

# Clinico-pathological conference



*Ramon Pujol*

**ESIM**

**Saas Fee, 18<sup>th</sup> January 2011**



## **Presentation of case**

- Male, 77 y.
- Severe epilepsy since 10 y.-old
- Post-op. pulmonary embolism, 50 y.-old
- Chronic bone pain
- Small-cell lymphoma, 9 m. before. Clinical improvement on Chlorambucil

# **Presentation of case**

- Bilateral pleural effusion
- Generalized edema
- Xanthelasma
- EKG, low diffuse QRS voltage
- Lab tests
  - Acute phase reactants
  - Hypoalbuminemia
  - Hypoxemia

# **Presentation of case**

- Pleural fluid, exudate, inflammatory, cultures neg.
- Pericardial fluid, similar
- Pneumothorax & pericardial tamponade
- Died after 4 weeks

Figure 1. Chest X-ray

n: 1

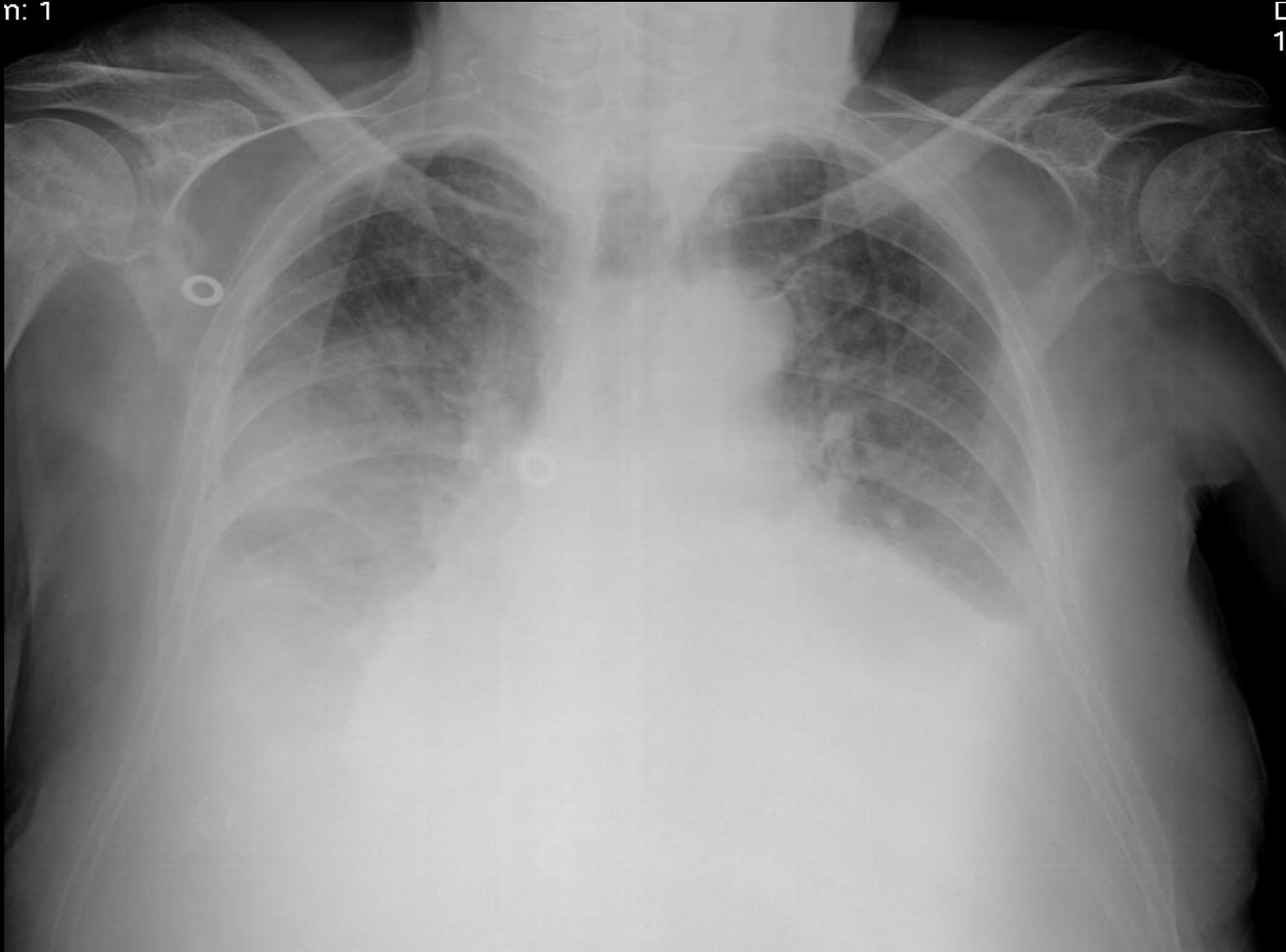


Figure 2. CT-scan

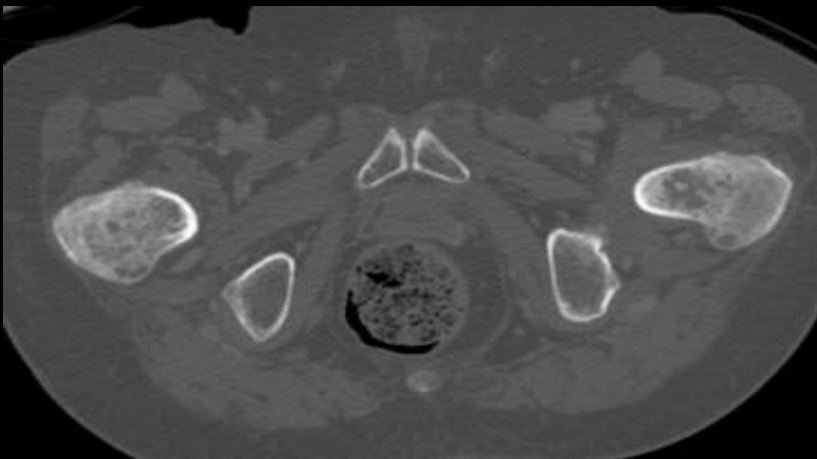
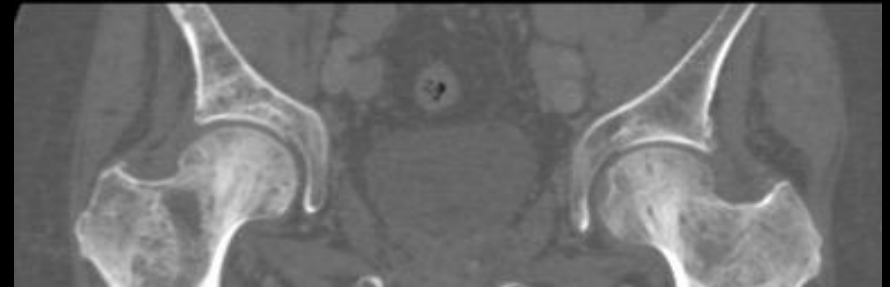
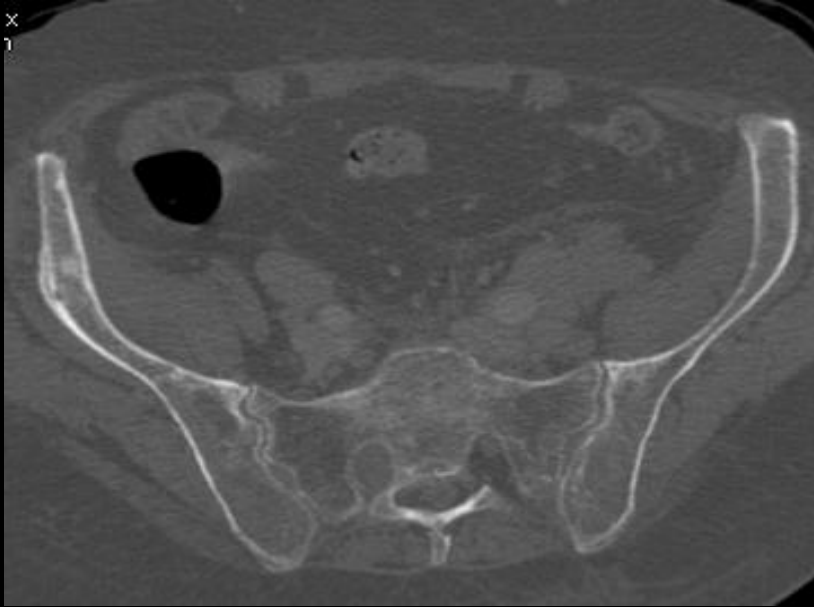


Figure 2. CT-scan



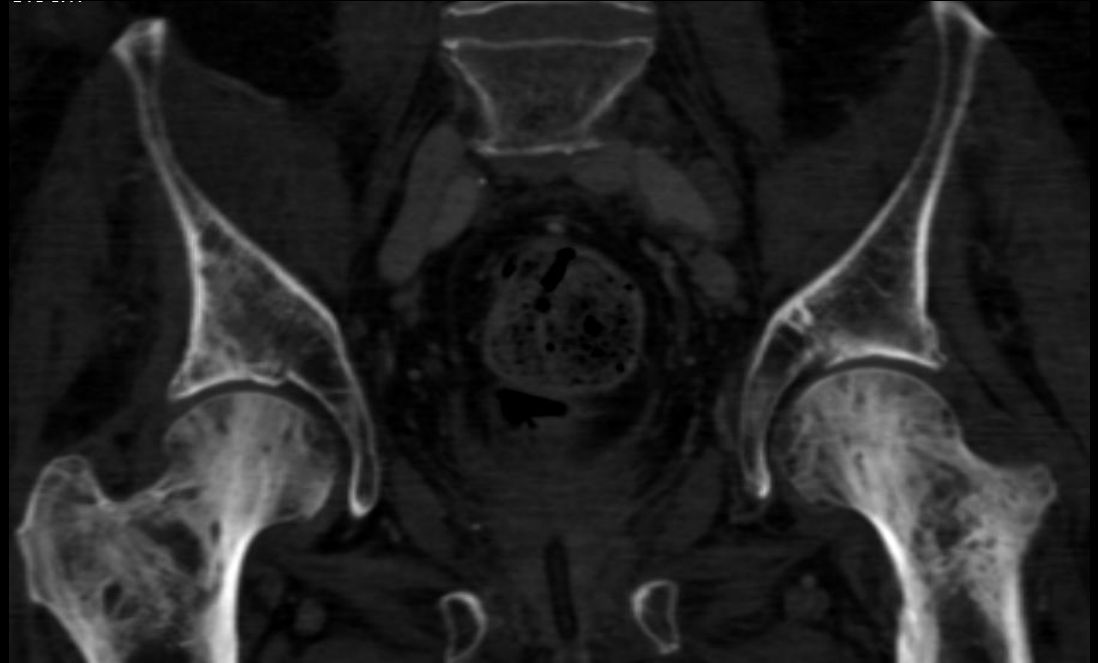
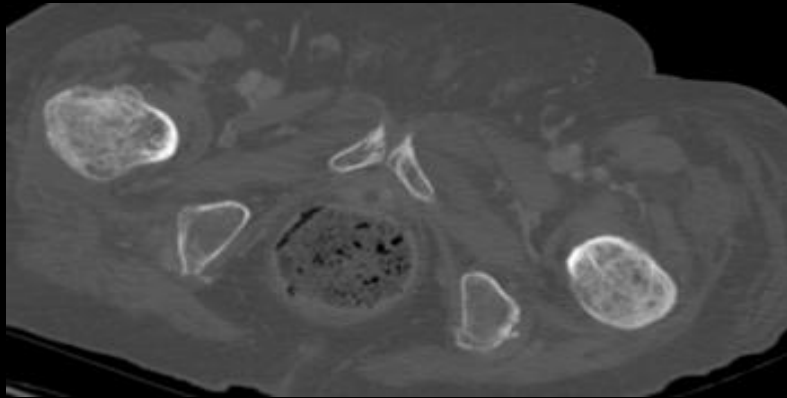
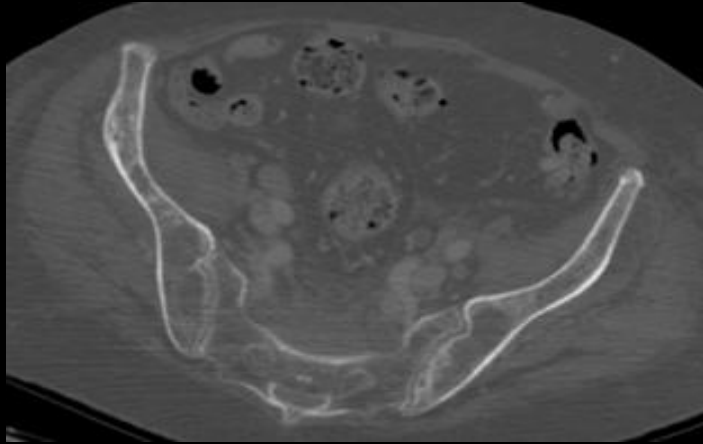


