Sarcoidosis

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Nuclear medicine, 2011

Epidemiology

- Female > Males
- 20-29 y and women over 50
- Sweden and Iceland → 60/100,000
- High risk group in the US:
  - African Americans ladies
    - More severe
- Higher prevalence in non-smokers

Initial symptoms

- The most common → restrictive lung disease symptoms
- General symptoms → fatigue, night sweats, weight loss, and erythema nodosum
- Organ specific symptoms
- As many as 50% → Asymptomatic

Clinical course

- ~ 2/3rd → remain stable or experience a remission within a decade after diagnosis, with few or no consequences thereafter.
- ~ 20% → pulmonary fibrosis.
- <5% die due to lung fibrosis with respiratory failure or cardiac or neurologic involvement

Poor prognosis

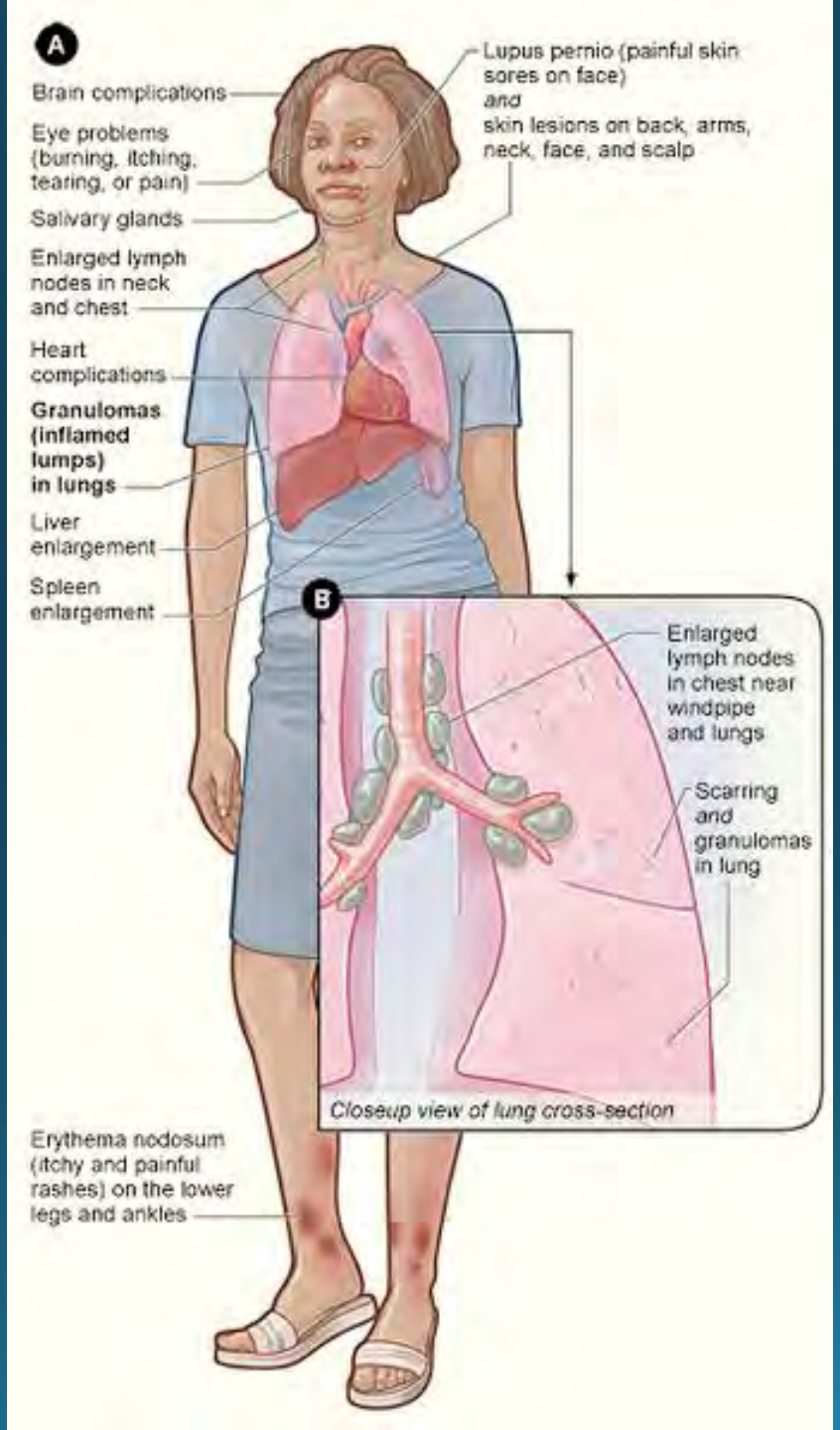
- Stage 2-3 pulmonary disease at the time of diagnosis
- Onset after the age of 40 years
- Black race
- Hypercalcemia
- Splenomegaly
- Osseous involvement
- Chronic uveitis
- Lupus pernio

