higher risk for this occurrence[9]. Patients with the syndrome exhibit procoagulant activity through the conversion of factor X to factor Xa[10]. The association between adenocarcinoma of the rectum and SVC syndrome has not been reported previously. Therefore, the implanted Port-A catheter could have induced SVC syndrome in this patient, who was free of tumor recurrence.

Gray et al. reported that direct thrombolytic therapy was an effective treatment strategy for SVC syndrome with an 88% success rate if symptoms presented for less than 5 days. However, the success rate reduced to 25% if symptoms lasted for longer. Urokinase and streptokinase could be used and 56% of patients had complete clot lysis and relief of symptoms. The efficacy of urokinase was superior to streptokinase, with fewer side effects[11]. Stenting therapy is also a choice to relieve patients with SVC syndrome with moderate to severe or rapidly worsening symptoms[12]. Long-term, low-intensity warfarin therapy using targeted INR1.5-2 is also a highly effective method of preventing recurrent venous thromboembolism[13].

In summary, this patient presented with SVC syndrome associated with thromboses in the BCV and SVC caused by a Port-A catheter. Sequential management including emergency thrombolytic therapy, removal of the catheter and subsequent anticoagulant therapy were effective treatments for this unpredicted but potentially fatal event.

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