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

Haroche et al. 2004; CV involvement, an overlooked feature of ECD. Medicine83(6) 317-392
CARDIOVASCULAR MANIFESTATIONS OF ECD
SITES OF INVOLVEMENT

Thoracic or Abdominal Aorta
Renal Arteries
Heart
  ▪ Pericardium
  ▪ Myocardium
  ▪ Endocaridum
  ▪ 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

ERDHEIM-CHESTER DISEASE
CARDIAC CHARACTERISTICS- COATED AORTA

ERDHEIM-CHESTER DISEASE
CARDIAC CHARACTERISTICS- COATED AORTA

ERDHEIM-CHESTER DISEASE

RENAAL 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.


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

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)

Dris et al. Cardiac Erdheim-Chester. Inter Med 48: 83-84, 2009)
ERDHEIM-CHESTER DISEASE
HEART INVOLVEMENT

Large Pericardial Effusion caused by ECD

Rossi E, et al. Cardiovascular involvement in Erdheim-Chester Disease: a Magnetic Resonance Imaging study on seven patients ECR 2014/c-0662
OBJECTIVES

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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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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**.
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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 PREFORMED

- **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 catherizations**: 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
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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