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Case report

Systemic-to-pulmonary venous shunt in a patient with non-Hodgkin lymphoma: A case report and review of the literature

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A B S T R A C T

We describe a case of a systemic-to-pulmonary venous shunt secondary to superior vena cava obstruction in a patient with newly diagnosed non-Hodgkin lymphoma. This rare condition manifested with symptoms of dyspnea and hypoxemia that were out of proportion to the pleural effusion diagnosed on chest imaging. Standard treatment of such rare collateral plexuses is observation. However, it is important for clinicians to be cognizant that in rare cases such plexuses can lead to right-to-left shunt complications such as embolism.

Case report

A 31-year-old man with no past medical history presented to the emergency department after 2 weeks of progressive dyspnea. His review of systems was positive for weight loss and night sweats. Physical examination revealed that the patient was hypoxic and unable to tolerate decubitus position. He had facial flushing and edema, and chest examination revealed decreased breath sounds in the right hemithorax. A chest computed tomography (CT) with contrast, injected through a left antecubital intravenous access, showed a large right-side pleural effusion and an 8-cm anterior mediastinal mass encasing both the superior vena cava (SVC) and the right brachiocephalic vein (Fig. 1). In view of these findings, the patient underwent video-assisted right thoracoscopic surgery with drainage of the effusion and pleurodesis. The biopsy of the mass and pleura was positive for T-cell lymphoblastic lymphoma, and chemotherapy was initiated.

After 2 weeks of chemotherapy the patient was still dyspneic and hypoxic and had no improvement in his facial edema. A repeat chest CT with contrast administered this time through a right antecubital intravenous access showed a slight decrease in the size of the mediastinal mass, a small loculated right pleural effusion, right internal jugular thrombosis and persistent narrowing of the SVC and right brachiocephalic vein (Fig. 2). Multiple new collateral vessels within the right chest wall and new bridging vessels across the pleura and mediastinum draining into the superior right pulmonary vein were now observed. There was also evidence of pleural enhancement (Fig. 2). The patient was diagnosed with a systemic-to-pulmonary venous shunt (SPVS), and no further treatment was offered for the unusual collaterals. The patient continued to receive treatment for lymphoma, but response was poor.

Discussion

SVC obstruction and secondary SVC syndrome are caused by malignancy in 85% of cases [1]. With venous flow obstruction, collateral venous plexuses are formed to maintain venous drainage from the upper extremities, head and neck into the right side of the heart. The most common collateral plexuses are the azygos, the internal/external mammary, the lateral thoracic and the vertebral [2–4]. SPVSs are an extremely rare form of collaterals after SVC obstruction. There are only 22 reports of SPVSs in the literature, and the majority of those cases were in patients with lung cancer.

Instead of draining into the right side of the heart, SPVSs drain systemic venous blood directly into the left side of the heart, causing a right-to-left shunt. The most common clinical manifestations of SPVS are dyspnea and hypoxia; rarely, paradoxical embolic events, cerebral abscesses and high cardiac output can occur [3,5,6]. In our patient, the right-to-left shunt was contributing to his symptoms of dyspnea and hypoxia, caused by the initial