CT-scan Prior to admission. Bone sections.  
February 2009





CT-scan Prior to admission. December 2009  
PELVIS-FEMUR AXIAL-SAGITAL



# Autopsy findings

## Dr. Xavier Sanjuan



Generalitat de Catalunya  
**Departament de Salut**

 **Bellvitge**  
Hospital

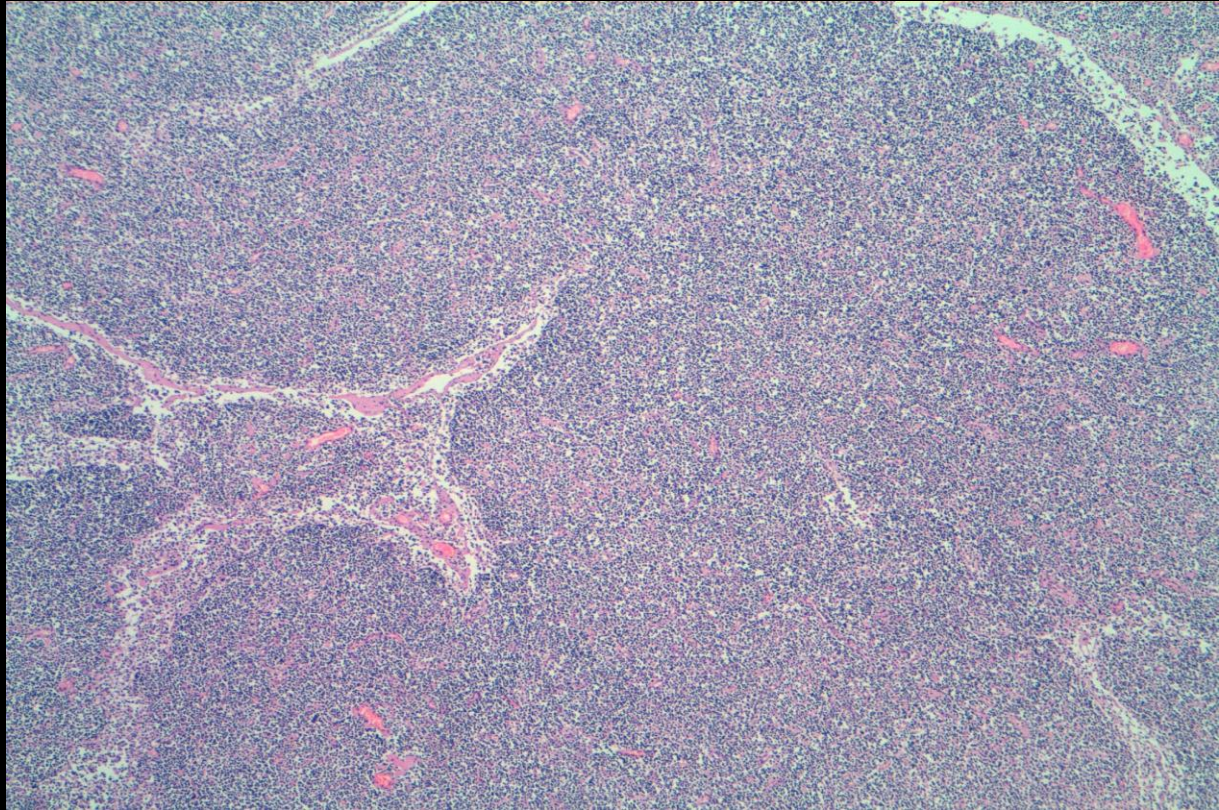
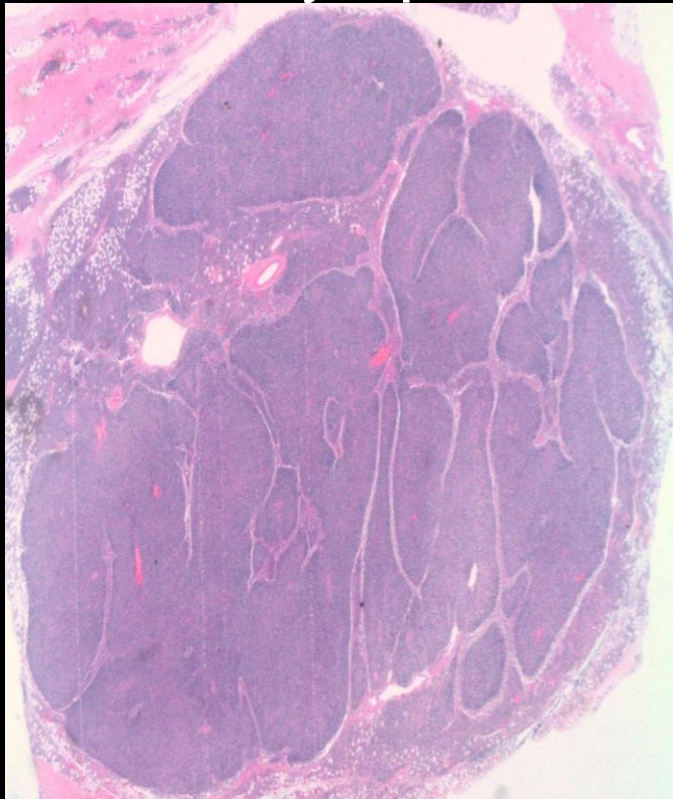
 Institut Català  
de la Salut



