Radiology Case Reports 15 (2020) 254–258

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Case Report

Stanford type IV venous collateral blood flow following complete chronic occlusion of the superior vena cava in a patient with lung cancer

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ARTICLE INFO

Article history:
Received 4 November 2019
Revised 5 December 2019
Accepted 16 December 2019

Keywords:
Collateral
Complete occlusion
Lateral thoracic pathway
Lung cancer
Stanford type IV
Superior vena cava syndrome

ABSTRACT

In superior vena cava occlusion, multiple collateral pathways develop to maintain venous drainage. Major patterns and pathways of venous collateral blood flow are well described, but rarely in complete chronic superior vena cava occlusion secondary to malignancy. A 59-year-old man with facial and upper extremity edema had a severely compressed superior vena cava at the initial diagnosis of stage IV mediastinal lung adenocarcinoma. The occlusion of superior vena cava progressed. After 10 months of treatment, the complete occlusion led to mild symptoms of hoarseness, muscle weakness, cough, and slight upper extremity edema. Venography clearly illustrated well-developed venous collateral blood flow through lateral thoracic, azygos-hemiazygos, and vertebral collateral venous pathways classified as Stanford type IV. The patient survived for a total of 20 months. He maintained Eastern Cooperative Oncology Group performance status of 1-2 until 2 months before death without severe symptoms of superior vena cava occlusion. This case described a rarely occurring venographic demonstration of well-developed Stanford type IV collateral pathway. Moreover, even with complete superior vena cava occlusion, well-developed Stanford type IV lateral thoracic collateral pathway can compensate for the venous flow without deterioration of performance status for a long period in certain cases.

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Acknowledgments: We would like to thank Editage (www.editage.com) for English language editing.
Declaration of Competing Interests: None.

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https://doi.org/10.1016/j.radcr.2019.12.010

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Introduction

Nonsmall cell lung cancer is the most common cause (approximately 50%) of malignant superior vena cava (SVC) syndrome, followed by small cell lung cancer (22%), and non-Hodgkin's lymphoma (12%) [1]. It often presents with progressive symptoms over several weeks, such as facial edema, dilated veins of head, neck, and chest, cough, dyspnea, and orthopnea. The severity of the symptoms varies widely and depends on the degree and rapidity of SVC narrowing, and the development of venous collaterals. Although SVC syndrome can have striking clinical presentation, it is rarely fatal (<1%), nor requires emergent intervention [1,2]. Chemotherapy, radiotherapy, or both effectively relieves SVC syndrome in 60% of patients with nonsmall cell lung cancer and 77% with small cell lung cancer. SVC stenting is considered for life-threatening symptoms such as confusion, obtundation, stridor, syncope without precipitating factors, hypotension, and renal insufficiency; and provides symptom relief in 95% of patients [3]. Owing to the successful management and poor prognosis of malignant SVC syndrome, clinical presentations and venous collateral pathways of chronic complete SVC occlusion have rarely been reported [4–7]. We report complete chronic SVC occlusion compensated by Stanford type IV collateral flow through lateral thoracic pathway in a patient with lung cancer.

Case report

A 59-year-old man was referred with short duration facial and upper extremity edema (1 week) and exertional dyspnea (1 month). He had a smoking history of 60 pack-years, hypertension, hyperlipidemia, and hyperuricemia. He also took a daily 100 mg aspirin tablet for common hepatic artery thrombosis diagnosed 2 years back. His chest X-ray showed mediastinal enlargement corresponding to the SVC and azygos vein that had existed since last year (Fig. 1A). A contrast-enhanced computed tomography (CECT) of the chest showed an abnormal mediastinal shadow measuring 35 × 30 mm with SVC compression (Fig. 1B and C). The enhanced SVC was severely compressed with accompanying dilation of the azygos vein. An 18F-fluorodeoxyglucose positron emission tomography revealed abnormal bilateral mediastinal and right hilar accumulation. He was diagnosed with moderate SVC syndrome with suspected mediastinal cancer.

The pulmonary lesion was not evident except for some lung cysts next to the mediastinal mass. A diagnostic video-assisted mediastinal tumor resection showed a rigid, white mass caudal to the azygos vein and anterior to the SVC. Resected mediastinal lymph nodes contained adenocarcinoma positive for cytokeratin (CK) 7, and negative for CK 20 and thyroid transcription factor-1 by immunohistochemistry. The pleural effusion showed adenocarcinoma cells. Hence, we made a final diagnosis of stage IV, T0N3M1a mediastinal lung adenocarcinoma, with pleural dissemination. Driver mutations of epidermal growth factor receptor gene or anaplastic lymphoma kinase gene were not detected.

