



ECD AND CARDIOVASCULAR MANIFESTATIONS

THE UNIVERSITY OF TEXAS
MD Anderson
Cancer Center
Making Cancer History®

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ERDHEIM-CHESTER DISEASE

DISCOVERY AND DEFINITION

Erdheim-Chester disease (ECD) is a rare non-Langerhans cell histiocytosis first described by Jakob Erdheim and William Chester in 1930

Infiltrate of lipid-laden macrophages (histiocytes), multinucleated giant cells, inflammatory infiltrate of lymphocytes and histiocytes in the bone marrow and a generalized sclerosis of the long bones sparing the epiphysis

ERDHEIM-CHESTER DISEASE

- 73% Men versus 27% women
- Mean age at diagnosis: 55 +/- 14 years (16-80)
- Very rare in the pediatric group
- Affects multiple organ systems including the cardiovascular system

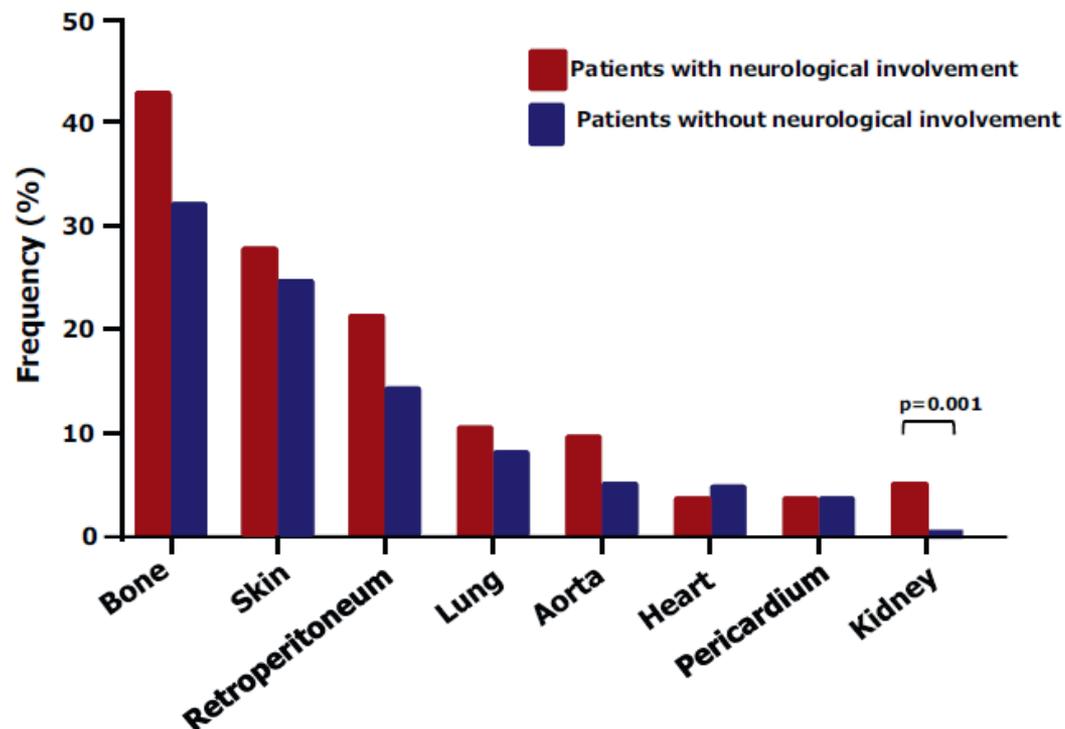


Fig. 3. Frequency of disease localizations in patients with ECD.