Good prognosis

- Common early-stage features:
  - fever
  - polyarthritis
  - erythema nodosum
  - bilateral hilar lymph node enlargement

(Löfgren syndrome)

85% remission rate


A
Brain complications
Eye problems (burning, itching, tearing, or pain)
Salivary glands
Enlarged lymph nodes in neck and chest
Heart complications
Granulomas (inflamed lumps) in lungs
Liver enlargement
Spleen enlargement
Lupus pernio (painful skin sores on face)
and
skin lesions on back, arms, neck, face, and scalp
B
Enlarged lymph nodes in chest near windpipe and lungs
Scarring
and
granulomas in lung
Closeup view of lung cross-section
Erythema nodosum (itchy and painful rashes) on the lower legs and ankles

Different subtypes

- Annular sarcoidosis
- Erythrodermic sarcoidosis
- Ichthyosiform sarcoidosis
- Hypopigmented sarcoidosis
- Löfgren syndrome
- Lupus pernio
- Morpheaform sarcoidosis
- Mucosal sarcoidosis
- Neurosarcoidosis
- Papular sarcoid
- Scar sarcoid
- Subcutaneous sarcoidosis
- Systemic sarcoidosis
- Ulcerative sarcoidosis

Pathophysiology

- Not well understood
- Accumulation of monocytes, macrophages and activated T-lymphocytes
- TNF-alpha, IFN-gamma, and IL-12, characteristic of a Th1-response
- Increased macrophage and CD_4 helper T-cell activation → accelerated but ineffective response
- Anergy→ increased risk of infections and cancer

Diagnosis

- Compatible clinical and radiologic findings
- Non-caseous granulomas and the absence of causative organisms
- Ruling out other potential etiologies

For every patient

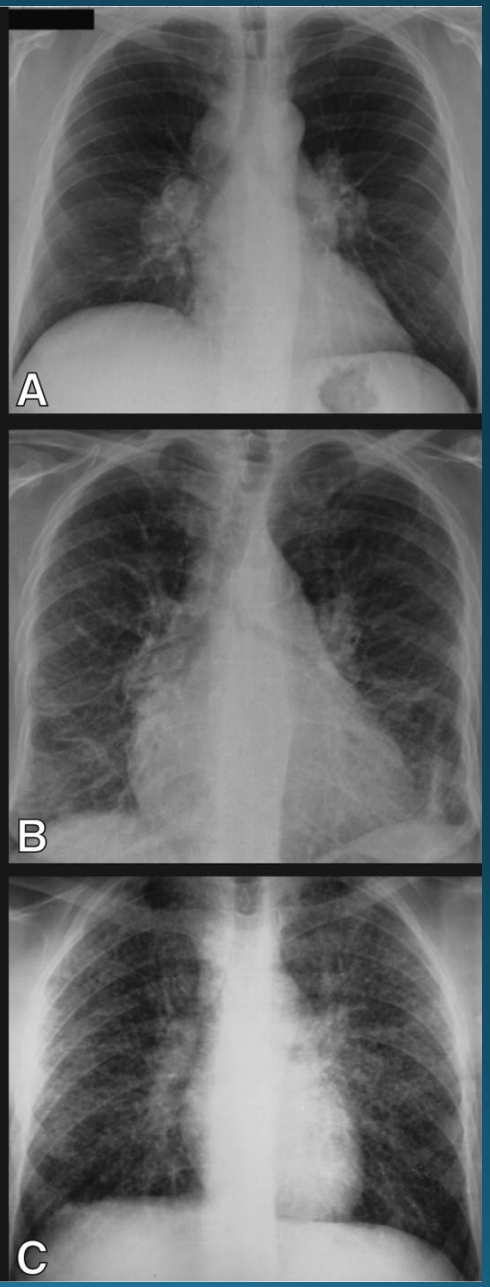
- Histologic verification
- Assessment of the extent and severity of organ involvement
- Assessment of whether disease is stable or likely to progress
- Determination of whether a patient might benefit from treatment

Lungs

- Primarily an interstitial lung disease → alveoli, small bronchi, and small blood vessels
- 90% of patients
- 50% → permanent pulmonary abnormalities
- 5 -15% → progressive fibrosis
- HRCT vs. CT

