SUPERIOR VENA CAVA THROMBOSIS

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Abstract

Superior vena cava syndrome (SVCS) is characterized by gradual, insidious compression or obstruction of the superior vena cava (SVC). The most common etiology of SVCS is related to malignancy such as bronchogenic CA and lymphoma. Other cases have a variety of causes such as chronic infections and chronic inflammations that involve the mediastinum. Spontaneous SVC thrombosis is also known to occur among rare causes. We report a case that presented with SVC thrombosis, of which the causes had not been identified. 

Keywords: superior vena cava, thrombosis, bronchogenic CA, lymphoma

A 69-year-old man presented with a 2-week history of worsening dyspnea, and swelling of the face, trunk and upper extremities. Five months earlier, he began dyspnea on exertion and when he performed some vigorous activities. These symptoms were gradually increasing. He neither sought medical examination nor received any investigation until the last 2 weeks, and his functional capacity worsened, with his face, trunk and upper extremities markedly swelling. He had had no endovascular catheterization, at any time especially central venous access line, and no illegal drug injections. He had a smoking history of 17 pack-years of cigarettes, but had abstained for the last 17 years. He also had a history of thyrotoxicosis and received an operation for this condition. After that he developed euthyroid stage and did not take any medication.

His physical examination revealed plethora, tachypnea, venous distension and edema of the head, neck, and upper extremities, with collateral vein development in the upper trunk. Other than these finding, his physical examination was normal and no clinical deep vein thrombosis was demonstrated elsewhere.

The chest radiograph obtained at presentation was normal on both views. There was no mediastinal widening.
Computed tomography of the thorax was performed and showed hypodense intraluminal filling defects consistent with intraluminal thrombosis in the right Internal jugular, Brachiocephalic and Azygos vein, and SVC caused distention of these vessels (Fig. 1-4). Neither internal obstruction nor an extrinsic compression that precipitates the thrombosis in these vessels was identified. The collateral circulation for this obstruction was not seen inside the thorax. Imaging of the abdomen was also performed and it did not find any abnormalities including the thrombosis. This cavity might be another possible source of thrombosis.

The patient’s laboratory investigations were normal for all causes of thrombophilia which included the conditions predisposed to hypercoagulable states (24-hour urine protein, ANA, Anti-cardiolipin, protein-C, protein-S) and the tumor marker (AFP, CA 19-9, PSA and CEA).
While on-going investigations were being conducted, we started treatment with intravenous heparin and followed that with oral warfarin. Despite keeping blood coagulation more prolonged (INR 2-3 times) than normal, the patient still deteriorated clinically. He had intractable hypoxia and hemodynamic compromise despite full mechanically ventilated and hemodynamic supports. We had discussed the risk and benefit from surgical procedure as a diagnostic for proper treatment and for the causes of thrombosis with his family, but they refused to allow us to operate. The patient died 4 weeks after the symptoms had begun and 2 weeks after the disease was diagnosed. An autopsy was performed within 10 hours after death.

**Autopsy finding**

The autopsy confirmed the radiologic findings of occlusive mural thrombi in the superior vena cava, right brachiocephalic vein and right internal jugular vein. The veins were dilated. The thrombi were limited to the right side in contrast to the left side. The lung, liver and spleen were congested. The pleural surfaces
showed dilated lymphatics and fibrinous pleuritis. Other organs including abdominal organs, the brain and spinal cord were unremarkable.

Histologically, these mural thrombi were organized and adhered to the venous wall from the superior vena cava and right brachiocephalic vein, respectively (Fig. 5). These thrombi were composed of fibrin deposits and fibroblastic proliferation with recanalization (Fig. 6 A-B). Neither definitive cancer cells nor granulomatous inflammation of the vascular wall was noted.

**Discussion**

Superior vena cava syndrome (SVCS) is characterized by gradual, insidious compression or obstruction of the superior vena cava.
Superior vena cava thrombosis (SVC). The typical symptoms of SVCS are most obvious when obstructive disease is almost complete. Patients with SVCS most often present with complaints of facial edema and erythema, swelling of the neck and/or arms, and visible dilatation of the veins in the upper extremity. Patients with SVCS may also complain of dyspnea, persistent cough and orthopnea. As the disease progresses, the symptoms may include hoarseness, periorbital edema, dysphagia, headaches, dizziness, syncope, lethargy, and chest pain.

