Occlusion of the superior vena cava (SVC) is a complication of malignant and benign diseases that compress, occlude, or invade the SVC, subsequently directing blood flow into collateral veins. SVC obstruction is associated with multiple venous collaterals. These include the azygos-hemiazygos, internal and lateral thoracic veins, intercostal veins and the vertebral venous plexus which all drain systemic veins from the upper extremities, head and neck into the right heart. Systemic-to-pulmonary venous shunts (SPVS) may occur in rare cases (1–9). These shunts drain systemic veins from the upper extremities, head and neck into the left heart. SPVS have been described as unusual collateral pathways that appear in SVC obstruction, and occasionally have been detected in patients with lung cancer that obstructed the SVC (2). To the best of our knowledge, this is the second reported case of Budd-Chiari syndrome associated with SVC obstruction (10). Our case is interesting because it features two points that have not been previously emphasized: First, the majority of cases in this issue have lung cancer occluding the SVC; our patient had thrombus in the SVC due to coagulopathy. Second, systemic to pulmonary venous communication was illustrated using multidetector computed tomography (CT) angiography.

Case report

A 43-year-old man was admitted to the hospital because of swelling of his face, neck and upper extremities which had started a month before. The patient also complained of progressive dyspnea and cough. He had been diagnosed with Budd-Chiari syndrome due to coagulopathy (protein C and S deficiency and homozygous factor V Leiden mutation) three months earlier. Physical examination revealed that he had a heart rate of 78/min, respiratory rate of 28/min, and blood pressure of 120/70 mmHg. Venous distension of the neck and upper thorax was recognized. Left jugular vein thrombosis was demonstrated by Doppler ultrasonography of the upper extremities. CT angiography was performed using a 16-row system (Lightspeed 16, GE Medical Systems, Milwaukee, Wisconsin, USA) to evaluate the possibility of pulmonary emboli. The exam was performed after the intravenous injection of 110 mL of ioversol (350 mgI/mL, Optiray; Tyco Healthcare, Pointe Claire, Quebec, Canada) through the right antecubital vein with a power injector at a rate of 4 mL/s. The scan was obtained 30 s after the start of injection. The scanning protocol was as follows: 1.25-mm slice thickness, a pitch of 1.375, reconstruction interval of 0.8 mm, 120 kV, 380 mA. Initially, there was no visible opacification of the main pulmonary artery and its branches (Fig. 1). The pulmonary veins, the left heart and aorta were intensely opacified before opacification of the right heart and the pulmonary artery. There were numerous strongly enhanced collaterals, abundant peripheral bridging veins and pleural enhancement in the right hemithorax.
We then performed a second scan on another day which was obtained 30 s and 100 s after the start of injection through the left antecubital vein to see whether chest wall veins were present on the left hemithorax. We also aimed to achieve optimum opacification of the pulmonary artery on the second scan. The scan was performed using the same parameters as the first MDCT angiography. The last examination established the presence of collateral veins and intense pleural enhancement in the left hemithorax (Fig. 3). A poor opacification of the main pulmonary artery and its branches was achieved at 100 s. No pulmonary thromboembolus was detected.

Discussion

SPVS are typically associated with SVC obstruction, usually caused by malignant tumors and rarely by a benign condition. To our knowledge, a few cases have been reported with SPVS in lung cancer in the literature (1–9). In the case presented here, there was no lung cancer occluding the SVC. Our patient has coagulopathy due to protein C and S deficiency and homozygous factor V Leiden mutation associated with Budd-Chiari syndrome and SVC occlusion.

The mechanism by which SPVS develops is not well known, but inflammation and adhesion of the pleura is considered to be essential for angiogenesis of bridging veins penetrating across the pleura (6). Pulmonary tuberculosis and pleural thickening are important risk factors in SPVS (2, 4, 6). The peripheral bridging veins appear as a thick band of high attenuation in the peripheral zone of the lung on CT. Bridging veins may simulate thick interlobular septa. Proper diagnosis is necessary in order not to misdiagnose other diseases that may cause interlobular septal thickening such as congesive heart failure. In cases of SPVS, the pulmonary veins can be opacified without prior opacification of the right heart and pulmonary arteries. Prominent pleural enhancement and bridging veins also help in diagnosing these shunts (3).

Progressive thrombosis of veins caused by underlying hypercoagulability lead to the development of collateral veins in unusual sites, including SPVS. Ito et al. (2) considered that SPVS that penetrate across thick pleural effusion indicate that the SPVS, once formed under the condition of pleural adhesion, was not broken by the accumulation of massive effusion that might have a potential to separate adhesion of the pleura.

Reported methods for depicting SPVS include radionuclide studies using 99m-Tc-aggregated albumin (7), conventional venography (9), and helical CT (4, 6, 8). CT venography has been
widely used to examine patients with suspected SVC syndrome because it can show the cause of SVC syndrome, the exact level of venous blockage, and the collateral pathways (3, 5). We were obliged to perform two scans in order to achieve satisfactory images to look for pulmonary emboli. Initially, the right antecubital vein was used for injection of contrast material. During the second scan the contrast material was injected through the left antecubital vein. We detected unexpectedly SPVS on both hemithoraces. In fact, when venous blockage is bilateral or the SVC is obstructed, the contrast material may be injected into both upper extremities to visualize SPVS. The ability to provide multiplanar and three-dimensional imaging is the greatest advantage of multidetector CT angiography. Three-dimensional reconstruction images enhance the delineation of complex venous anatomy and help the clinician to understand SPVS along with SVC obstruction.

In conclusion, prominent pleural enhancement, highly attenuating pulmonary veins and opacification of the left heart chambers without opacification of the right heart chambers will help in diagnosing SPVS in patients with SVC obstruction. CT angiography is a useful tool to demonstrate the venous collateral pathways such as SPVS.

References