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SUPERIOR VENA CAVA SYNDROME ASSOCIATED WITH RIGHT-TO-LEFT SHUNT THROUGH SYSTEMIC-TO-PULMONARY VENOUS COLLATERALS

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INTRODUCTION

Superior vena cava (SVC) obstruction is associated with the gradual development of venous collaterals. We present a rare form of systemic-to-pulmonary subpleural collateral pathway that developed in the bridging subpleural pulmonary veins in a 54-year-old woman with complete SVC obstruction. This uncommon collateral pathway represents a rare form of acquired right-to-left shunt due to previous pleural adhesions with an increased risk of stroke due to right-to-left venous shunting, which requires lifelong anticoagulation.

CASE REPORT

A 54-year-old woman presented with intermittent swelling of the face and arms. Her past surgical and medical history included pleurodesis for recurrent left-sided pleural effusions and Factor V Leiden deficiency disorder, for which she was taking warfarin. This was further complicated by recurrent hypercoagulable events including deep venous thrombosis and pulmonary embolisms. On examination, her body temperature was 36.8°C, heart rate was 94 beats per minute, blood pressure was 132/68 mm Hg, and respiratory rate was 18 breaths per minute. Clinical examination also revealed bilateral lung basal crackles, and prominent engorged vasculature in the neck and anterior chest wall.

Multidetector computed tomographic (MDCT) venography of the chest was performed. Coronal reformation images (Fig. 1A), maximum-intensity-projection images (Fig. 1B, C), and volume-rendered images (Fig. 1D) demonstrated complete chronic SVC obstruction (Fig. 1A) with numerous resultant paravertebral and chest wall venous collaterals.
Fig. 1. Multidetector computed tomographic venography images of chest in 54-year-old woman with complete SVC obstruction and abundant venous collaterals.

A. Coronal reformatted image demonstrates complete chronic SVC obstruction with abundant adjacent mediastinal collateral venous drainage (white thin arrow). B-D. Axial (B) and oblique sagittal maximum-intensity-projection images (C) along with volume-rendered images (D) reveal multiple venous collaterals in chest wall (white bold arrows) and paravertebral regions (arrowhead), representing involvement of lateral thoracic and paravertebral collateral pathways. Note dense contrast in patent left-sided venous collaterals (yellow bold arrow, B-D) extending from chest wall, crossing pleura, subpleural pulmonary veins and lingular lobe before draining into left superior pulmonary vein, which is consistent with connections between lateral thoracic pathway and systemic-to-pulmonary venous collateral pathway and bridging subpleural pulmonary veins.

SVC = superior vena cava

(經濟 1D) 位的外側胸壁和椎旁動脈側枝的參與。此外，左側未閉塞的肺靜脈側枝（黃色粗箭頭，B-D）從胸壁延伸，穿越肺葉，向下肺葉，最終排入左上肺靜脈，這與外側胸壁動脈和系統動脈-肺靜脈側枝路徑和橋接下肺靜脈的連接一致。

SVC = 上腔靜脈

(圖 1D)，代表了外側胸壁和椎旁動脈側枝的參與。此外，左側未閉塞的肺靜脈側枝（黃色粗箭頭，B-D）從胸壁延伸，穿越肺葉和下肺葉，最終排入左上肺靜脈，這與外側胸壁動脈和系統動脈-肺靜脈側枝路徑（SPVC）和橋接下肺靜脈的肺靜脈的參與一致。

DISCUSSION

This is an extremely rare case of SVC syndrome due to a hypercoagulable state presenting with a right-to-left anticoagulation with warfarin was initiated to reduce the risk of stroke. Angioplasty or stenting of the SVC was not recommended, because the SVC occlusion was secondary to a hypercoagulable state, rather than due to external compression, which in most cases, is due to malignancy. Due to the presence of extensive collaterals, embolization of the venous collaterals was not performed, and the patient was discharged after symptomatic improvement.
extracardiac shunt through bridging pleuro-pulmonary venous collaterals demonstrated on MDCT. Malignancy is the most common cause of SVC syndrome, and it accounts for two-thirds of the cases, while the remaining one-third of the cases have a benign etiology. SVC obstruction due to benign causes is most commonly due to indwelling large-bore venous catheters or pacet wires, while the other benign causes include fibrosing mediastinitis, aortic aneurysm, or infectious diseases (3, 6). However, regardless of its cause, SVC syndrome develops secondary to the obstruction of the SVC or brachiocephalic veins, resulting in venous congestion; thus leading to the formation of prominent venous collaterals to facilitate drainage of the venous blood to the heart (1). Commonly, SVC syndrome presents as facial edema, as in our patient, along with lightheadedness, engorged superficial head and neck veins, and orthopnea; while neurologic symptoms and laryngeal edema can occur in cases with severe SVC obstruction (1, 7).

Multidetector computed tomography is particularly beneficial in the diagnosis of SVC obstruction and SPVC (1, 2, 6). MDCT is less invasive, and it is comparable or even superior to conventional venography in assessing the site of obstruction, collateral venous pathways and associated thoracic abnormalities (1). In addition to the routine two-dimensional axial images, maximum-intensity projection and 3-dimensional volume-rendering reconstructions of MDCT can accurately demonstrate the anatomy of the great vessels, presence of SPVC and tortuous draining veins in detail (1, 2). The presence of SVC occlusion with venous collaterals is a reliable criterion for diagnosing SVC syndrome on CT, which has a sensitivity and specificity of 96% and 92%, respectively (8, 9).

Four major thoracic venous collateral pathways have been reported in the literature, and they include the azygos-hemiazygos pathway, internal/external mammary pathway, lateral thoracic pathway, and paravertebral pathway (1, 2, 4, 5); while unusual pathways include SPVC and portocaval routes (4, 5). Among the SPVC pathways, the connection between the brachiocephalic vein and the superior pulmonary vein via the bronchial venous plexus is the commonly reported pathway (4). However, our case had an uncommon type of SPVC, in which these pleuro-pulmonary venous collaterals were developed due to previous left-sided pleurodesis that was performed for recurrent left-sided pleural effusions. Angiogenesis within the pleural adhesions between the chest wall and lungs has been postulated as an underlying mechanism for the formation of bridging subpleural pulmonary veins (2), which drain into the left superior pulmonary vein. Thus, our patient had an acquired type of SPVC, with pleural adhesions and chronic inflammatory processes being the major precipitating factors (4). Overall, SPVC can be clinically important as the venous collaterals bypass the pulmonary circulation and may potentially result in intracranial emboli and high cardiac output failure (4), which fortunately were not observed in our patient. This uncommon venous collateral pathway represents a rare form of acquired right-to-left shunt due to previous pleural adhesions and represents a potential risk factor for stroke due to right-to-left venous shunting, which requires lifelong anticoagulation due to the concomitant presence of factor V Leiden deficiency and SPVC.

REFERENCES