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# December 2021 Pulmonary Case of the Month: Interstitial Lung Disease with Red Knuckles

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## *History of Present Illness*

A 56-year-old man was referred for a second opinion on recent onset of diffuse parenchymal lung disease. He had started noting mild dyspnea with yard work approximately in March 2021. His symptoms progressed over the next month with increasing shortness of breath and some fever. He presented to outside emergency department on April 17, 2021 and chest CT showing patchy ground-glass opacities with some areas of irregular consolidation (Figure 1).

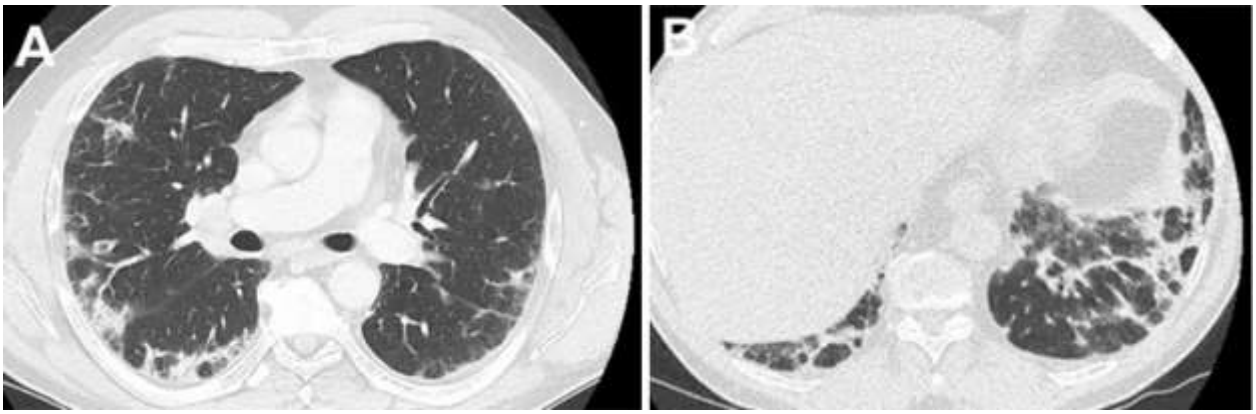


Figure 1. Representative images from the thoracic CT in lung windows from outside emergency room visit.

He was subsequently seen by an outside pulmonologist and started empirically on prednisone (50 mg/day). An outside lung biopsy had been performed which showed nonspecific interstitial pneumonitis. There was some improvement in his symptoms and his prednisone dose was reduced to 20 mg/day; however, his symptoms subsequently worsened with saturations noted to drop to 85% with any ambulation. He also had

swelling of his left face and a biopsy of the parotid gland with the findings suggestive of malignancy, possibly melanoma.

What should be ***done at this time***?
1. History and physical examination
2. Repeat the open lung biopsy
3. Repeat the parotid biopsy
4. 1 and 3
5. All of the above

**Correct!**
**1. History and physical examination**

First things first, the patient needs to be seen and have a good history and physical. Repeated biopsies of lung or the parotid gland would rarely be performed unless absolutely necessary.

His past medical history (PMH), family history (FH), social history (SH), and medication list (Meds) are below:

- PMH: hyperlipidemia, hypertension
- Family History: Father with h/o "pulmonary fibrosis"
- SH: smoked ½ pack per day for 10 years quitting in 1996
    - He had previously vaped (nicotine, none recently)
    - He denied illicit drug use
- Meds: rosuvastatin, sertraline, prednisone 55 mg/day, Bactrim DS taken 3 times weekly, clonazepam 0.5 mg daily

Physical examination revealed the previously noted left facial swelling along with redness over the knuckles which the patient said itched intensely (Figure 2).

Figure 2. Photograph of the patient's hands.