Staging of Sarcoidosis on the Basis of Chest Radiographs

STAGE 0	No abnormalities	5%–10%
STAGE 1	Lymphadenopathy (fig. A)	50%
STAGE 2	Lymphadenopathy + pulmonary infiltration (fig. B)	25%–30%
STAGE 3	Pulmonary infiltration (fig. C)	10%–12%
STAGE 4	Fibrosis	5% (up to 25% during the course of the disease)



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Typical lung findings

- Lymphadenopathy: bilateral hilar, right paratracheal
- Micro or macro nodules
- Lymphangitic spread: peribronchovascular, subpleural, interlobular septal
- Fibrotic changes
- Bilateral perihilar opacities
- **Upper- and middle-zone predominant**

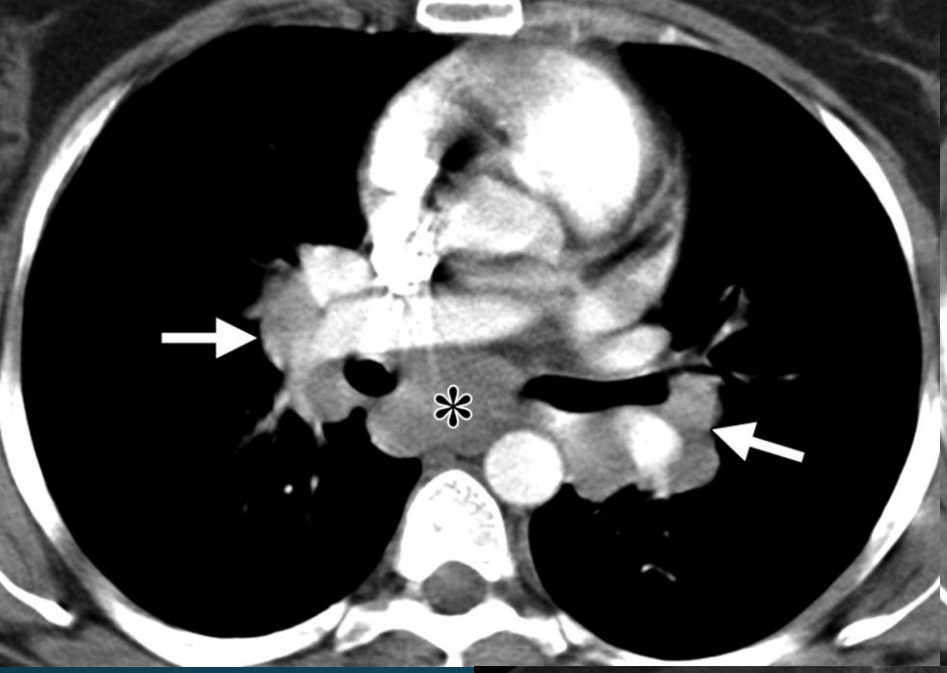
Reversible vs. irreversible lung lesions