At treatment initiation, the patient had mild facial edema with Eastern Cooperative Oncology Group performance status (ECOG PS) 1. Four courses of carboplatin and pemetrexed were administered. The patient declined radiotherapy as it entailed a long hospital stay. During treatment, he experienced intermittent swelling of his face and both arms, left shoulder pain, headache, and cough that were managed symptomatically. After 4 courses of chemotherapy, a repeat CECT revealed stable disease with no change in tumor size or SVC stenosis. Thrombosis of the right internal jugular vein was seen, and anticoagulation changed from aspirin to warfarin. Nivolumab was initiated 5 months after diagnosis as second-line therapy. At 8 months, the patient suffered cardiac tamponade from carcinomatous pericarditis. At 10 months, a decrease in mediastinal tumor size was confirmed after 8 cycles of nivolumab; however, the SVC was completely occluded (Fig. 2).

Venography performed to evaluate collateral venous pathways showed that venous blood from both arms entered into bilateral lateral thoracic veins, flowed through anterior and posterior intercostal veins, retrogradely traversed the azygos and hemiazygos veins to reach the bilateral lumbar veins, and ultimately entered the iliac vein and inferior vena cava to drain into the right atrium (Fig. 3). This was also depicted in the volume rendered 3-dimensional (3D) CT images (Fig. 4). His symptoms were not severe to warrant endovascular treatment. Eight months of disease control was obtained with 14 cycles of nivolumab. He maintained ECOG PS 1 with chronic yet mild symptoms of hoarseness, muscle weakness, cough, and slight upper extremity edema. He survived another 7 months with docetaxel, erlotinib, and nivolumab readministration. His total clinical course since diagnosis was 20 months. He maintained an ECOG PS of 1-2 until 2 months before death. His SVC occlusion was almost compensated by the collateral venous pathways.

Discussion

This case illustrates the well-developed Stanford type IV venous collateral blood flow through lateral thoracic, azygos-hemiazygos, and vertebral pathways in a patient with complete chronic occlusion of SVC due to lung adenocarcinoma. Venography clearly demonstrated the collateral venous flows from both arms into bilateral lateral thoracic veins, anterior or posterior intercostal veins, retrograde flow into azygos and hemiazygos vein to enter the bilateral lumbar veins, iliac veins and finally the inferior vena cava to drain into the right atrium. Stanford and Doty classified the major venographic patterns in SVC obstruction into 4 types; type I: partial obstruction of SVC with patency and antegrade flow in azygos vein, type II: near-complete obstruction of SVC with patency and antegrade flow in azygos vein, type III: complete obstruction of SVC with reversal of azygos blood flow, and type IV: complete obstruction of SVC and azygos system with development of chest-wall and internal mammary collaterals [8–10]. The venographic type depends on the level of obstruction, above or below the origin of the azygos vein. When the azygos vein is obstructed at the orifice together with the SVC, the connection of SVC and azygos vein is completely lost resulting in the de-