# A10-00016

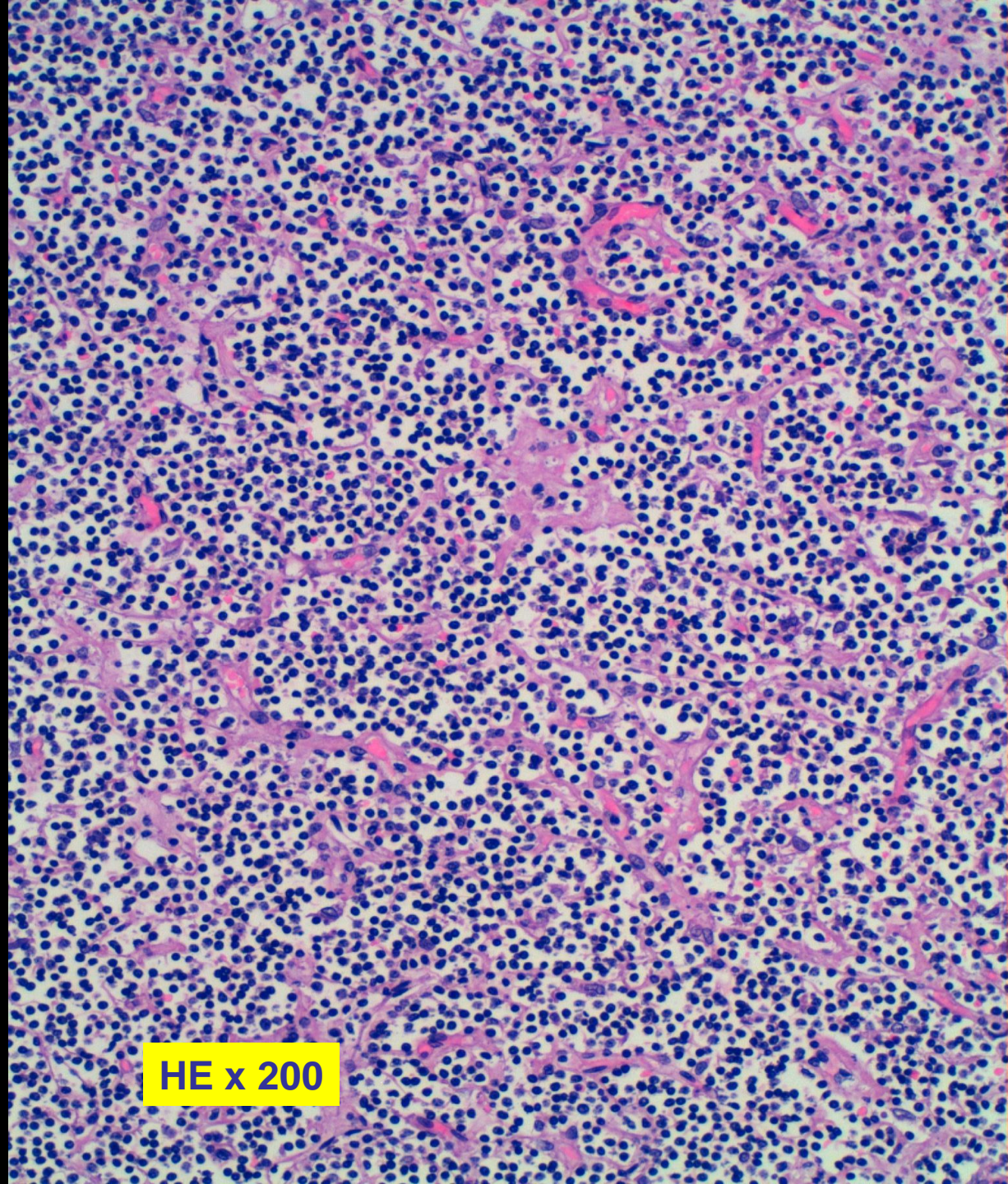
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- **Small-cell Lymphocytic Lymphoma**
  - Paravertebral (Thoracic and lumbar) & Mesenteric Lymphadenopathies