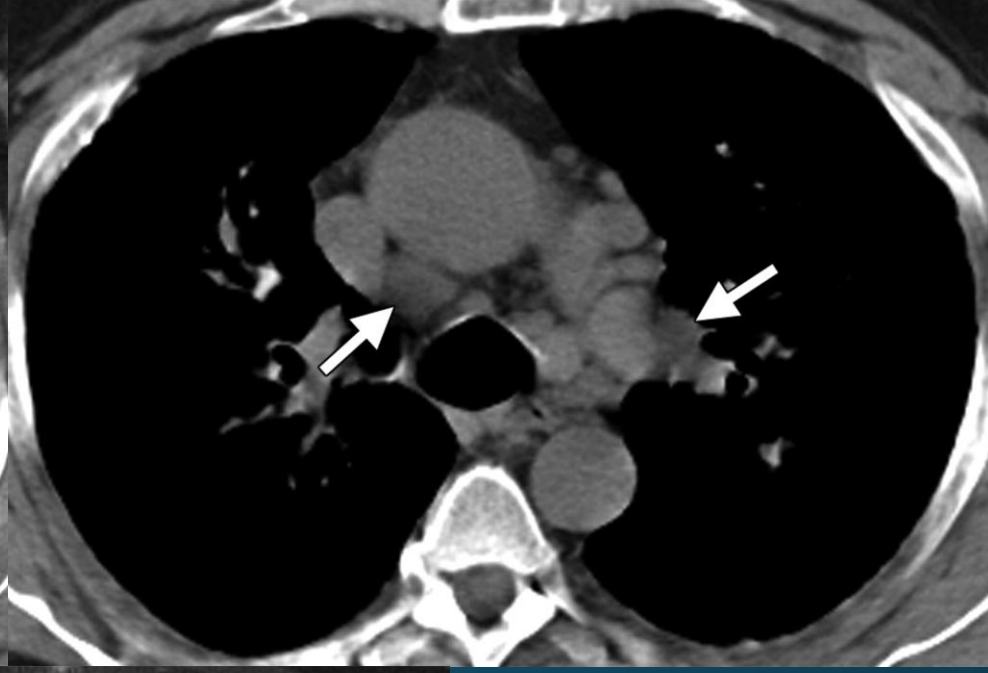
Reversible

- Micronodules, macronodules
- Consolidation
- Ground-glass opacities
- Interlobular septal thickening
- Intralobular linear opacities
-

Irreversible parenchymal abnormalities

- Honeycomb-like opacities, cysts, bullae, emphysema
- Architectural distortion
- Traction bronchiectasis, bronchiolectasis
- Volume loss in upper lobes, retraction of hila
- Mycetoma (in 10% of patients with end-stage sarcoidosis and a preexisting cavity)