OBJECTIVES

1. Describe and illustrate the cardiovascular manifestations of ECD
2. Cardiac signs and symptoms in patients with ECD
3. Describe the cardiac evaluation of patients with ECD at MD Anderson Cancer Center
4. Treatment options for cardiovascular manifestations

OBJECTIVES

1. Describe and illustrate the cardiovascular manifestations of ECD

CARDIOVASCULAR MANIFESTATIONS OF ECD

Cardiovascular manifestations are now reported to be up to 70% of patients

Seem to be higher in older patients

Mean age of patients with ECD involvement of the heart is 60 years

CARDIOVASCULAR MANIFESTATIONS OF ECD

SITES OF INVOLVEMENT

Thoracic or Abdominal Aorta

Renal Arteries

Heart

- Pericardium
- Myocardium
- Endocardium
- Coronary Arteries
- Heart valves

ERDHEIM-CHESTER DISEASE

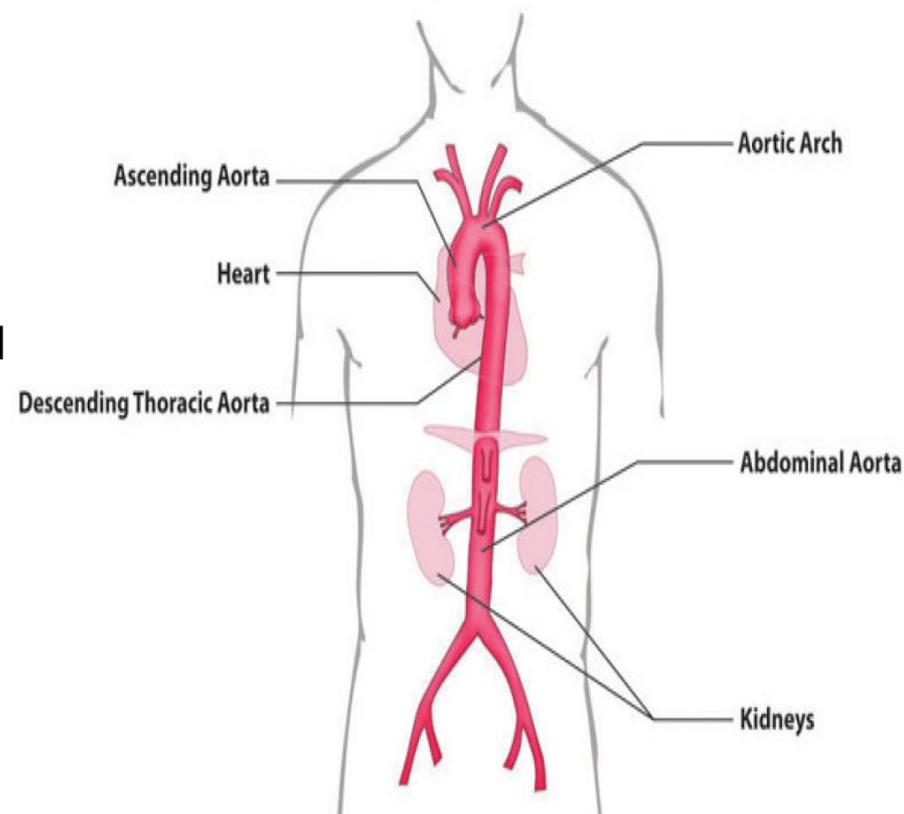
THORACIC AND ABDOMINAL AORTA

The most frequent involvement is the circumferential sheathing of the thoracic and/or abdominal aorta (66%)

When the whole aorta is sheathed, Serratrice et al. coined the term 'coated aorta' (38%)

The infiltration may often spread to the main aortic branches and effect other organs