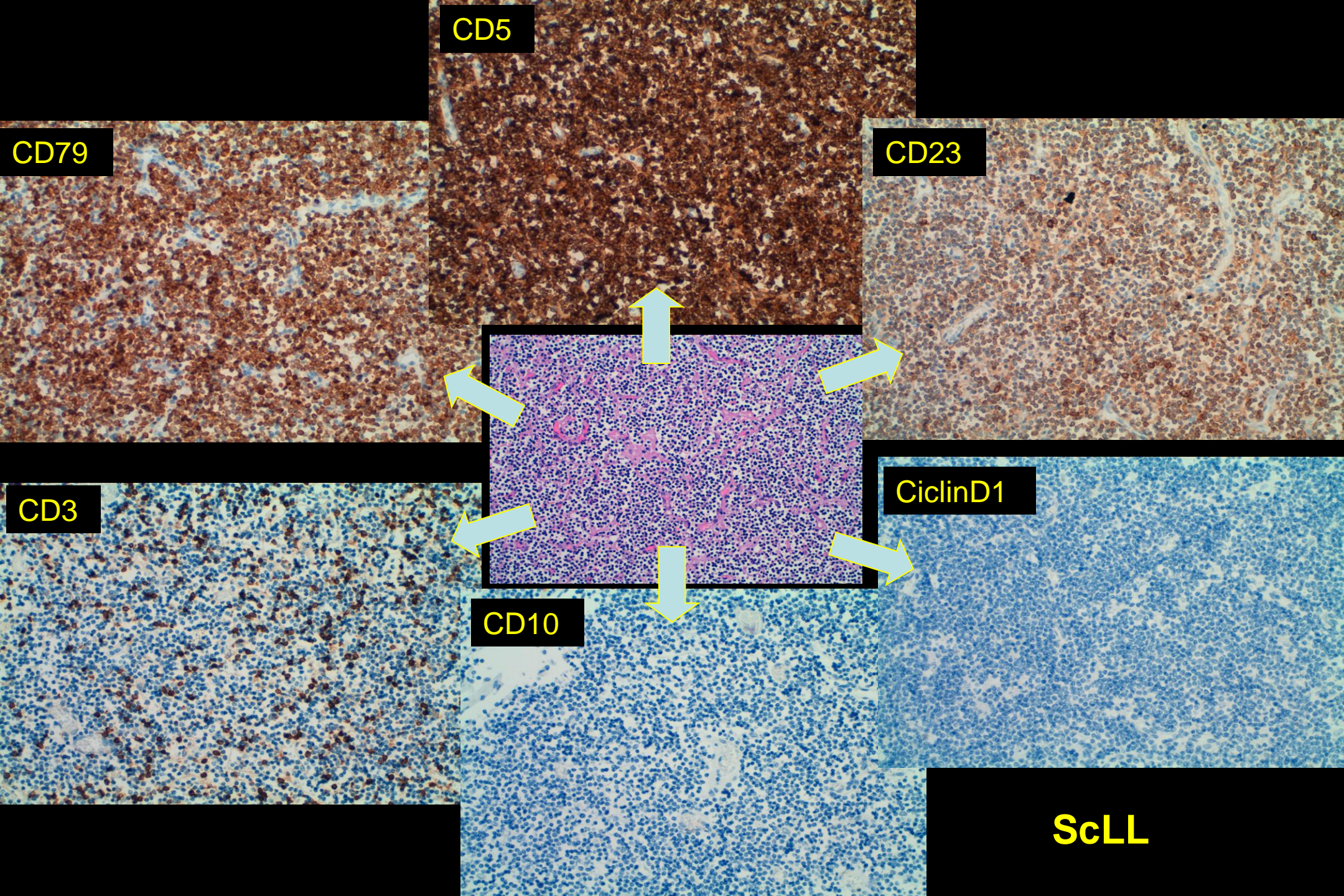


# Retroperitoneal Lymphadenopathy



HE x 200







- Erdheim-Chester Disease

- Bone marrow
- Pericardial effusion (250 ml) & infiltration of pericardium and epicardial fat
- Perirenal and periadrenal
- Meningeal enhancement and tumoral nodules
- Pleural effusion (R 150 ml; L 100 ml) and infiltration of visceral pleura

# Erdheim-Chester Disease

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- First report in 1930 by Chester
- Non Langerhans form of histiocytosis
- Over 300 cases reported so far

## Classification of Histiocytosis syndromes

1. Langerhans-cell Histiocytosis  
(prev. Histiocytosis X)
  - Eosinophilic granuloma
  - Hand-Schüller-Christian disease
  - Letterer-Siwe disease
2. Non-Langerhans-cell Histiocytosis
  - Hemophagocytic lymphohistiocytosis
  - Rosai-Dorfman disease
  - Reticulohistocytosis
  - **Erdheim Chester disease**
3. Malignant Histiocytic disorders
  - Acute Monocytic leukemia
  - Histiocytic lymphoma
  - Malignant histiocytosis



# Histologic diagnosis

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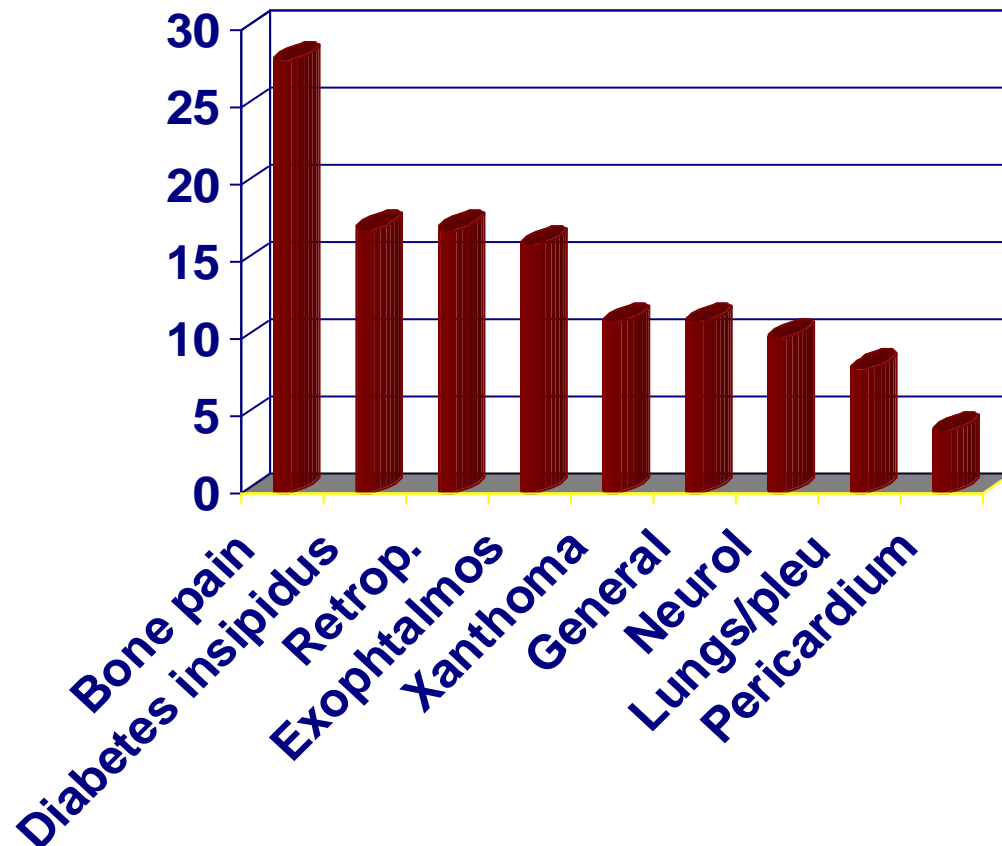
- Infiltration of foamy histiocytes nested among polymorphic granuloma and fibrosis or xanthogranulomatosis.
- CD68 +, CD1a -, S-100 protein +/-.

# Erdheim-Chester Disease.

## Review of 59 cases.

Veyssier-Belot et al. Medicine (Baltimore) 1996;75:157-

- Mean age  $53 \pm 14$  yr.
- 33 M/26 F
- Mortality 59 %  
(mean follow-up  
2,7 yr.)