What is the patient's likely ***underlying rheumatologic disease***?
1. Amyopathic dermatomyositis
2. Psoriatic arthritis
3. Rheumatoid arthritis
4. Sjogren's syndrome
5. Systemic lupus erythematosus

**Correct!**
**1. Amyopathic dermatomyositis**

The patient's skin rash might be several things but is most consistent with Gottron papules which are pathognomonic for dermatomyositis/polymyositis (DM/PM). A skin biopsy was performed which was nonspecific. However, given the clinical

presentation the dermatology consultant was confident in a diagnosis of amyotrophic (without muscle involvement) dermatomyositis.

What should be ***done at this time***?

1. Repeat the chest CT scan
2. Repeat the open lung biopsy
3. Serum rheumatology panel
4. 1 and 3
5. All of the above

**Correct!**
**4. 1 and 3**

It is rarely necessary to repeat surgical biopsies. Given that the Gottron nodules, a rheumatology panel is justified (Table 1).

Table 1. Serum rheumatology panel.

- SS-A/Ro Ab, IgG, S: <0.2
- SS-B/La Ab, IgG, S: <0.2
- Sm Ab, IgG, S: <0.2
- RNP Ab, IgG, S: <0.2
- Scl 70 Ab, IgG, S: <0.2
- Jo 1 Ab, IgG, S: <0.2
- CPK and aldolase were both normal

Repeating the CT scan is also reasonable (Figure 3).

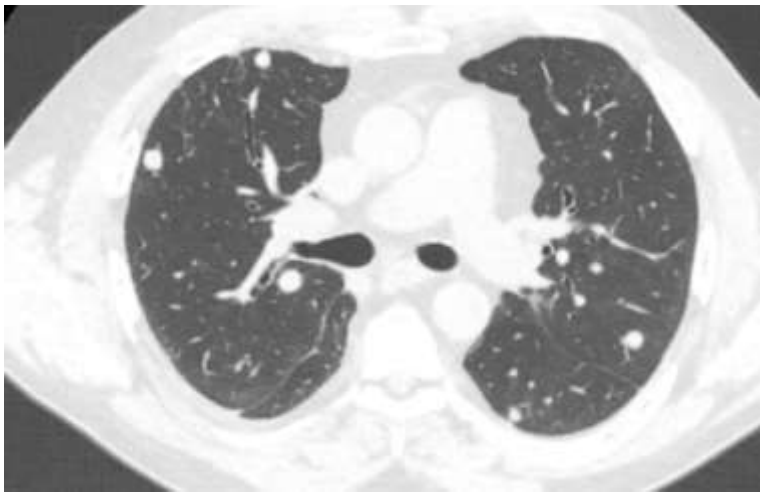

Figure 3. Representative image in lung windows from the repeat thoracic CT scan.

Which of the following are ***true*** regarding the patient's repeat thoracic CT scan and serum rheumatology panel?

1. The new lung nodules are dermatomyositis nodules and indicative of progressive disease
2. The new lung nodules raise a concern for metastatic cancer
3. The normal serum rheumatology panel is inconsistent with a diagnosis of dermatomyositis
4. 1 and 3
5. All of the above

**Correct!**
**2. The new lung nodules raise a concern for metastatic cancer**

Although a Jo 1 antibody is one of the more frequent antisynthetase antibodies associated with DM/PM, a negative result does not exclude DM/PM as there are other less common antibodies associated with these disorders and more extended testing for myositis-associated antibodies is recommended if there is a strong clinical suspicion. To our knowledge there is no such thing as "dermatomyositis nodules".

DM/PM is associated with various cancers and the detection of multiple new lung nodules is concerning for a cancer with lung metastasis.

A positron emission tomography (PET) scan was ordered because of the concern for possible cancer (Figure 4).