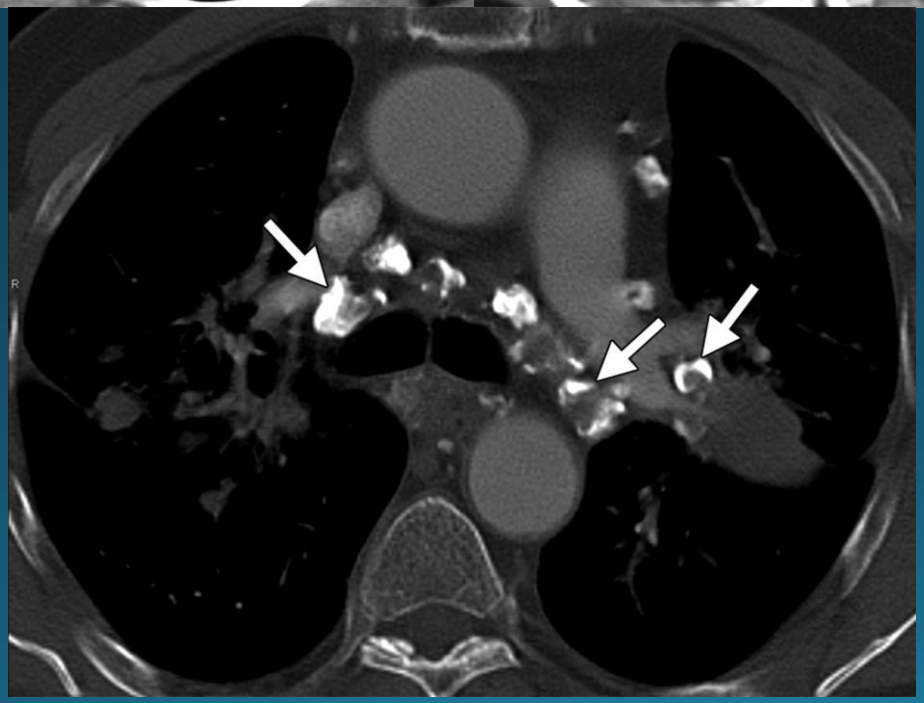


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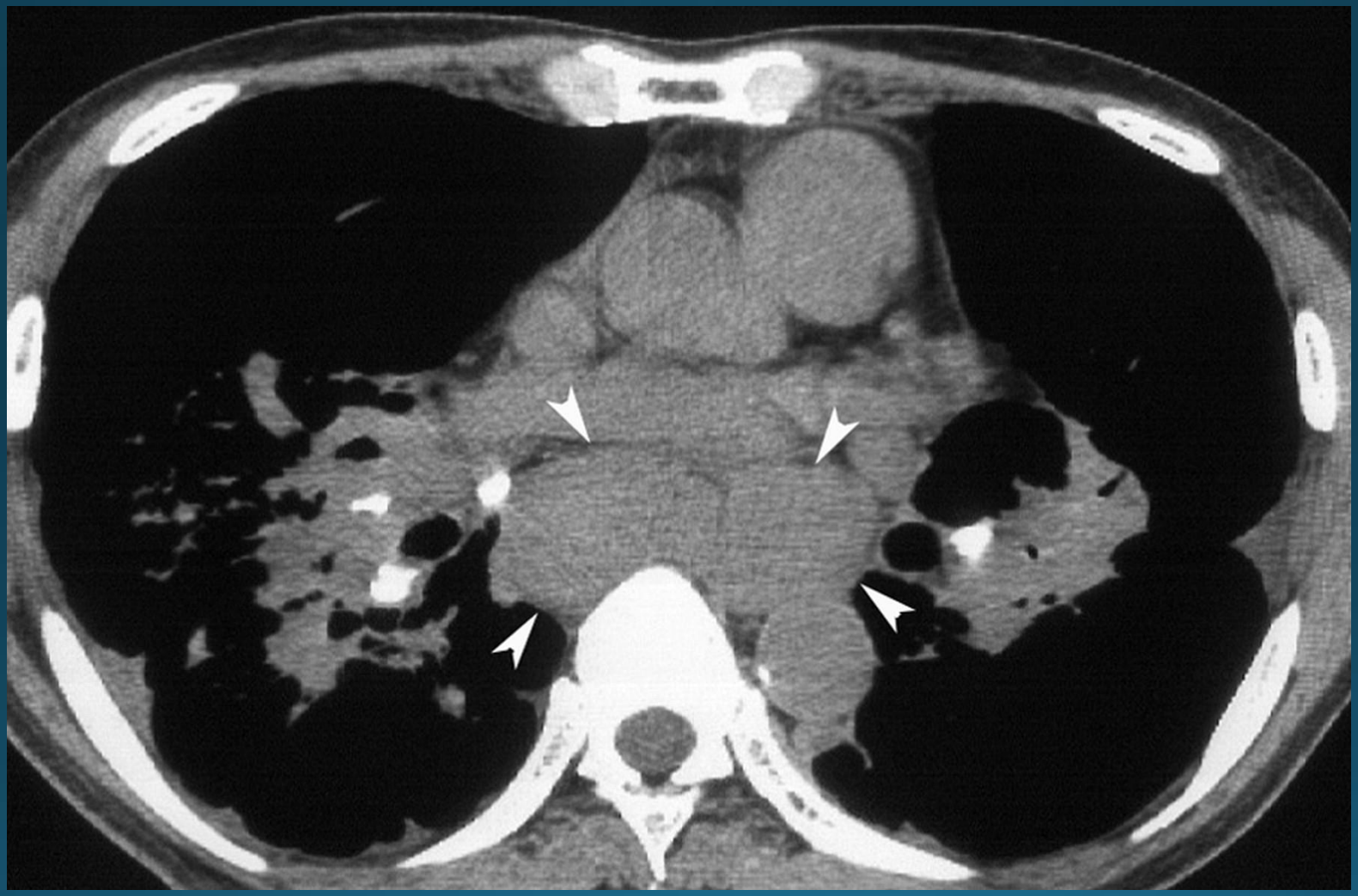
Figure 3. Mediastinal adenopathy in a 60-year-old man.



