

Yale University

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Re: Lenor

MR # 175 59 33

Service: Interstitial Lung Disease Clinic

Referring Physician: Joao Nascimento, M.D.

Date of Birth: 1/20/1963

Date of Service: 11/10/2003

CLINIC SUMMARY:

CHIEF COMPLAINT:

Ms. Lenor is a 40-year-old white female from Connecticut who is referred by Dr. Nascimento of Bridgeport, Connecticut, for a second opinion regarding the management of interstitial lung disease in the setting of polymyositis.

HISTORY OF THE PRESENT ILLNESS:

Ms. presented in May 1999 with severe weakness and was diagnosed at Yale-New Haven Hospital with polymyositis. She had a CPK of 11,000. The details of that hospital record are not available to me. She was initiated on treatment with oral prednisone at 60 mg QD and subsequently was started on methotrexate. She has been under the care of Dr. Nascimento since that time. She has also been treated with IV-Ig for approximately one year which she felt did initially help her, although she became refractory to it. She is currently taking Remicade monthly, Imuran, and prednisone. She reports severe retractable cough. She has limited exertional tolerance although apparently does not desaturate with exertion. She denies fevers, chills, nausea, vomiting or diarrhea. She does complain of nausea following the Remicade and is generally quite ill after taking it. Over the last year she feels as though she has worsening lung function, but what is most difficult for her is the severe, intractable cough. Her mother had tuberculosis but Ms. never had tuberculosis herself, as far as she knows. She is a lifelong nonsmoker and is a strict vegetarian.

PAST MEDICAL HISTORY:

1. Polymyositis.
2. Interstitial lung disease.
3. Raynaud's.
4. Cataracts.

CURRENT MEDICATIONS: prednisone 30 mg QD, azathioprine 100 mg QD, Remicade monthly (? dose).

REVIEW OF SYSTEMS:

Notable for cataracts. She denies gait disturbances. She does complain of easy bruisability. She has intermittent nausea. She denies symptoms of reflux. The remainder of her review of systems are unremarkable.

PHYSICAL EXAMINATION:

She is a well appearing female who appears slightly younger than her stated age. Blood pressure is 98/50, heart rate is 75, oxygen saturation is 100% at rest, temperature is 98. HEENT is unremarkable. Neck is supple with full range of motion. Extraocular muscles are intact. Sclera are anicteric and non-injected. There is no thyroid enlargement. Auscultation of the chest reveals late inspiratory crackles about one-third of the way up the posterior lung fields. Cardiovascular exam shows a normal S1 and S2 with no murmurs, rubs or gallops. Abdomen is soft and nontender with no organomegaly. Extremities show no cyanosis, clubbing or edema.

REVIEW OF AVAILABLE DATA:

1. A chest CT scan with high resolution cuts from 11/7/03 showed predominantly lower lung zone reticular changes with some subpleural honeycombing and traction bronchiectasis. The pattern is consistent with usual interstitial pneumonia.
2. Pulmonary function tests performed on 8/14/03 revealed a forced vital capacity of 2.23 (51% predicted), FEV1 of 1.68 (50% predicted), and total lung capacity of 2.99 (45% predicted). Diffusing capacity was 12.92 (56% predicted, uncorrected), however, it normalized for alveolar volume.

ASSESSMENT:

Ms. Lenor is a delightful 40-year-old female who is referred by Dr. Nascimento for further opinion regarding the management of her interstitial lung disease in the setting of polymyositis. There is a reported history of a Jo-1 positive antibody. She has been treated with a variety of medications including prednisone, methotrexate, Imuran and IV-Ig. The best results I have seen with interstitial lung disease in the setting of a Jo-1 antibody has been with cytoxan. Ms. has not received cytoxan. I discussed the possibility of either oral or IV cytoxan with Dr. Nascimento and he agrees it is a reasonable consideration. He has extensive experience with the use of the drug and is comfortable initiating therapy. The pattern on chest CT scan is most consistent with the usual interstitial pneumonia pattern, although most patients with polymyositis when they have a biopsy reveal a nonspecific interstitial pneumonia pattern. We also discussed the possibility of consideration for lung transplant, but given the extent of her muscle disease I think it is unlikely that she would be considered a candidate. We also talked about management issues for the refractory cough. One possibility could well be related to her nausea and potential reflux symptoms. Many patients who do not complain of reflux particularly with restrictive lung disease can have significant evidence of reflux when evaluation is performed. I would therefore recommend a referral to a gastroenterologist for a pH probe as well as upper endoscopy. She should also have an ENT evaluation to look at her vocal cords. Treatment with a proton pump inhibitor may significantly improve her cough. I would also recommend she have an echocardiogram to determine whether she has any evidence of pulmonary hypertension.