Anatomy of the Aorta



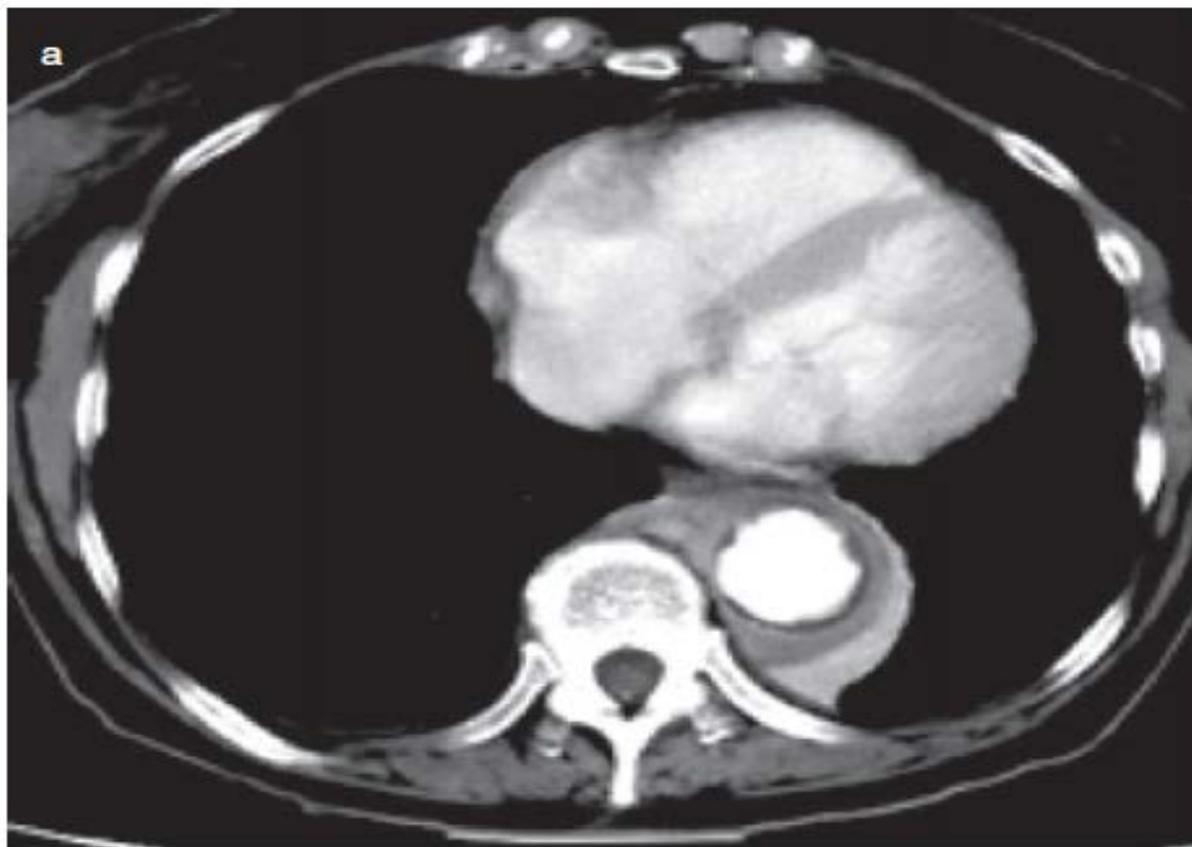
ERDHEIM-CHESTER DISEASE

CARDIAC CHARACTERISTICS- COATED AORTA



ERDHEIM-CHESTER DISEASE

CARDIAC CHARACTERISTICS- COATED AORTA



ERDHEIM-CHESTER DISEASE

RENAL ARTERY INVOLVEMENT

The periarterial infiltration may often spread to the main aortic branches

Involve the renal arteries



Fig. 1. (a) 'Coated aorta'. Computed tomography of the chest with intravenous contrast showing periaortic fibrosis of the descending thoracic aorta. (b) Computed tomography of the abdomen with oral and intravenous contrast showing perivascular fibrosis surrounding the right renal artery (arrow). Absent left kidney consistent with previous left nephrectomy.

ERDHEIM-CHESTER DISEASE

AORTA AND KIDNEYS



ECD infiltration encircling the abdominal aorta (arrow) and also surrounding both kidneys (arrowheads).