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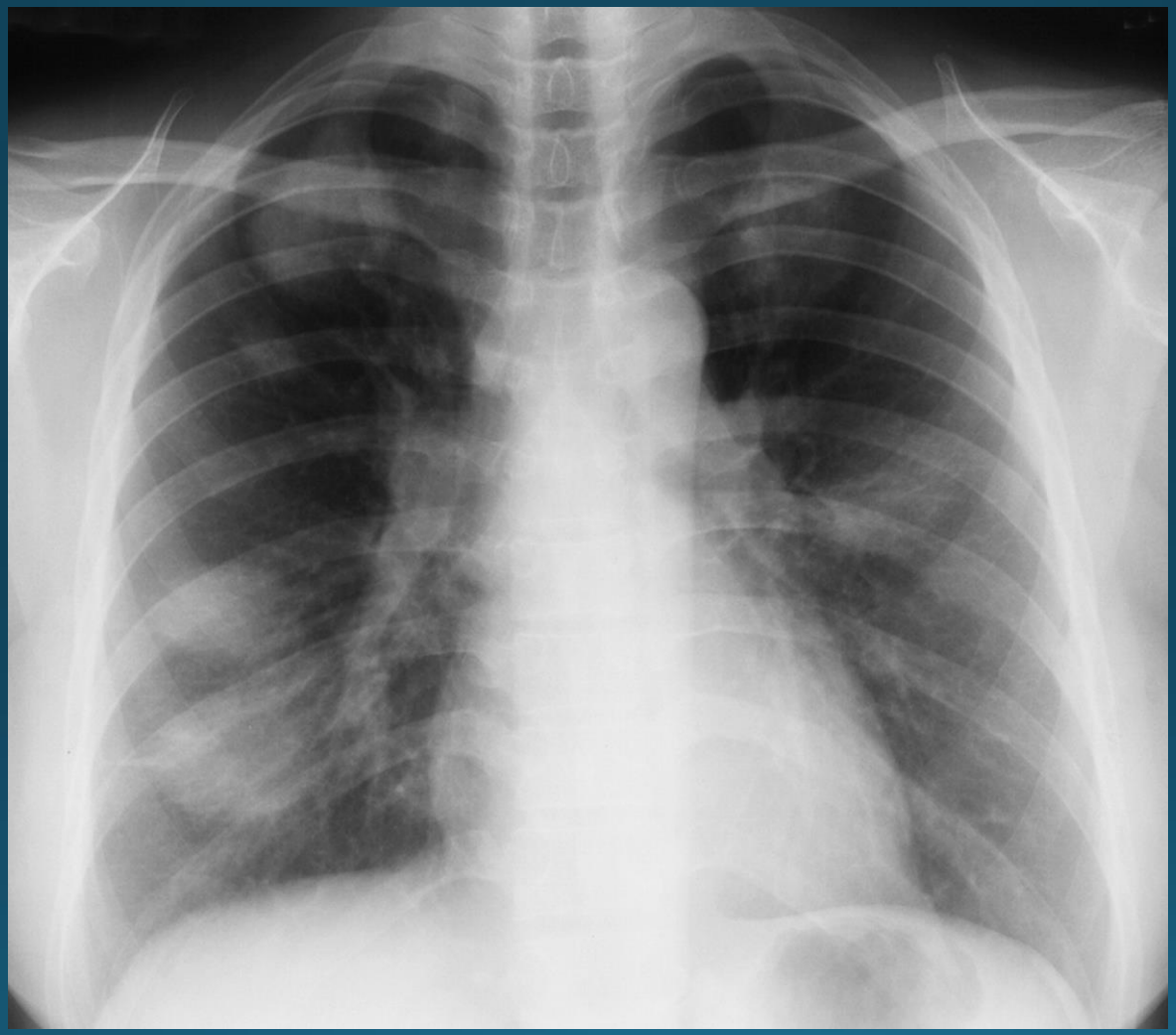
Pulmonary sarcoidosis in a 26-year-old woman.



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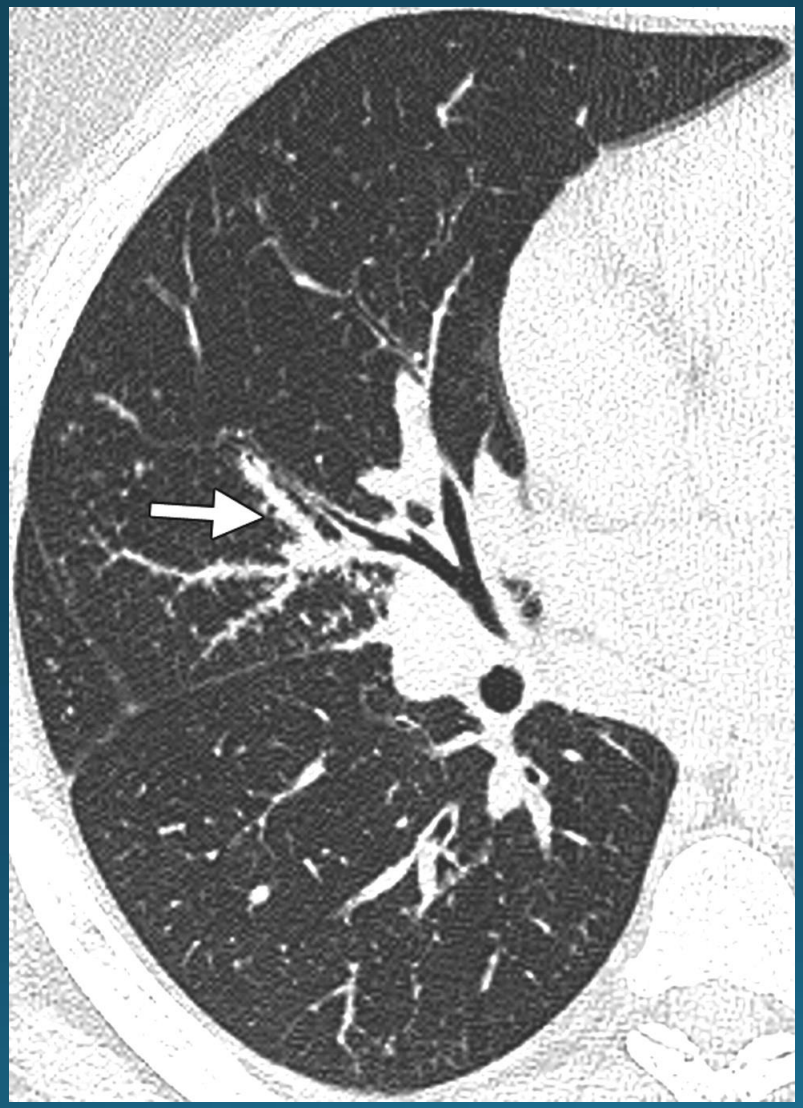

