EJ antibody antisynthetase syndrome

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DESCRIPTION

A woman in her 50s presented with several months of dyspnoea, joint pain and skin changes. She denied muscle weakness or dysphagia. Physical exam revealed hyperkeratosis and cracking at multiple fingertips ([figure 1](#)). Muscle strength was normal. CT of the chest revealed honeycombing and ground glass opacities in her bilateral lower lung lobes ([figure 2](#)). Laboratory testing revealed creatine kinase of 277 U/L (0–163 U/L), aldolase of 19.7 U/L (<8.1 U/L) and positive anti-EJ antibody greater than 100 SI (<11 SI). She was diagnosed with EJ antibody antisynthetase syndrome based on skin, lung and laboratory findings. She was started on prednisone 60 mg by mouth daily and mycophenolate mofetil 500 mg by mouth two times per day. After 1 month of treatment, her dyspnoea had stabilised and her mechanic's hands resolved.

Antisynthetase syndrome is characterised by the presence of an anti-aminoacyl tRNA synthetase antibody (in this case, anti-EJ antibody) with one or more of the following clinical features: Raynaud's phenomenon, arthritis, mechanic's hands or hiker's feet, interstitial lung disease, fever and myositis.¹ Interstitial lung disease is a prominent internal organ manifestation of the antisynthetase syndrome.^{2,3} Mechanic's hands is a cutaneous manifestation consisting of hyperkeratosis and cracking of the hands and fingertips. Hiker's feet is the same phenomenon occurring on the skin of the feet and tips of the toes.⁴ These skin findings should raise clinical suspicion for antisynthetase syndrome and prompt an investigation of anti-aminoacyl tRNA synthetase antibodies as well as a thorough history to determine if other manifestations of the disease

Figure 2 The patient's CT scan of the chest showing interstitial lung disease.

are present. The Connor's criteria and the Solomon's criteria are both useful diagnostic tools that clinicians can use in cases where antisynthetase syndrome is suspected.^{2,3} The presence of an anti-aminoacyl tRNA synthetase antibody is required for diagnosis in both of these criteria.

Learning points

- ▶ The presence of an anti-aminoacyl tRNA synthetase antibody and one or more of the following clinical features should raise suspicion for antisynthetase syndrome: Raynaud's phenomenon, arthritis, mechanic's hands or hiker's feet, interstitial lung disease, fever and myositis.
- ▶ Mechanic's hands and hiker's feet should raise suspicion for antisynthetase syndrome.
- ▶ Interstitial lung disease is a prominent feature of antisynthetase syndrome.
- ▶ The Connor's criteria and the Solomon's criteria are useful tools for aiding in the diagnosis of antisynthetase syndrome.

Figure 1 The patient's hands with cracking at the distal fingertips and cracking of the skin on the lateral and medial sides of the patient's fingers characteristic of mechanic's hands.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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