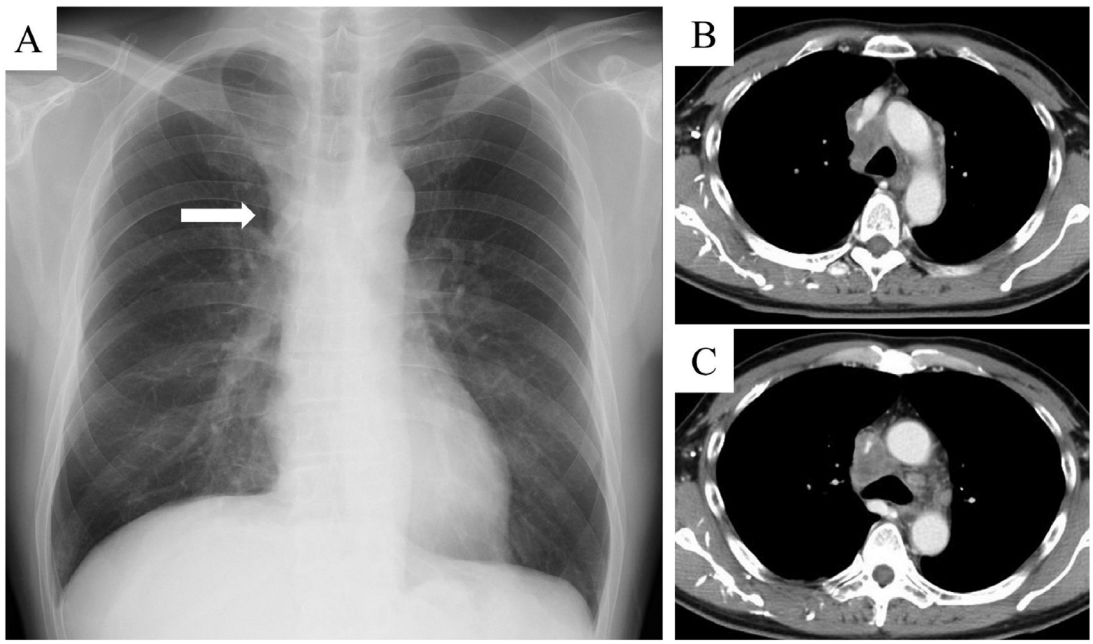

Fig. 1 – (A) Initial posteroanterior chest radiograph showed mediastinal enlargement in place of superior vena cava and azygos vein (arrow). (B, C) A chest contrast-enhanced computed tomography detected 35 × 30 mm abnormal mediastinal shadow that compressed the superior vena cava. The enhanced superior vena cava was severely compressed with accompanying dilation of the azygos vein.

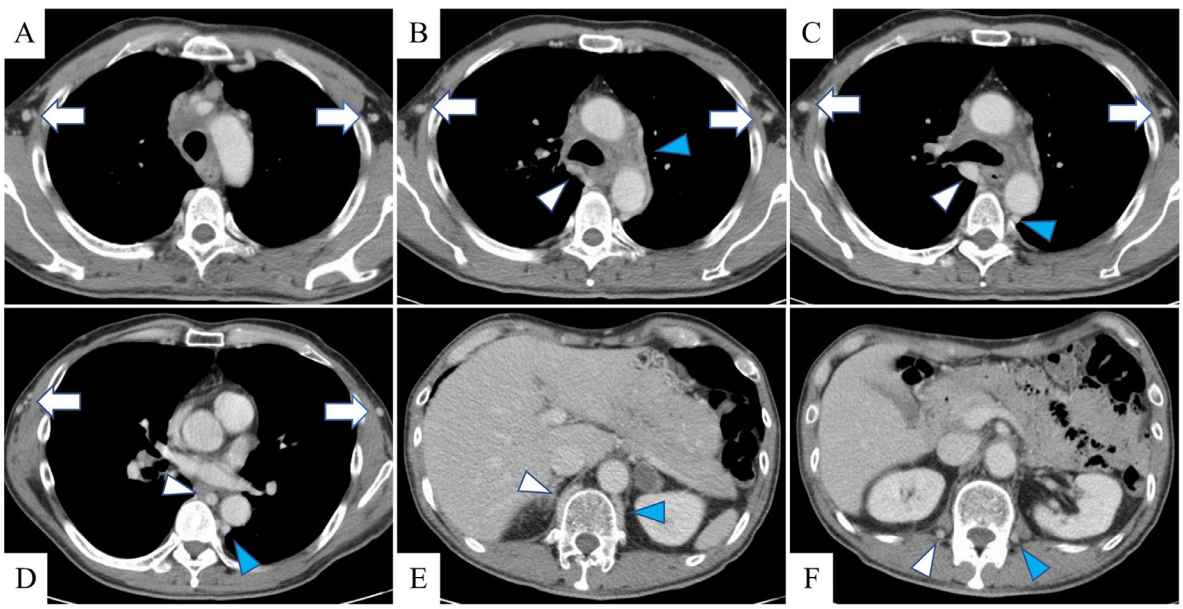

Fig. 2 – Mediastinal tumor size decreased after 8 cycles of nivolumab. Complete occlusion of superior vena cava was seen. Right and left lateral thoracic veins were seen as collaterals (white arrow in A-D). Azygos vein drained retrogradely into the right lumbar vein (white arrowhead in B-F). Left superior intercostal vein (blue arrowhead in B) drained to hemiazygos vein (blue arrowhead in C-E) and down to left lumbar vein (blue arrowhead in E, F). (Color version of figure is available online.)

