Case of the Month

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Each month, we will present a challenging Case of the Month for **The Green Journal** readers, who must use their clinical acumen to arrive at the correct answer. We will also post the case each month on the *Journal's* web site (http://www.elsevier.com/locate/ajmselect). Several possible answers may be consistent with the case presentation; use your best judgment. Please send your answer (one per respondent) and indicate the case to which you are responding to **The Green Journal** at editors@amjmed.org or via FAX to (415) 447-2799. Only those answers with a complete mailing address will be considered. The correct answer will appear in the January issue of the *Journal*. The first five persons who submit correct answers will receive a free one-year subscription to the *Journal*. Colleagues of Drs. Walensky and Hellmann at Johns Hopkins are not eligible for this month's case. We will offer special recognition to the clinicians with the most correct answers at the end of the year. If you would like to contribute a case, please submit a brief synopsis (<250 words) to the editorial office. **Am J Med. 1998; 105:544–545.** ©1998 by Excerpta Medica, Inc.

A 32-year-old man presented to the emergency room with shortness of breath and increasing dyspnea on exertion for 2 weeks. Two weeks before admission, a rheumatologist diagnosed dermatomyositis as the explanation for the patient's 4-month history of weakness, arthralgias, myalgias and a scaling, erythematous rash over the eyelids, ears, elbows, and extensor surfaces of the fingers. The rash and myalgias improved on prednisone 60 mg per day, but 1 week later the patient reported hoarseness, a nonproductive cough, and increasing dyspnea on exertion. A computed tomography (CT) scan of the chest showed atelectasis and bilateral interstitial infiltrates. On the evening of admission his breathlessness worsened acutely, prompting his visit to the emergency department.

His past medical history was remarkable for a positive tuberculin skin test (PPD); he was treated with isoniazid for 1 year (compliance unknown). His current medications were salmeterol by metered dose inhaler, prednisone 60 mg daily, and calcium carbonate. The patient was married, had stopped smoking cigarettes 5 years prior, occasionally drank alcohol, and denied using illicit drugs. Over the previous 3 months, he had lost 14 kg as a result of painful oral ulcers that resolved before beginning prednisone therapy. He denied fevers, chills, orthopnea, paroxysmal nocturnal dyspnea, and chest pain.

On exam, the patient appeared well and was speaking in full sentences with a hoarse voice. His vital signs were a temperature of 35.7°C, blood pressure of 139/80 mm Hg, pulse of 106 beats per minute and respiratory rate of 32 breaths per minute. The O_2 saturation was 94% on room air. The oropharynx was remarkable for several nontender ulcers on the tongue. There was no palpable lymphadenopathy. Lung exam revealed fine bibasilar crackles. There was no jugular venous distention. Heart sounds were normal. The abdomen was without organomegaly. All joints had full range of motion and were without erythema or effusion. Telangiectasias were present at the palpebral margins, and there was a scaling, erythematous rash over the posterior surface of the neck and behind the ears. Periungual erythema was present on all digits and violaceous scaling macules covered all hand and finger joints. Neurologic exam, including assessment of strength, was normal.

Laboratory values were remarkable for a white blood cell count of 5,700 per μL, with 8% band forms, 77% neutrophils, 12% lymphocytes, and 3% monocytes. The erythrocyte sedimentation rate was 90 mm/h. Electrolytes were within normal limits. Serum creatine kinase level was 262 U/L (normal 0–175 U/L), aspartate aminotransferase (AST) level was 76 U/L (normal 0–35 U/L), and alanine aminotransferase (ALT) level was 81 U/L (normal 0–40 U/L), with normal levels of serum bilirubin and alkaline phosphatase. Tests for hepatitis B and C were negative. With the patient wearing nasal prongs delivering 2 L of oxygen per minute, the arterial blood gas was pH of 7.48, PaO_2 100 mm Hg, $PaCO_2$ 33 mm Hg with a calculated bicarbonate of 24 mmol/L. A chest radiograph showed bilateral lower lobe interstitial and alveolar infiltrates. Electromyography and nerve conduction studies revealed an irritative myopathy. Muscle biopsy was nondiagnostic. Dermatology consultants were impressed by the "classic" rash, and punch biopsy was histopathologically consistent with dermatomyositis. Blood tests for anti-nuclear antibodies, anti-double-stranded DNA, rheumatoid factor, anti-Ro, anti-La, p/c-ANCA, anti-Hu, and anti-Jo-1 were negative. Pulmonary function tests were consistent with restrictive lung disease. Review of the chest CT scan demonstrated a 2 cm irregular right lower lung mass, small mediastinal lymph nodes, and minimal diffuse lower lung scarring. An induced sputum grew *Candida albicans* at 3 days.

On the 12th hospital day, the patient had a fever 38.4°C with deteriorating respiratory status, and he eventually required mechanical ventilation.

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What test should be performed next? What is the diagnosis?

ANSWER TO THE NOVEMBER CASE OF THE MONTH

Last month's patient (submitted by Dr. Richard Sohn) had platypnea (breathlessness with upright posture) and orthodeoxia (hypoxemia with upright posture) due to a patient foramen ovale. This condition is caused by position-dependent right-to-left intrapulmonary or intracardiac shunting of blood, and can also be caused by pulmonary emboli, pulmonary arteriovenous malformation, and following pneumonectomy. The diagnosis was made by transesophageal echocardiography with bubble study in both supine and upright positions. There were no intracardiac communications when the patient was supine, but marked interatrial shunting was present when she sat upright. At surgery, her dilated aorta was seen to be compressing the right atrium, causing intermittent right-to-left shunting through the patent foramen ovale. Following surgery, this woman's platypnea and orthodeoxia completely resolved.

Correct answers to the September case (hepatic tuberculosis) were provided by Alberto León Dain (Córdoba, Argentina), Susan E. Wiegers (Philadelphia, Pennsylvania), and Ganesh C. Kudva (St. Louis, Missouri). They will receive a free one-year subscription to **The Green Journal**.