# Bone lesions

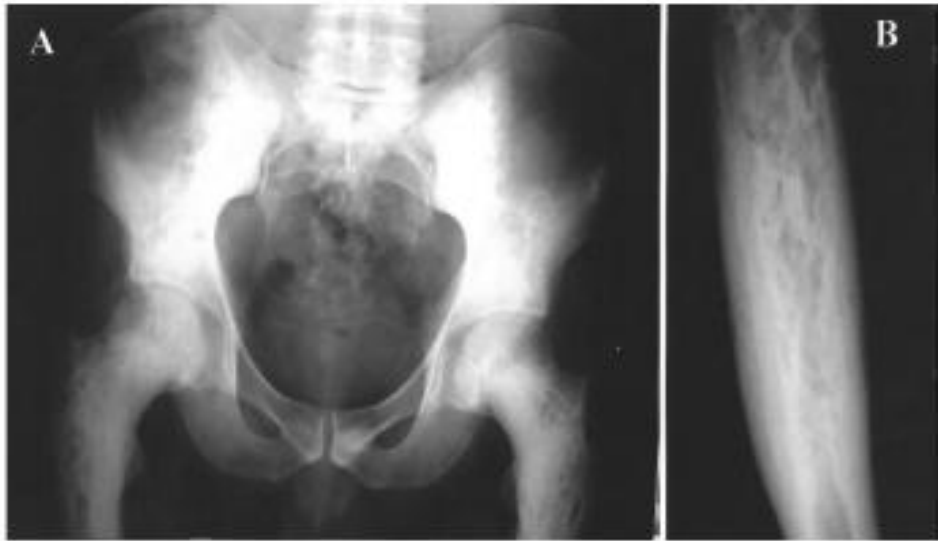
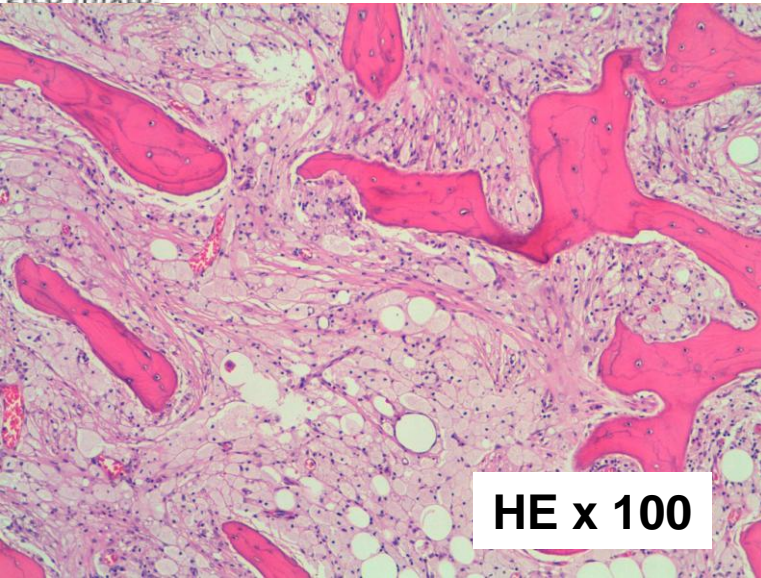
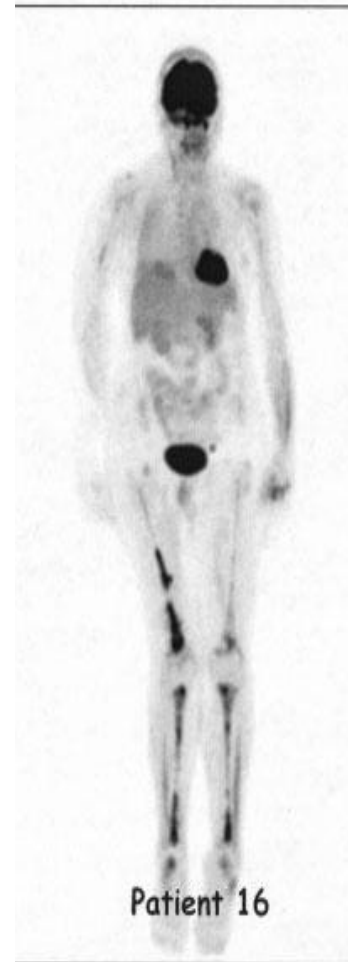


Fig. 1. A: Radiografía frontal de la pelvis del paciente en la que puede apreciarse la alternancia de áreas de hueso esclerótico con lesiones osteolíticas que involucran ambos iliacos y fémures. B: Detalle del fémur izquierdo en el que se observa un patrón patológico mixto.



- Pain in
  - Knee/leg ++++
  - Ankle/shoulder ++
  - Back/cervical +
- Osteosclerotic +++
- Osteolytic +
- Patchy increased density +



# Retroperitoneal

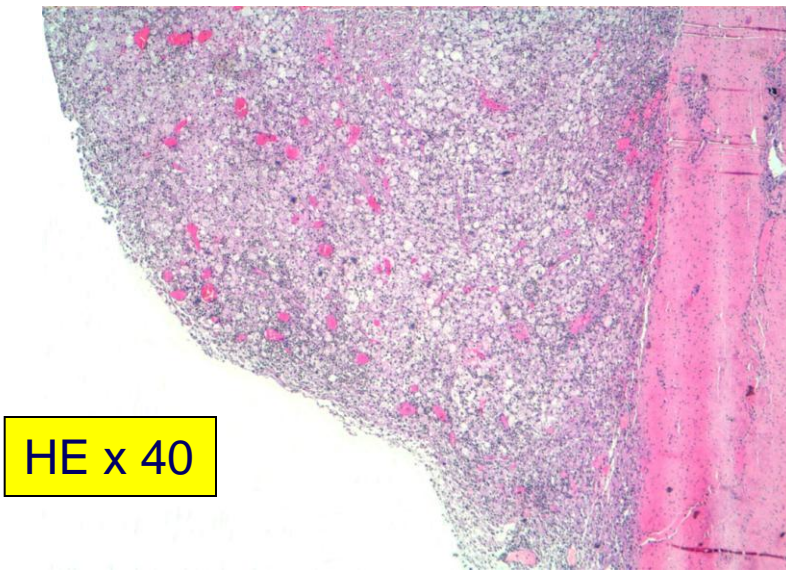
- Rarely symptomatic
- Dysuria, abdominal pain, enlarged kidneys
- CT-scan showing retroperitoneal and/or pelvic infiltration



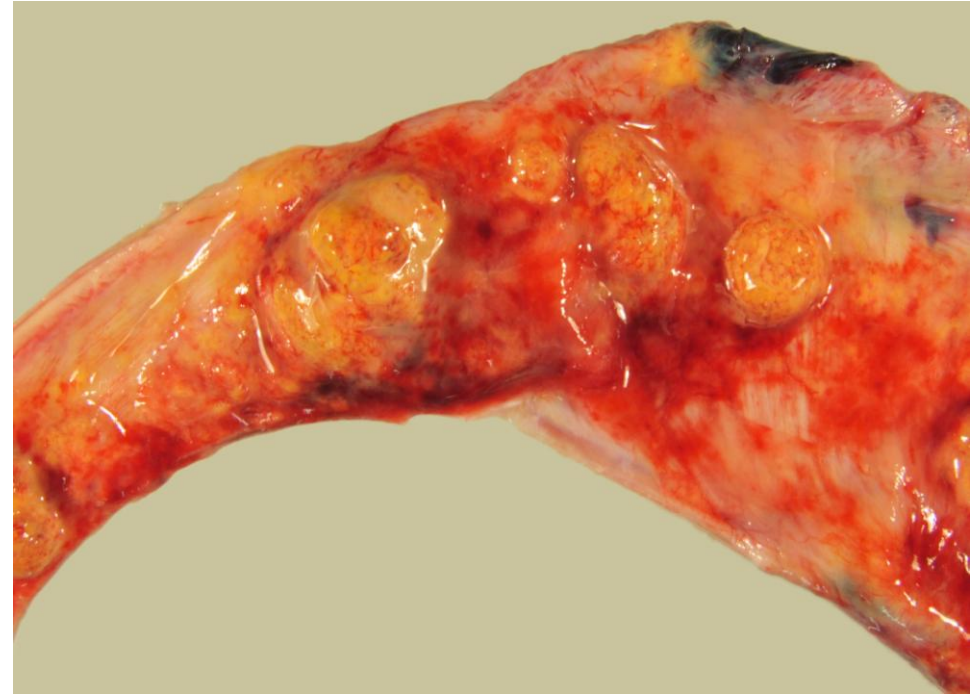


# Neurological

- Diabetes insipidus
- Epilepsy
- Change in mental status
- Stroke
- Thickening of the dura



