Erdheim-Chester Disease and the lungs

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What is the problem?

Histiocyte

THIS CELL ISN'T RIGHT! I MUST DESTROY IT QUICKLY BEFORE IT RELEASES A BUNCH OF NEW VIRUSES.
What is the problem?

• Too many histiocytes (cells which normally fight infections)

• Can infiltrate all or some organs including the lungs

• Causes scar-like tissue

• This makes it hard for oxygen to get from the air into the lungs, which can make it hard to breathe
How many patients have lung involvement?

- One-quarter to one-half of patients will have pulmonary involvement
  - Pleural involvement
  - Parenchymal involvement
What are the symptoms?

- Cough
- Shortness of breath
- Fatigue
- Chest pain
What testing do I need?

- Pulmonary function test
- 6 minute walk test
- Imaging
- Bronchoscopy
Pulmonary function test (breathing test)

- Pulmonary function testing measures how well you are breathing
- A Complete Pulmonary Function Test often takes 1 ½ hours to complete and can include:
  - Spirometry
  - Lung volumes measurements
  - Diffusing capacity
Pulmonary function test—spirometry

- It involves breathing in as deeply as you can, and then breathing out as hard and as fast as you can into a tube.
Pulmonary function test - lung volumes

- This test measures how much air is in your lungs. In ECD there is a decrease in the amount of air in your lungs, also called restriction.

- The test involves sitting in a large glass box that looks like a phone booth. While you are in this box, you will breathe in and out through a tube.
Pulmonary function test—diffusing capacity

- This test measures how well oxygen gets from your lungs into your blood. It involves breathing in a certain gas, and then breathing out into a tube.
6 minute walk test

- This test measures how far you can walk in 6 minutes. It also measures how much oxygen is in your blood before and after you walk for 6 minutes.
- While you walk, you will wear a sensor on your finger that measures how much oxygen is in your blood.
- Healthy subjects can typically walk 400 to 700 m.
Imaging

• Chest x-ray
• Computerized tomography (CT) scan
Chest X-ray

• The lung damage associated with ECD disease often shows up in characteristic patterns on chest X-rays

• Occasionally, the chest X-ray is normal
Glass Test Tube

Air

Fat

Water

Bone

Water + Metal
Computerized tomography (CT) scan

- CT scanners use a computer to combine X-ray images taken from many different angles to produce cross-sectional images of internal structures.
Bronchoscopy

• In this procedure, we remove fluid or a very small tissue sample — generally no larger than the head of a pin — using a small, flexible tube (bronchoscope) that's passed through your mouth or nose into your lungs.

• The serious risks of bronchoscopic biopsy include bleeding or a deflated lung, which may require treatment.

• More common side effects are temporary sore throat and hoarseness.
Bronchoscopy with bronchoalveolar lavage

Double-contoured histiocytes in the bronchoalveolar lavage fluid of a patient with Erdheim-Chester disease. (arrow)

Diagnosis

• Pulmonary ECD is considered highly probable when

  • Presence of radiological hallmarks of interstitial lung disease in ECD patients

  • The detection of CD68(+), CD1a(−) histiocytes in bronchoalveolar lavage fluid confirms the diagnosis of pulmonary ECD

Treatment

- Not all patients with ECD disease require treatment
- Treatment varies with most patients undergoing several kinds of treatments
- Pulmonary involvement is not the main indication for therapy
- Symptoms due to pleural effusion, low oxygen or cough may need symptomatic treatment
Pleural effusion - fluid around the lungs

• Draining pleural effusion - Thoracentesis
Pleural effusion - fluid around the lungs
Low oxygen

- Supplemental oxygen
- When oxygen is below 88-89%
Oxygen therapy

• Using oxygen can't stop lung damage, but it can:
• Make breathing and exercise easier
• Prevent or lessen complications from low blood oxygen levels
• Reduce blood pressure in the right side of your heart
• Improve your sleep and sense of well-being
• You're most likely to receive oxygen when you sleep or exercise, although some people may use it round-the-clock
Cough

• Cough suppressants
  • Over the counter
  • Prescription
Prognosis

- The researchers concluded that pulmonary involvement of ECD can add to the morbidity of the disease but has limited impact on the overall prognosis of the disease.
Important !
Eat well
Remain active
NO smoking !!!!!
Thank you