

Systemic to pulmonary venous shunt and the focal hepatic hot spot sign from SVC obstruction in Behcet's disease

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DESCRIPTION

A 60-year-old Thai man presented with the first diagnosis of Behcet's disease^{1,2} which illustrated the history of recurrent aphthous ulcers, positive pathergy test and several abnormal vascular manifestations. He had generalised central cyanosis with an inappropriate response to oxygen supplement, multiple signs of venous insufficiency and chronic bilateral transudative pleural effusions. Given profound hypoxemia with difficulty to correct, a right-to-left shunt was suspected. Neither arteriovenous malformations, liver cirrhosis nor intracardiac shunts were found from initial investigations. However, transthoracic echocardiography with agitated saline findings were compatible with a non-cardiac right-to-left shunt. Therefore, intravenous administration of radio-labelled, macro-aggregated albumin (Technetium ^{99m}Tc macro aggregated albumin (Tc-99m MAA)) in perfusion scan and aerosolised Tc-99m phytate in ventilation scan (V/Q scan) were performed which revealed no ventilation-perfusion mismatches. However, the perfusion scan findings revealed significant radiotracer accumulations in the brain (figure 1), heart, left lobe of liver, spleen and kidneys. The radiotracer particles are normally trapped in pulmonary capillary bed,^{3,4} the presence of extrapulmonary tracing is suggestive of right-to-left shunt. The whole-body CT demonstrated a small-sized, non-opacified superior vena cava (SVC) causing SVC obstruction (figure 2A). There were also numerous abnormal collateral vessels along great vessels, mediastinum and anterior chest wall (figure 2B-C) suggesting a shunt from systemic to pulmonary circulation which could contribute to right-to-left shunt physiology. Moreover, there were several wedge-shaped areas of intensely enhancing lesion at the left lobe of liver which is consistent with focal hepatic hot spot sign (figure 2D-F).⁵ This sign is a focal sign of the cavoportal collateral pathway in chronic SVC obstruction.⁶ There was also an extensive intraluminal thrombus causing nearly total inferior vena



Figure 1 Perfusion scan showed significant radiotracer accumulation in the brain.

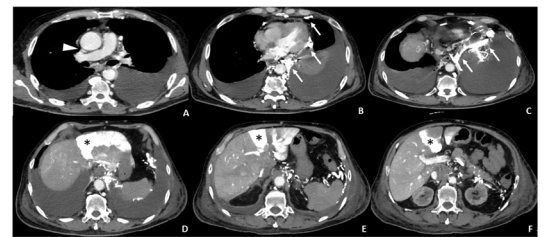


Figure 2 Axial view of chest CT showed a small-sized superior vena cava (A, arrowhead) multiple abnormal collateral vessels (B-C, arrow) and a focal enhancing lesion at the left lobe of liver (D-F, asterisk).

cava (IVC) occlusion (figure 3). With regard to extensive venous thrombosis, further investigations were done and revealed no evidence of antiphospholipid antibodies nor protein C and S deficiency.

In conclusion, our patient's clinical syndrome is consistent with Vasculo-Behcet's disease manifesting with extensive venous thrombosis and severe vasculopathy. The unusual collateral circulations resulting in systemic to pulmonary venous shunt



Figure 3 Coronal view of abdominal CT showed extensive intraluminal thrombus in inferior vena cava (arrow).



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is rarely reported in Behçet's disease. The cavoportal collateral circulation (focal hepatic hot spot sign) reflecting chronic SVC obstruction is seen in this case.

Learning points

- ▶ Right-to-left shunt should be aware in profound or difficult-to-correct hypoxia.
- ▶ Vasculo-Behçet's disease is one of the disease subtypes mainly leading to arterial or venous vasculopathy.
- ▶ A focal hepatic hot spot sign represents a cavoportal collateral circulations which could be found in chronic superior vena cava obstruction.

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