HE x 40

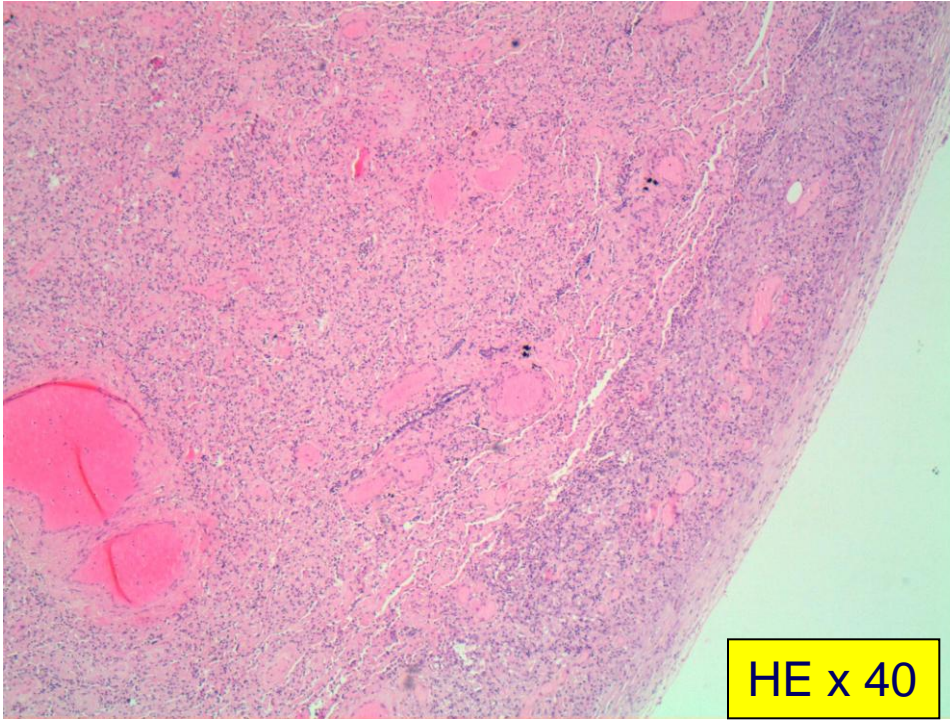


# Lungs & pleura

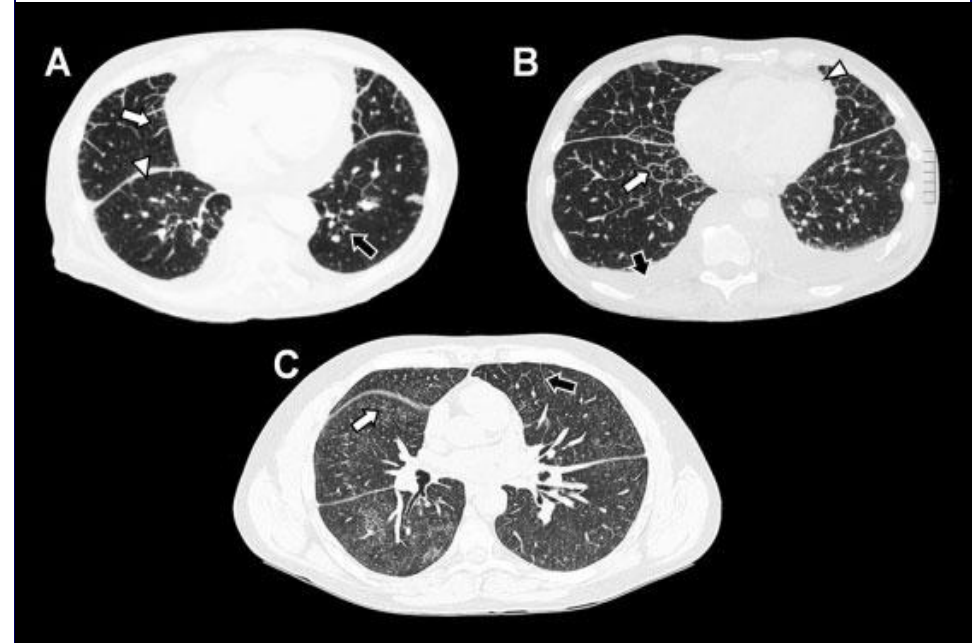
Arnaud et al. Arthritis & Rheumatism

Vol. 62, No. 11, November 2010, pp 3504–3512

- Shortness of breath, cough
- Interstitial lung disease
- Pleural effusion
- Respiratory failure