ERDHEIM-CHESTER DISEASE

HEART INVOLVEMENT

ECD can involve the:

- Pericardium (most frequent, 42%)
- Myocardium
- Endocardium

Pericardial involvement can sometimes be complicated by fluid accumulation around the heart

- pericardial effusion
- tamponade

ERDHEIM-CHESTER DISEASE

HEART INVOLVEMENT

Retrospective review of 37 patients who underwent cardiovascular screening (MRI and/or heart CT-scan) and found 70% of 37 patients with abnormal heart imaging:

- 49% exhibited abnormal infiltration of the right heart
- 30% of whom had a 'pseudo-tumoral' infiltration of the right atrium,
- 19% had an infiltration of the auriculoventricular sulcus

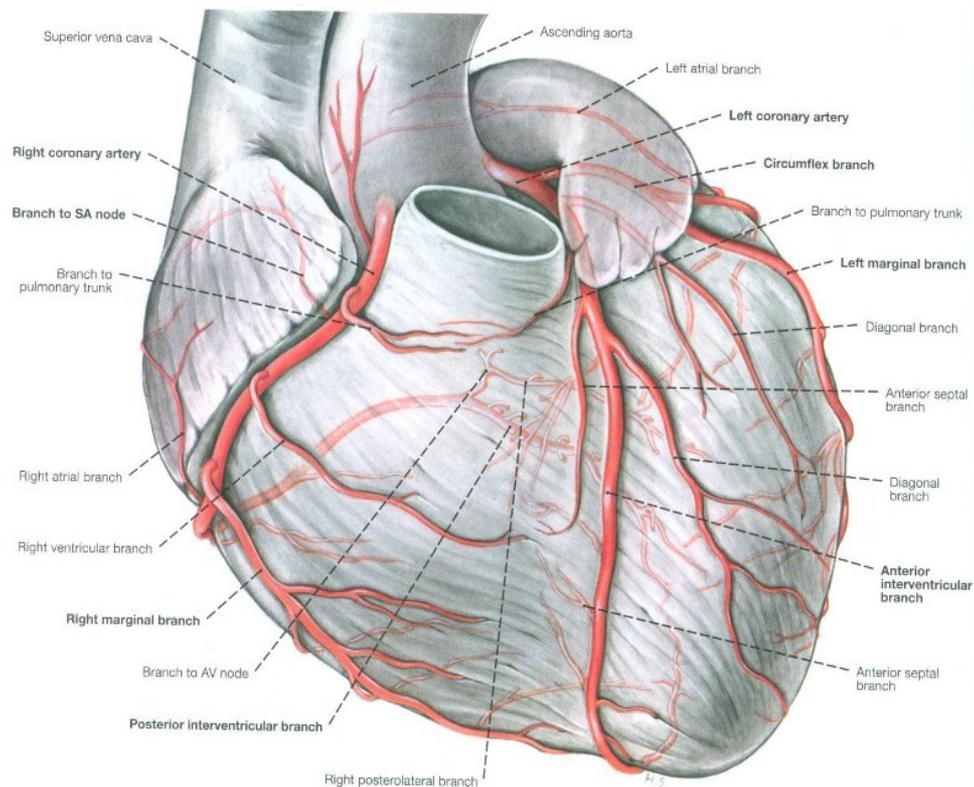
ERDHEIM-CHESTER DISEASE

CARDIAC CHARACTERISTICS

Other findings include

- Pericoronary infiltration (15 cases)
- Symptomatic aortic and mitral valve regurgitation (17%)