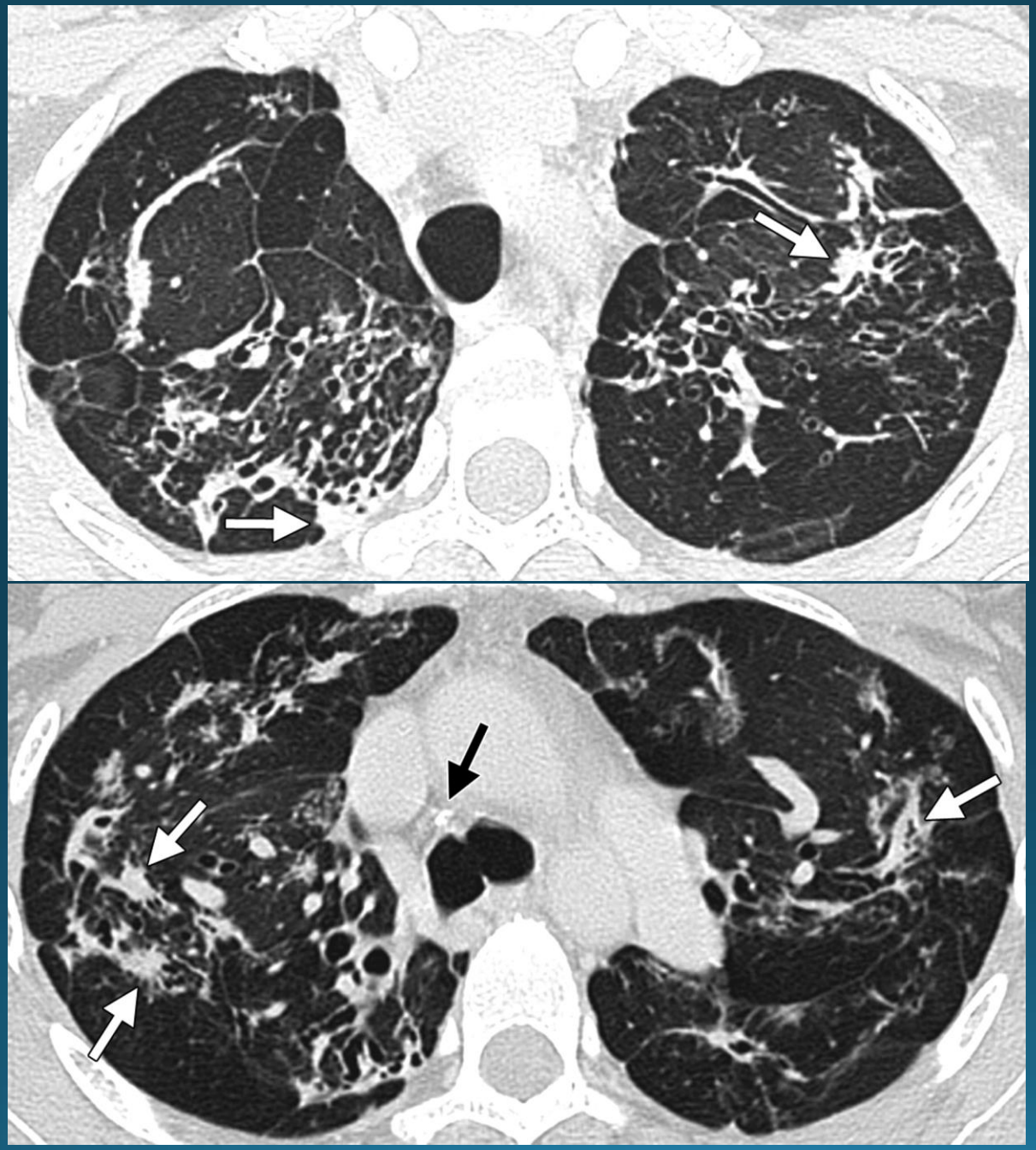

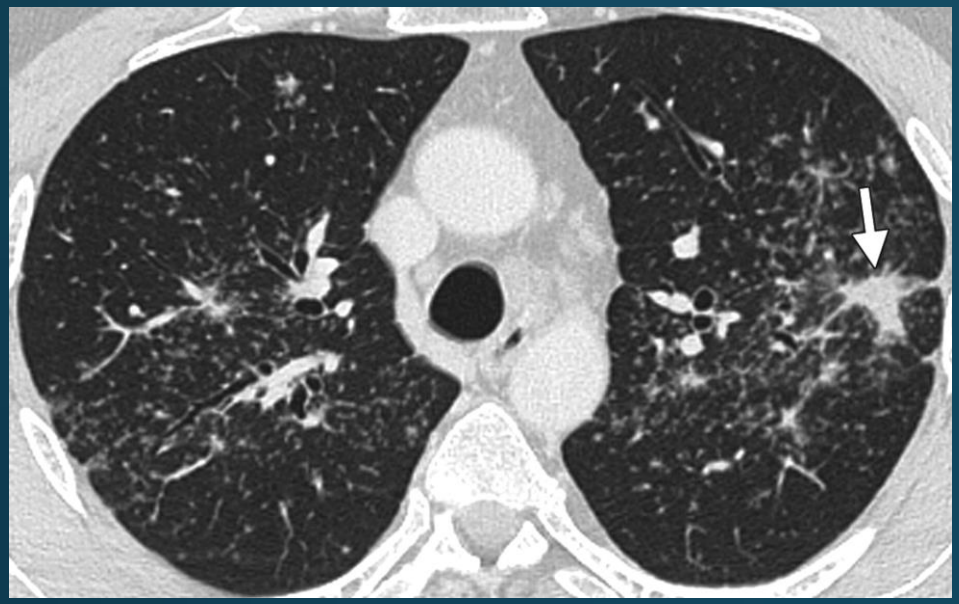