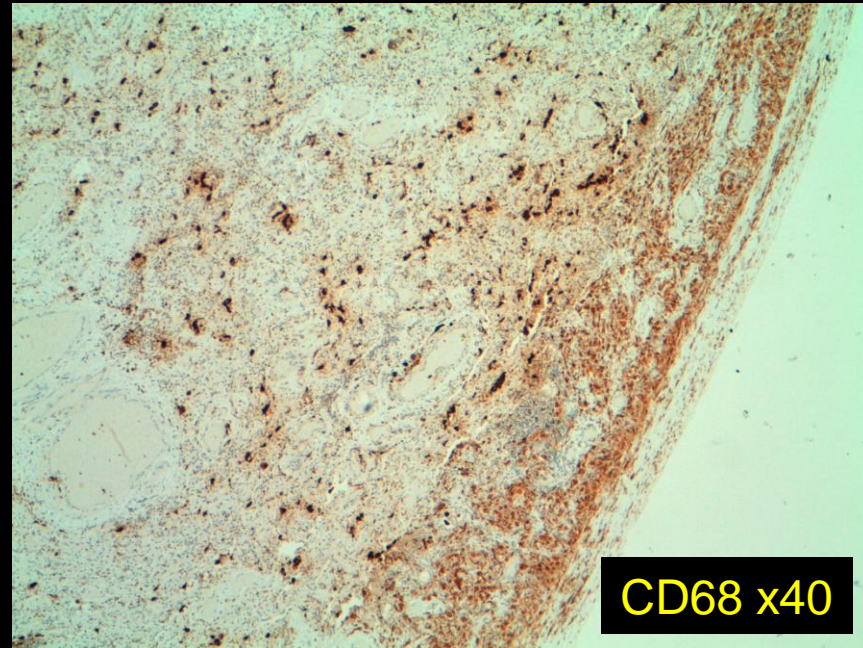
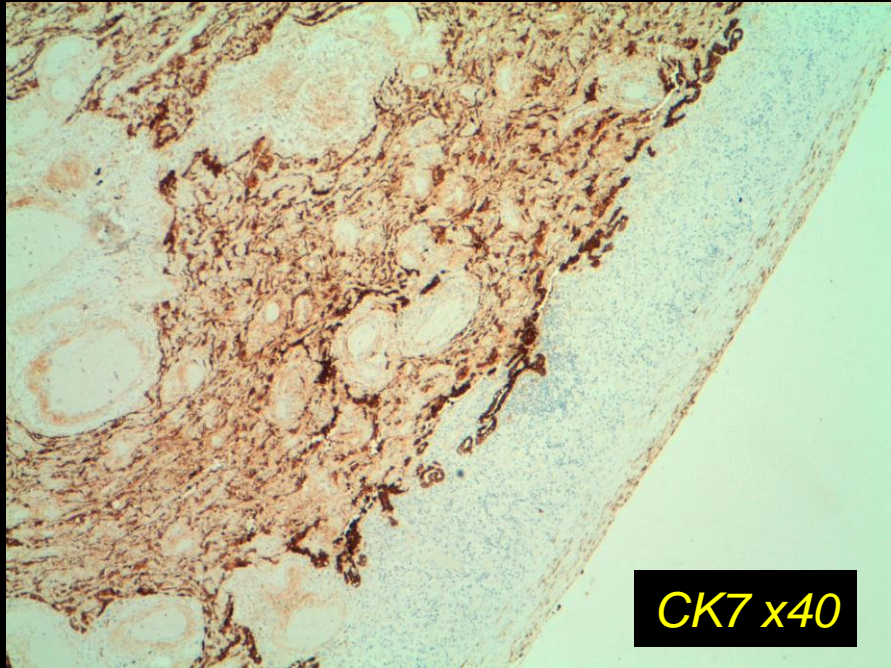
HE x 40



Interlobular septal thickening  
Thickening of the subpleural space  
Bilateral pleural effusion  
Small pericardial effusion



# Epithelial vs histiocytic markers



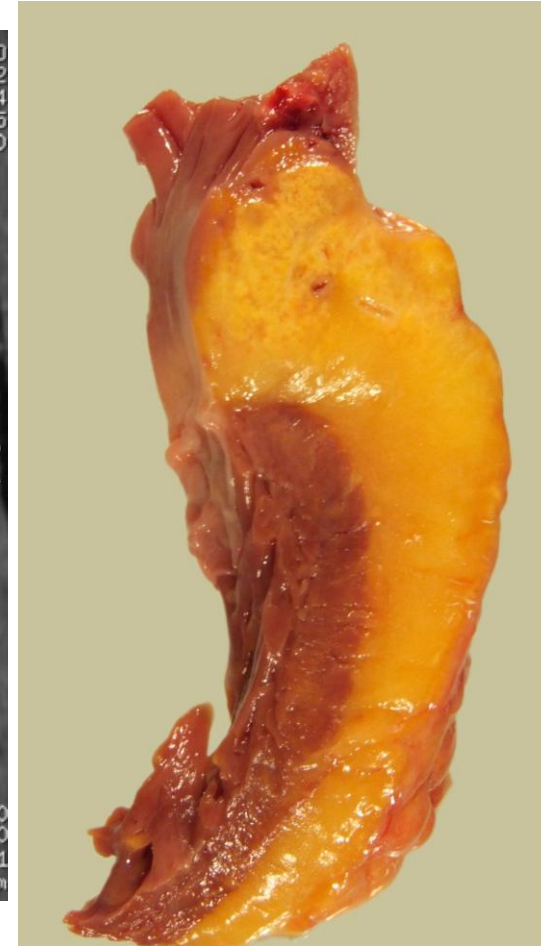


# Pericardium & large vessels

- Pericardium
- Myocardium
- Heart valves
- Coronary arteries
- Aorta & aortic branches
- Cava and pulmonary veins
- Systemic hypertension



**“coated aorta”**



**Stiff pericardium**

# Erdheim-Chester and malignancy (?)

**A. Simiele et al. Anales de Medicina Interna 2004; 21:593-**

## ABSTRACT

*Erdheim-Chester disease is an idiopathic systemic histiocytosis histologically different to Langerhans histiocytosis which is characterized by symmetric sclerotic bone lesions, predominantly affecting the diaphyseal and metaphyseal areas of large bones. It is not classified within malignant histiocytosis, but the clinical course tends to be aggressive with a poor response to different schedules of treatment. In rare cases the association of both entities has been reported: Erdheim-Chester disease and Langerhans histiocytosis.*

*We present a new case with histological data of both histiocytosis whose clinical course included bone and muscle pain, insipidus diabetes, exophthalmos, bilateral symmetrical sclerosing bone lesions and a cerebellar syndrome.*

# Erdheim-Chester and malignancy (?)

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- Association of Langerhans cell histiocytosis with malignant neoplasms.  
*Egeler RM, Neglia JP, Puccetti DM, Brennan CA, Nesbit ME*  
*Cancer 1993;71(3):865-73*
  - The relation of Langerhans cell histiocytosis to acute leukemia, lymphomas, and other solid tumors. The LCH-Malignancy Study Group of the Histiocyte Society.  
*Egeler RM, Neglia JP, Aric M, Favara BE, Heitger A, Nesbit ME, Nicholson HS*  
*Hematol Oncol Clin North Am 1998;12(2):369-78.*
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# Final message

*“ Although we tend to explain all the findings under a single diagnosis, it is quite frequent to find two or more coexisting diagnoses responsible of the patient’s disorders”*