Huge Mediastinal Mass with SVC Syndrome Accompanying Numerous Chest Wall Collateral Vessels

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Abstract

A 47-year-old man was referred to our hospital because of dyspnea, cough and weight loss. On physical examination, marked dilatation of thoraco-superficial epigastric venous anastomosis was found. The chest wall collateral vessels revealed enlarged head-to-toe flow, suggesting complete obstruction of the SVC and one or more of the major caval tributaries, including the azygos system. Thoracic CT demonstrated that a huge anterior mediastinal tumor completely obstructed the superior vena cava. He was diagnosed with Hodgkin lymphoma of the nodular sclerosis type, Stage III B based on the biopsy specimen from the right subcutaneous lumbodorsal mass.

Key words: Hodgkin lymphoma, SVC syndrome, chest wall collateral vessels, anterior mediastinal tumor

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Introduction

Although the most common type of anterior mediastinal lymphoma is nodular sclerosing Hodgkin lymphoma (HL), the risk of superior vena cava (SVC) syndrome is rare in HL compared to non-Hodgkin lymphoma (non-HL) \((1-4)\). There are many reports concerning anterior mediastinal tumors with SVC syndrome. However, reports on SVC syndrome referring to the collateral vessels are scarce. Herein, we report a case of SVC syndrome with a discussion on the genesis of collateral vessels.

Case Report

A 47-year-old man was hospitalized because of cough for two months prior to admission, dyspnea for two weeks and weight loss (20 kg in 7 months). He claimed dilatation of superficial vessels on his trunk for a month. He had difficulty in walking, lying down and sleeping because of dyspnea. He was afebrile with a BP of 120/60 mmHg and pulse rate 112 beats/min. His respiratory rate was 24 breaths/min with an oxygen saturation of 97% under an ambient air. His face and neck were flushed, erythematous and edematous. His left supraclavicularly lymph nodes were firm, fixed, circumscribed, and rubbery palpable. A right subcutaneous lumbodorsal mass with anasarca was detected. In addition, marked dilatation of thoraco-superficial epigastric venous anastomosis was found, as shown in Fig. 1, panel A (white arrowhead). All of those veins revealed head-to-toe flow. Although his lungs were clear to auscultation bilaterally, breathing sound of left side was decreased and percussion was dull. His blood examination showed an increased WBC count (15,600/\(μ\)L; seg. 88.0%, eosino 0.3%, baso 0.2%, mono 5.4%, lymph 6.2%), LDH (580 IU/L), soluble IL-2 receptor level (12,200 U/mL) and erythrocyte sedimentation rate (83 mm/h). The chest roentgenogram depicted a huge mediastinal shadow accompanied by left pleural effusion (Fig. 1, panel B). Thoracic CT detected inhomogeneous enhanced huge mass of 16 cm in diameter in anterior medias-
Figure 1. Panel A: Thoraco-superficial epigastric venous anastomosis is seen. Panel B: Chest roentgenogram shows huge mediastinal mass accompanied by left pleural effusion. Panel C: Lymphonodes of paraaortic and paravertebral area (double arrow) and right subcutaneous lumbo-dorsal mass (arrow) as well as azygos vein (arrow head) are demonstrated on CT. Moderate bilateral pleural effusion and ascites are also detected. Panel D: Thoracic CT shows that the huge mass invade and completely obstructs the superior vena cava (asterisk), bilateral brachiocephalic veins and azygos vein. Dilatation of the thoraco-superficial epigastric venous anastomosis (white arrow head), internal thoracic vein (black arrow) and mediastinal venous plexus (black arrow head) are depicted. Vertebral veins and vertebral venous plexus are normal in size (white arrow).