Involvement of the electrical system of the heart-MDACC experience

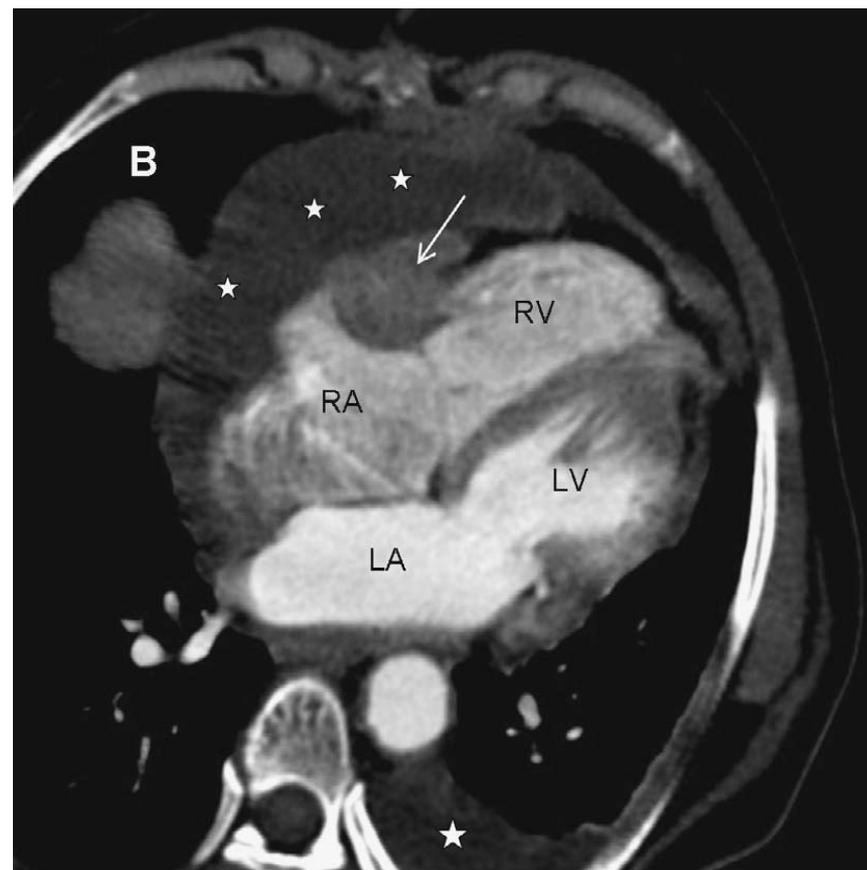


ERDHEIM-CHESTER DISEASE

HEART INVOLVEMENT

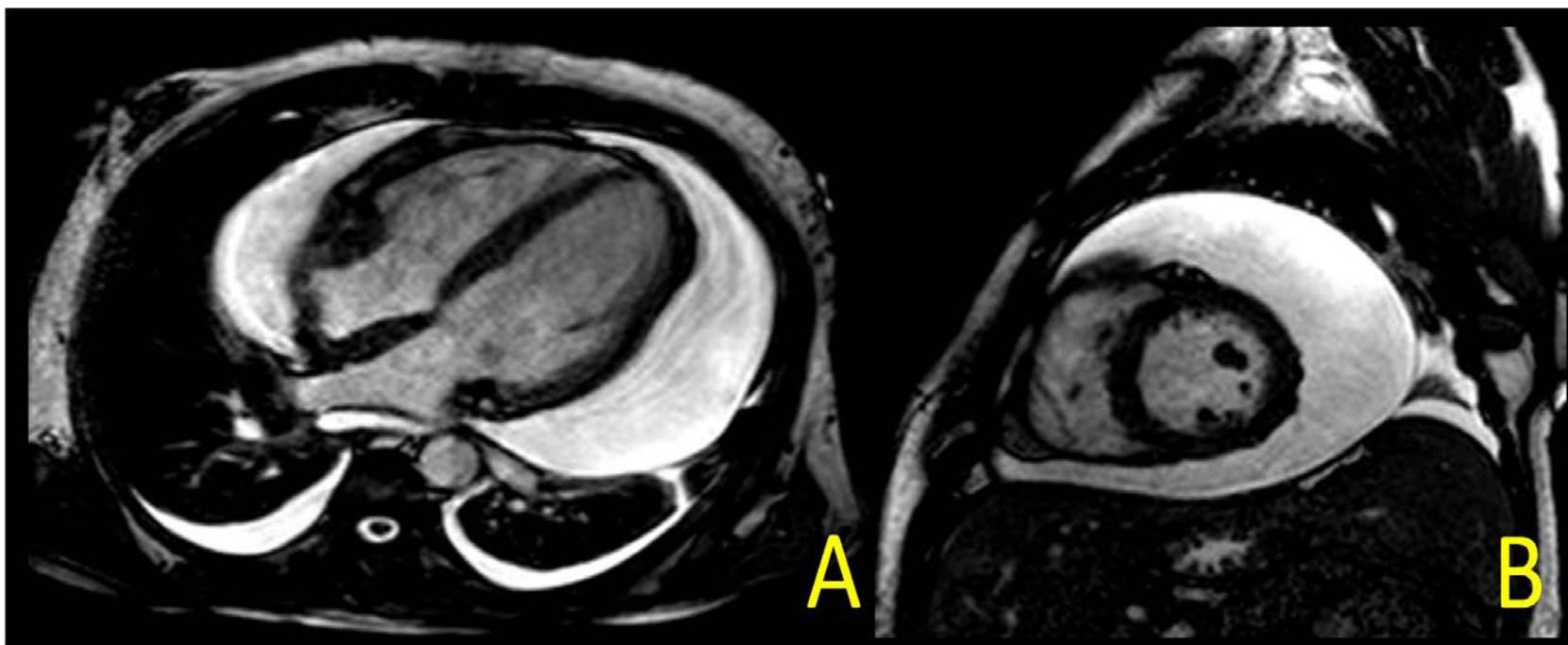
CT scan of the chest shows:

