Pictorial Quiz

An unusual cause for cyanosis

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CASE DESCRIPTION

A 50-year-old female presented with a 10-month history of shortness of breath on exertion and cyanosis. There was no history of orthopnea or paroxysmal nocturnal dyspnea. She smoked twenty cigarettes a day and gave a 30-pack year smoking history. She had a background of chronic liver disease with portal hypertension related to alcohol and hepatitis C, and there was a history of substance abuse. On examination, she was comfortable at rest and centrally cyanosed with an oxygen saturation of 83% on room air. There was no finger clubbing. Her respiratory and cardiovascular system examinations were normal. There were no signs of heart failure. The differentials considered were pulmonary arterial hypertension (secondary to chronic obstructive pulmonary disease, substance abuse or chronic thromboembolic disease) and intra- or extra-cardiac shunts (pulmonary arteriovenous malformations, hepatopulmonary syndrome). Gas transfer was mildly reduced on pulmonary function tests, and a transthoracic echocardiogram was normal. Oxygen saturation improved to 95% on lying flat suggesting orthodeoxia. Her arterial blood gas showed a PaO_2 of 54 mmHg and a high A-a gradient of 71 mmHg. Computed tomography pulmonary angiogram revealed bilateral diffusely prominent pulmonary vasculature predominantly at the bases [Figures 1 and 2]. Bubble contrast echocardiography [Figure 3] showed bubbles densely filling the left heart through the pulmonary veins after a delay of three cardiac cycles, suggesting a large extracardiac shunt. A diagnosis of severe hepatopulmonary syndrome with platypnea-orthodeoxia was made. This increases morbidity and mortality in patients with chronic liver disease,[1] and liver transplantation is the only treatment option.[2]

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QUESTIONS

Image quiz

Q1: The differentials for Figure 3 include:

1. Patent foramen ovale
2. Large atrial septal defect
3. Intrapulmonary shunt
4. All of the above.

Multiple-choice question

A patient with chronic liver disease presents with shortness of breath and cyanosis on exertion. On evaluation, she has a raised A-a gradient and contrast echocardiography showed an extracardiac shunt.

Q2: What is the most likely diagnosis?

1. Portopulmonary hypertension
2. Hepatopulmonary syndrome
3. Pulmonary arteriovenous malformation
4. Budd–Chiari syndrome.

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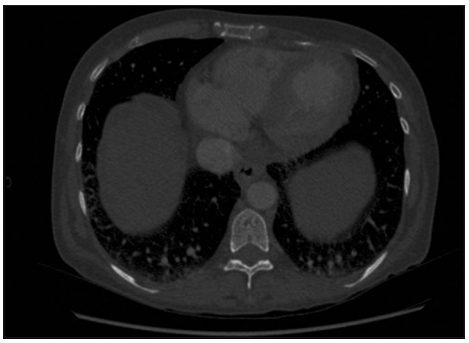

Figure 1: Computed tomography of the pulmonary angiogram (cross-sectional view) revealed bilateral diffusely prominent pulmonary vasculature predominantly at the bases

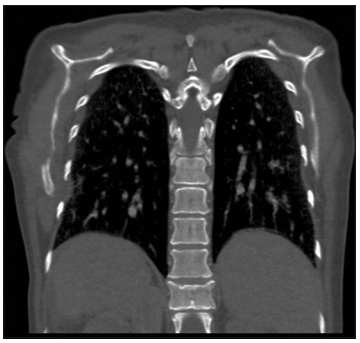

Figure 2: Computed tomography of the pulmonary angiogram (coronal view) revealed bilateral diffusely prominent pulmonary vasculature predominantly at the bases

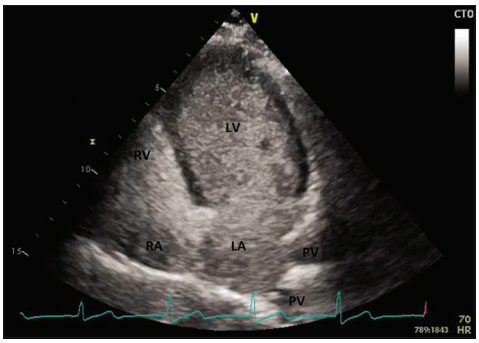

Figure 3: Bubble contrast echo showing the right and left heart filled with agitated saline (RV: Right ventricle; LV: Left ventricle; RA: Right atrium; LA: Left atrium; and PV: Pulmonary veins)

ANSWERS

Answer 1: Correct answer 4.
Answer 2: Correct answer 2.

Hepatopulmonary syndrome is a rare cause of cyanosis due to intra pulmonary shunts in patients with chronic liver disease. Orthodeoxia (oxygen desaturation causing cyanosis in the erect position, which resolves on lying down) is commonly seen. An increased A-a gradient and evidence of intrapulmonary shunt on contrast echocardiography in the background of chronic liver disease are diagnostic of hepatopulmonary syndrome.

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Conflicts of interest
There are no conflicts of interest.

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