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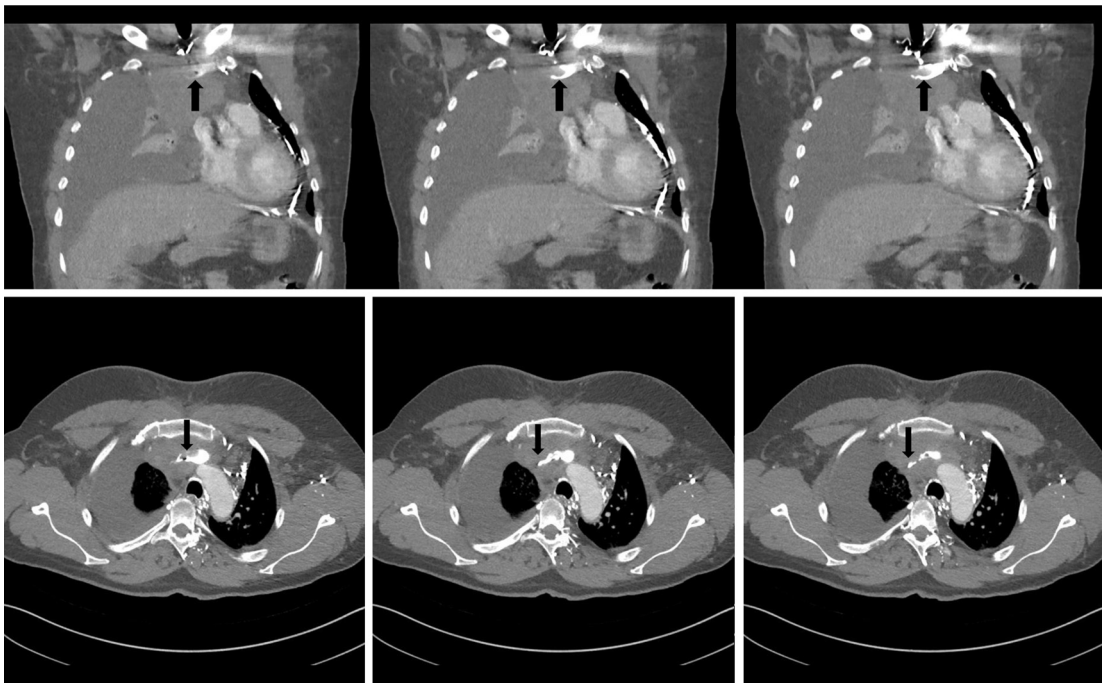

Fig. 1. Transverse (top) and coronal (bottom) CT scan images obtained with contrast administered through a left antecubital intravenous access, showing obstruction of the SVC by a mediastinal mass (arrow).

pleural effusion. After drainage of the fluid, the severity of the symptoms was out of proportion to the remnant small pleural effusion, which could be explained by the presence of the SPVS. No other signs of right-to-left shunt complications were evidenced in the patient.

SPVSs are categorized as 1) congenital, 2) anatomical or 3) acquired [3,5]. Congenital causes of SPVS consist of aberrant pulmonary veins with anomalous reversed flow, embryonic remnants of anastomosis between the cardinal system and pulmonary veins and a persistent left SVC that drains into the left atrium or