- pericardial and pleural effusion (stars)
- infiltration of the atrioventricular groove and right atrial wall as a soft-tissue pseudomass of low intensity (arrow)



ERDHEIM-CHESTER DISEASE

HEART INVOLVEMENT



Large Pericardial Effusion caused by ECD

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- 2. Cardiac signs and symptoms in patients with ECD**

ERDHEIM-CHESTER DISEASE

PATIENT SIGNS AND SYMPTOMS

Hypertension

Chest pain

Heart failure symptoms

- Shortness of breath
- Swelling

Dizziness and syncopal events

Palpitations

ERDHEIM-CHESTER DISEASE

PATIENT SIGNS AND SYMPTOMS

Hypertension

- Occurs as infiltration occurs in the main branches of the aorta
- Can involve the renal arteries
- This can cause renovascular hypertension



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ERDHEIM-CHESTER DISEASE

PATIENT SIGNS AND SYMPTOMS

Coronary artery involvement may produce **chest pain** symptoms

Pericardial involvement which can result in pericardial effusion that results in **shortness of breath**

Symptomatic leaky aortic and mitral valves which cause shortness of breath and **retaining fluid**

OBJECTIVES

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3. **Describe the cardiac evaluation of patients with ECD at MD Anderson Cancer Center**

MDACC CARDIAC EVALUATION

IMPORTANT POINTS

- Comprehensive cardiac evaluation using multimodality imaging/studies understanding the cardiac manifestations of ECD
- Systematic cardiac evaluation of manifestations in ECD that are not always clinically evident

MDACC CARDIAC EVALUATION

STUDIES PERFORMED

- **ECG:** to look for arrhythmias, heart block, cardiac intervals
- **Echocardiograms:** to assess heart function and valves
- **CT, MRI, and/or PET scans** for disease localization, degree of involvement, and progression
- **Stress test and heart catheterizations:** in patients with chest pain
- **Cardiac monitors:** to evaluate of hearth rhythm abnormalities

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ERDHEIM-CHESTER DISEASE

TREATMENT GOAL FOR CARDIOVASCULAR MANIFESTATIONS

Goal directed: Improve the symptoms and lifestyle of our ECD patients

Try to reduce further cardiac symptoms in our patients with close monitoring

- Comprehensive and repeated clinical evaluation
- Supplemented by imaging studies

ERDHEIM-CHESTER DISEASE

TREATMENT OPTIONS AT MDACC/MHM

Hypertension

- medical management
- Screening for renal artery stenosis
- Renal artery stenting

Coronary artery involvement

- Cardiac stenting
- Bypass surgery

Pericardial involvement

- Pericardiocentesis or pericardial window (minimally invasive procedure has been described) to remove fluid around the heart

ERDHEIM-CHESTER DISEASE

TREATMENT OPTIONS AT MDACC/MHM

Heart Valve involvement

- Valve Replacement

Heart rhythm abnormalities

- Pacemaker placement
- Heart ablation procedures to eradicate abnormal heart rhythms

CONCLUSION

ECD has a myriad of cardiovascular manifestations that we see at MDACC

We use a comprehensive clinical and multimodality imaging evaluation for our patients with ECD

We can offer a wide range of treatments to our patients to improve symptoms and decrease cardiovascular complications

THANK YOU

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