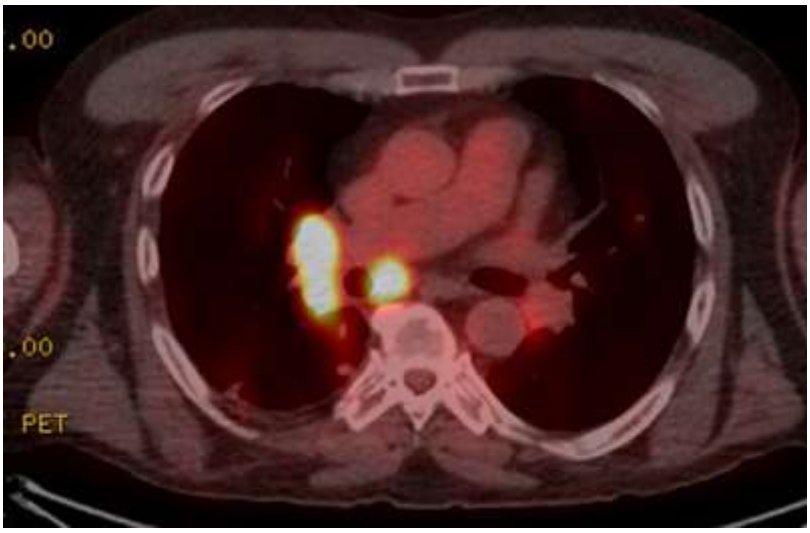


Figure 4. PET scan showing increased uptake in the right perihilar and left peritracheal regions.

What should be ***done at this time***?

1. Bronchoscopy with biopsy of the lung nodules and the appropriate lymph node stations
2. Cancer screening
3. Order of an extended panel for myositis-related antibodies
4. 1 and 3
5. All of the above

**Correct!**
**5. All of the above**

After repeatedly warning against repeated biopsies, it seems almost hypocritical to now recommend a biopsy of the lung nodules and lymph nodes. However, the detection of the new nodules suggests the possibility of a new disease process. Bronchoscopy with endobronchial ultrasound was performed with biopsy of 4R, 7 and 10R lymph node stations. The histology showed poorly differentiated sarcoma, possibly of endothelial origin. This was similar to the pathology on the outside biopsy of the parotid gland. An extended panel for myositis-related antibodies demonstrated the presence of anti-U2 RNP (ribonuclear) antibody, supporting a diagnosis of dermatomyositis.

A diagnosis of amyopathic dermatomyositis with rapidly progressive interstitial lung disease (ILD) was made.

Which of the following are ***true*** regarding amyopathic dermatomyositis?

1. ILD is seen in 60-80% of patients with DM/PM
2. Lung histologic findings can include NSIP, UIP, organizing pneumonia (OP) and diffuse alveolar damage (DAD)
3. Dermatomyositis is associated with a 6-fold higher risk of malignancy than the general population.
4. 1 and 3
5. All of the above

**Correct!**
**5. All of the above**

ILD occurs in 60% to 80% of patients with DM/PM and is even higher with amyopathic dermatomyositis. NSIP is reported to be the most common histologic pattern, followed by DAD. Lung biopsy is usually not necessary if ILD is present in patient with clinical findings of DM/PM. Corticosteroids are the usual initially recommended treatment for ILD associated with DM/PM.

Amyopathic dermatomyositis is seen in about 20% of patients with dermatomyositis and is characterized by cutaneous findings Gottron papules on the extensor aspects of metacarpal and interphalangeal joints, periungal erythema or facial heliotrope rash with no muscle involvement or only minimal weakness.

Dermatomyositis is associated with a 6-fold higher risk of malignancy than the general population. The risk is highest with the first year after diagnosis and decreases slowly over the subsequent 5 years. Cancer may present prior to the diagnosis, concurrently (as in the patient presented), or after the diagnosis of DM/PM. It is recommended that cancer screening be done, although there is not a consensus on the specific screening procedures.

## *References*


1. Waldman R, DeWane ME, Lu J. Dermatomyositis: Diagnosis and treatment. J Am Acad Dermatol. 2020 Feb;82(2):283-296. [CrossRef] [PubMed]
2. Udkoff J, Cohen PR. Amyopathic Dermatomyositis: A Concise Review of Clinical Manifestations and Associated Malignancies. Am J Clin Dermatol. 2016 Oct;17(5):509-518. [CrossRef] [PubMed]
3. Long K, Danoff SK. Interstitial Lung Disease in Polymyositis and Dermatomyositis. Clin Chest Med. 2019 Sep;40(3):561-572. [CrossRef] [PubMed]