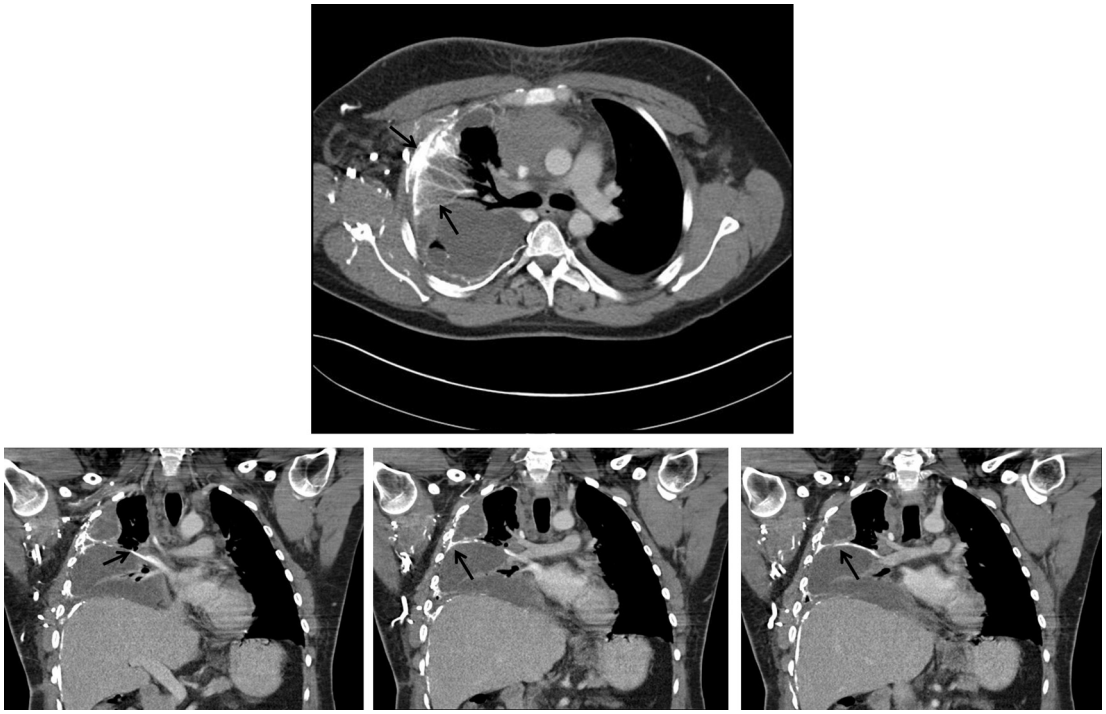

Fig. 2. Transverse (top) and coronal (bottom) views of CT scan images obtained with contrast administered through a right antecubital intravenous access, showing pleural enhancement (top, arrows) and bridging veins connecting intercostal and new chest wall collaterals draining into the pulmonary vein (bottom, arrows).

pulmonary veins [3,5]. Anatomical variations occur when there are preexisting bronchial venous plexuses connecting the bronchial and pulmonary veins [3,5]. If there is SVC obstruction, the systemic venous pressure rises, causing incompetence of the intervening valves and leading to venous reflux into the pulmonary veins [3]. Acquired SPVSs are formed in chronic inflammatory conditions such as tuberculosis, mediastinitis, chest radiation and chronic pleural effusions [7–9].

Liebow was the first to describe an acquired SPVS. He observed that in severely emphysematous lungs, chronic inflammation led to the formation of new intrapleural vessels [9]. He also described the development of these abnormal pulmonary collaterals upon ligation of the pulmonary veins of dogs [9]. During chronic inflammatory states, growth factors are released into the intrapleural space, leading to angiogenesis. The newly created vessels cross the pleura and create plexuses communicating the innominate and pulmonary veins [2,5,6,9]. If inflammation continues, the vessels will surround the airways and hilum, creating the bridging veins that on CT are observed crossing from the pleura to the mediastinum and into the pulmonary veins. In our patient, the SPVS was most likely secondary to the inflammation caused by the large pleural effusion, a mechanism already described in the literature [2,7,8]. We cannot completely rule out anatomical variations in this case, however; better imaging, such as venography, would be required to make that determination.

Various imaging modalities are available for the diagnosis of SPVS. CT venography and conventional venography have the advantage of three-dimensional reconstruction and better visualization of the site of venous blockage, vessel anatomy, delineation of the collaterals and venous hemodynamics [4,10]. Tests such as scintigraphy with technetium-99m, echocardiography with agitated saline through all extremities, cardiovascular CT, magnetic resonance imaging/angiography and cardiac catheterization are described in the literature as diagnostic methods for SPVS [2,4,8,10,11]. Conventional CT with intravenous contrast has become the most common mode of diagnosis due to its wide availability.

The typical findings on chest CT suggestive of SPVS are 1) evidence of intravenous contrast in the left cardiac chambers before its appearance in the right chambers, 2) evidence of multiple thick enhancing peripheral bridging veins and 3) prominent pleural enhancement—all in the setting of SVC obstruction [3,5,10]. In our patient's CT scan there was evidence of pleural enhancement and thick peripheral bridging veins draining into the pulmonary vein (Fig. 2). There was no evidence of early left chamber filling because the CT scan was not performed on first pass of the contrast. Our finding of an SPVS was obvious only when contrast was injected through the upper right extremity, not the left, suggesting that the shunt drained only the upper right extremity and right hemithorax.

Treatment for SPVSs is usually observant, as the majority of cases have been associated with only minimal hypoxia. However, the patient should be treated for the underlying cause of SVC obstruction and management of symptoms of SVC syndrome. In cases of anatomical or congenital SPVS, surgical or endovascular treatments have been described [6]. In our literature search we did not find reports of SPVSs that resolved after resolution of the SVC obstruction. An explanation might be that mortality in these cases was close after an SPVS diagnosis.

Conclusion

While in patients with acquired SPVSs intervention might not be necessary, physicians should recognize them in patients with SVC syndrome as a cause of hypoxemia. Treatment in our case was observant, as the patient did not develop severe complications from right-to-left shunting. Further follow-up and observation will help determine whether the patient's SPVS resolves or causes any further harm.

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