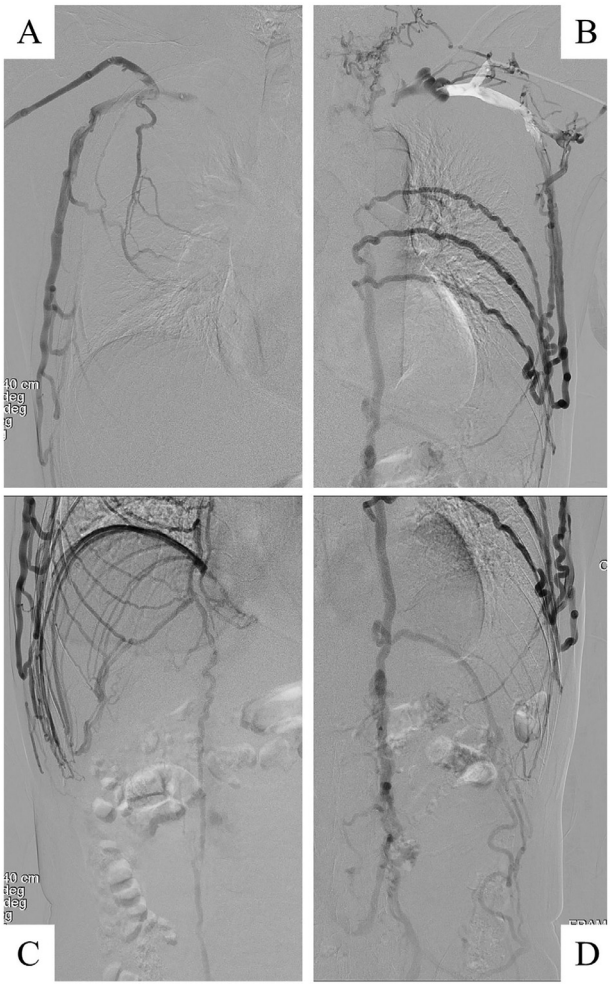

Fig. 3 – Venography clearly showed that collateral venous flow from both arms reached bilateral lateral thoracic veins, then the anterior or posterior intercostal veins followed by retrograde flow into azygos and hemiazygos veins emptying into the bilateral lumbar veins.

velopment of type IV collateral flows. Moreover, 4 main collateral pathways of SVC obstruction are acknowledged; azygos-hemiazygos pathway, internal and external mammary pathway, lateral thoracic pathway, and vertebral pathway [11–14]. In our case, the main venous flow from both arms was the lateral thoracic pathway.

Stanford admitted that the chest-wall collaterals were hard to visualize by venography [8,9]. Similarly, in our case, the collaterals of bilateral superior intercostal veins and internal thoracic veins were not seen on venography but were identified on CECT. The collaterals seen exclusively on CT may represent pathways from the head and neck. Clear venographic visualization of such well-developed Stanford type IV collateral pathways is valuable for its rarity. Such collaterals form with a chronic course of venous obstruction and collateral development.

The well-developed lateral thoracic collateral venous pathway compensated for the completely obstructed SVC, and the patient's symptoms were mild to moderate during his entire clinical period. After complete SVC occlusion, he survived 10 months with good ECOG PS. The symptomatic severity of SVC syndrome can vary widely and depends on collateral development. A slowly progressive SVC obstruction over several months could result in well-developed collaterals, as seen in our case. As the symptoms were not severe, we did not perform the endovascular treatment. The prognosis of malignant SVC syndrome is poor, with approximately 6 months of survival [1,2,7,15]. Our patient survived for 20 months, indicating that appropriate diagnosis and treatment can achieve prolonged survival in the era of cancer immunotherapy even in patients with malignant SVC syndrome [16].

In conclusion, this case clearly illustrated the well-developed Stanford type IV venous collateral blood flow through lateral thoracic, azygos-hemiazygos, and vertebral pathways by venography. Even with complete occlusion of the SVC, well-developed Stanford type IV lateral thoracic collateral venous pathway can compensate for sufficient venous flow without deterioration of ECOG PS for a long period in certain cases.

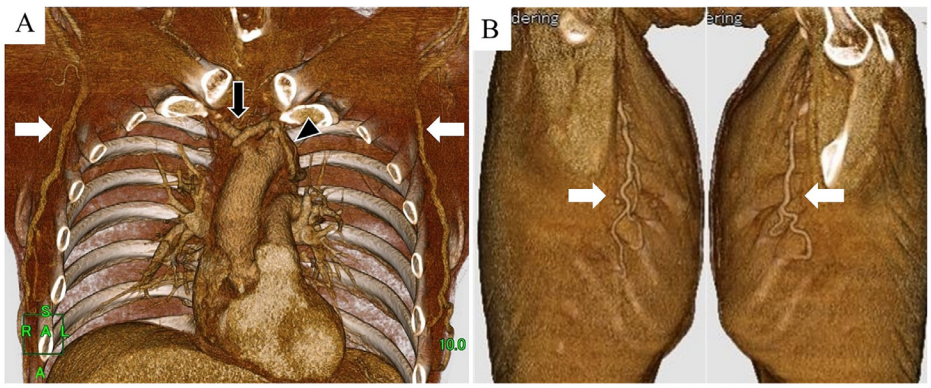

Fig. 4 – Volume rendered 3-dimensional computed tomography showed dilated right and left lateral thoracic veins as collateral pathways (white arrows in A, B). The left brachiocephalic vein was occluded before superior vena cava (black arrow). The left superior intercostal vein emerged from the left brachiocephalic vein toward the hemiazygos vein (black arrowhead).

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