Prior to modern antibiotics, infectious causes including syphilis, tuberculosis, and fungi occurred with almost equal frequency. Today, the most common etiology of SVCS is related to malignancy. The most common malignancy-related SVCS is bronchogenic CA, which accounts for nearly 80% of cases. Lymphoma accounts for approximately 15% of cases. Other cases have a variety of causes, including infectious and catheter-related etiologies. Increasingly, dialysis catheters and pacemaker leads are becoming associated with superior vena cava syndrome due to thrombosis.

Spontaneous SVC thrombosis is also known to occur among the rare causes such as hypercoagulable stages, pulmonary arteriovenous malformations, Crohn’s and Behcet’s disease, chronic lead exposure, thrombosis secondary to thoracic outlet syndrome, and ovarian hyperstimulation syndrome (OHSS) in patients who underwent in vitro fertilization following stimulation with a GnRH analog.

Radiological assessment includes plain radiography, CT, trans-esophageal ultrasound, MRI, angiograms and isotope scan.

Reported treatment modalities are medical treatment with thrombolyis therapy continued with anticoagulant; percutaneous interventions such as mechanical thrombectomy, transluminal venoplasty and stent placement, and open surgical thromboembolectomy.

The role of thrombolytic therapy and subsequent anticoagulation for SVCS has been demonstrated as effective in treating central venous catheter induced thrombosis in patients whose thrombosis occurred acutely. However, in our case the onset of thrombosis was unspecified and more likely to be chronic from the evidence of obstruction causing development of collateral circulation such as the dilatation of the superficial veins of the thorax. Therefore, we did not give our patient a thrombolytic drug. According to the pathologic results from the autopsy; the thrombi were composed of fibrin deposits and fibroblastic proliferation rather than fresh thrombi, which may explain the resistance to our anticoagulant treatment and no response to thrombolytic therapy.

Although factor-V Leiden mutation is another genetic predisposed to the hypercoagulable state, a laboratory test was not available at our institute and, furthermore, this hypercoagulable state has never been reported before in Thailand.

We reported a case of SVC thrombosis in a man without any specifiable causes. This is the first case of its type in our institute and Thailand.

Although extensive investigation was performed, we did not find any specific causes to finalize a diagnosis of idiopathic SVC thrombosis in this patient. A reason for this is because there are many possibilities that remain in modern and advanced medical science that need to be uncovered such as unknown genetic predisposing defects in the coagulating cascade, and physiological change of some low flow stages such as severe dehydration that causes thrombophilia.
ภาวะลิ่มเลือดอุดตันเส้นโลหิตดำซูเปอร์เวนา

ทีเรกิกุล ธีรกิตติกุล, พ.บ., นิรัชร์ เลิศประเสริฐสุข, พ.บ.

บทคัดย่อ
กลุ่มอาการซูเปอร์เวนาคาวาคือภาวะที่มีการอุดตันของเส้นโลหิตดำซูเปอร์เวนาคาวาซึ่งเกิดจากการทุกข์ทรมานอย่างค่อยเป็นค่อยไป สาเหตุส่วนใหญ่จะเกิดจากการทุกข์ทรมานเส้นเลือดที่มีตอนข์ทรมานเนื่องกิจกรรมของไฟเบอร์กล้าในกล้าในสมองหรือกล้าในกระดูกสันหลัง บริเวณที่ทำการแสดงอาการนั้น พบได้ในกล้าใหญ่ที่มีภาวะลิ่มเลือดอุดตันเส้นโลหิตดำซูเปอร์เวนา ทำให้อาการกิจกรรมของเส้นโลหิตดำซูเปอร์เวนาของโรคกิจกรรมที่มีภาวะลิ่มเลือดอุดตันเส้นโลหิตดำซูเปอร์เวนา ทำให้อาการกิจกรรมของเส้นโลหิตดำซูเปอร์เวนาของโรคกิจกรรมที่มีภาวะลิ่มเลือดอุดตันเส้นโลหิตดำซูเปอร์เวนา ทำให้อาการกิจกรรมของเส้นโลหิตดำซูเปอร์เวนาของโรคกิจกรรมที่มีภาวะลิ่มเลือดอุดตันเส้นโลหิตดำซูเปอร์เวนา ทำให้อาการกิจกรรมของเส้นโลหิตดำซูเปอร์เวนาของโรคกิจกรรมที่มีภาวะลิ่มเลือดอุดตันเส้นโลหิตดำซูเปอร์เวนา ทำให้การกิจกรรมของเส้นโลหิตดำซูเปอร์เวนาของโรคกิจกรรมที่มีภาวะลิ่มเลือดอุดตันเส้น

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