astinum, which expanded along the paraaortic lymph nodes to paravertebral area (Fig. 1, panel C: double arrow), and right subcutaneous lumbo-dorsal mass (Fig. 1, panel C: arrow). There was a moderate amount of bilateral pleural effusion and ascites. Thoracic CT showed that the huge mass invaded and completely obstructed the superior vena cava (SVC) (Fig. 1, panel D: asterisk), bilateral brachiocephalic veins and azygos vein resulting in the marked development of subcutaneous collateral veins, such as thoraco-superficial epigastric venous anastomosis (Fig. 1, panel A, D: white arrowhead), internal thoracic vein (Fig. 1, panel D: black arrow) and mediastinal venous plexus (panel D: black arrowhead). On the 2nd hospital day, biopsy from the right subcutaneous lumbo-dorsal mass (Fig. 1, panel C, arrow) which extended successively to the paravertebral and mediastinal tumors and supraclavicular lymph nodes was performed. A biopsy specimen revealed the presence of Hodgkin cells and Reed-Sternberg cells in the fibrosclerotic background. Immunohistochemically, those were CD30 and CD 15 positive (Fig. 2). Thus, he was diagnosed with Hodgkin lymphoma (HL), nodular sclerosis type, Stage III, B. He responded well to the 6 courses of ABVD (Doxorubicin, Bleomycin, Vinblastine, Dacarbazine) treatment and the collateral vessels on his trunk disappeared.

Discussion

The most common malignant causes of superior vena cava (SVC) syndrome are non-small cell lung cancer (approximately 50% of patients), small cell lung cancer (approximately 25% of patients), lymphoma, and metastatic lesions (each approximately 10%) (1). The risk of SVC syndrome is high in T-cell lymphoblastic lymphomas (T-LBLs), moderate in mediastinal large B-cell lymphoma (MLBCL), and rare in HL in spite of its higher incidence as an anterior mediastinal tumor (2–4). The prevalence of SVC syndrome is higher in non-HL than HL. The mode of spread is rather lymphatic in HL, and contiguous and/or hematogenous in non-HL, which may imply the higher affinity to vascular systems in the latter (7). The present case was nodular sclerosing HL, and completely obstructed the SVC.

Stanford et al (5) analyzed venacavograms in 27 patients
Figure 2. Hodgkin cells and Reed-Sternberg cells are seen in the fibrosclerotic background. Immunohistochemically, those cells are CD30 and CD15 positive.

Figure 3. Thoraco-superficial epigastric venous anastomosis (thoraco-abdominal vein and superficial epigastric vein) and internal thoracic vein are receiving whole flow from upper limb, head and neck because of complete obstruction of azygos vein and SVC. Head-to-toe flow of those veins is partially running to the inferior vena cava (IVC) via intercostals veins, azygos and hemi-azygos veins.

with SVC syndrome and reported that there are four patterns of venous collateral return. 1) type I, partial obstruction (up to 90% stenosis) of the SVC with patency of the azygos vein, 2) type II, near-complete to complete obstruction (90-100%) of the SVC with patency and antegrade flow through the azygos vein and into the right atrium, 3) type III, complete obstruction of SVC with reversal of azygos blood flow, 4) type IV, complete obstruction of the SVC and one or more of the major caval tributaries, including the azygos system. Recent reports showed additional collateral venous patterns such as anterior jugular venous system using CT or multi-detector row CT with multiplanar and 3D imagines (6). In the present case, there was complete obstruction of SVC at the cavitral junction, which satisfied Type IV classification by Stanford et al (Fig. 3). Thoracic CT demonstrated the following developed collateral pathways: the internal thoracic route (Fig. 1. panel D: black arrow), the thoraco-superficial epigastric venous anastomosis (Fig. 1. panel A and D, white arrowhead). Thus, the complete obstructive process of azygos vein as well as SVC is considered to end in the development of thoraco-superficial epigastric venous anastomosis and the internal thoracic route, because those veins take the whole flow from the upper limbs, head and neck. On the other hand, here the flow of vertebral venous system (the vertebral veins and vertebral venous plexus; Fig. 1. panel D: white arrow) and azygos vein (Fig. 1. panel C; arrowhead) running to the inferior vena cava (IVC) were depicted normally, suggesting their flow was small in quantity, originating from thoraco-superficial epigastric venous anastomosis and the internal thoracic route via intercostals veins (Fig. 3). We confirmed
the fact that if the azygos veins do not work properly because of the proximal obstruction of SVC, the thoraco-superficial epigastric venous anastomosis and the internal thoracic vein could play a major part of collateral pathway. Thus, if the head-to-toe flow of superficial veins is observed on chest-abdominal wall, we should suspect complete obstructive process of SVC as well as azygos systems. The present case is reminiscent of a figure in an anatomic atlas (Fig. 3).

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References


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