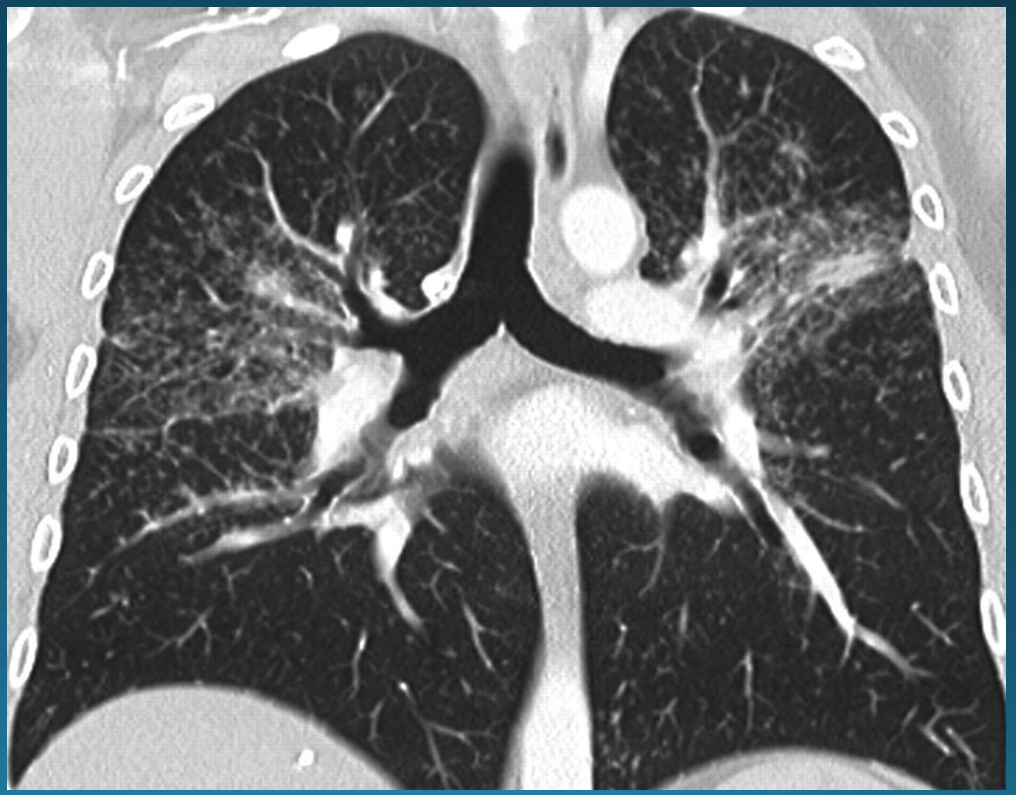

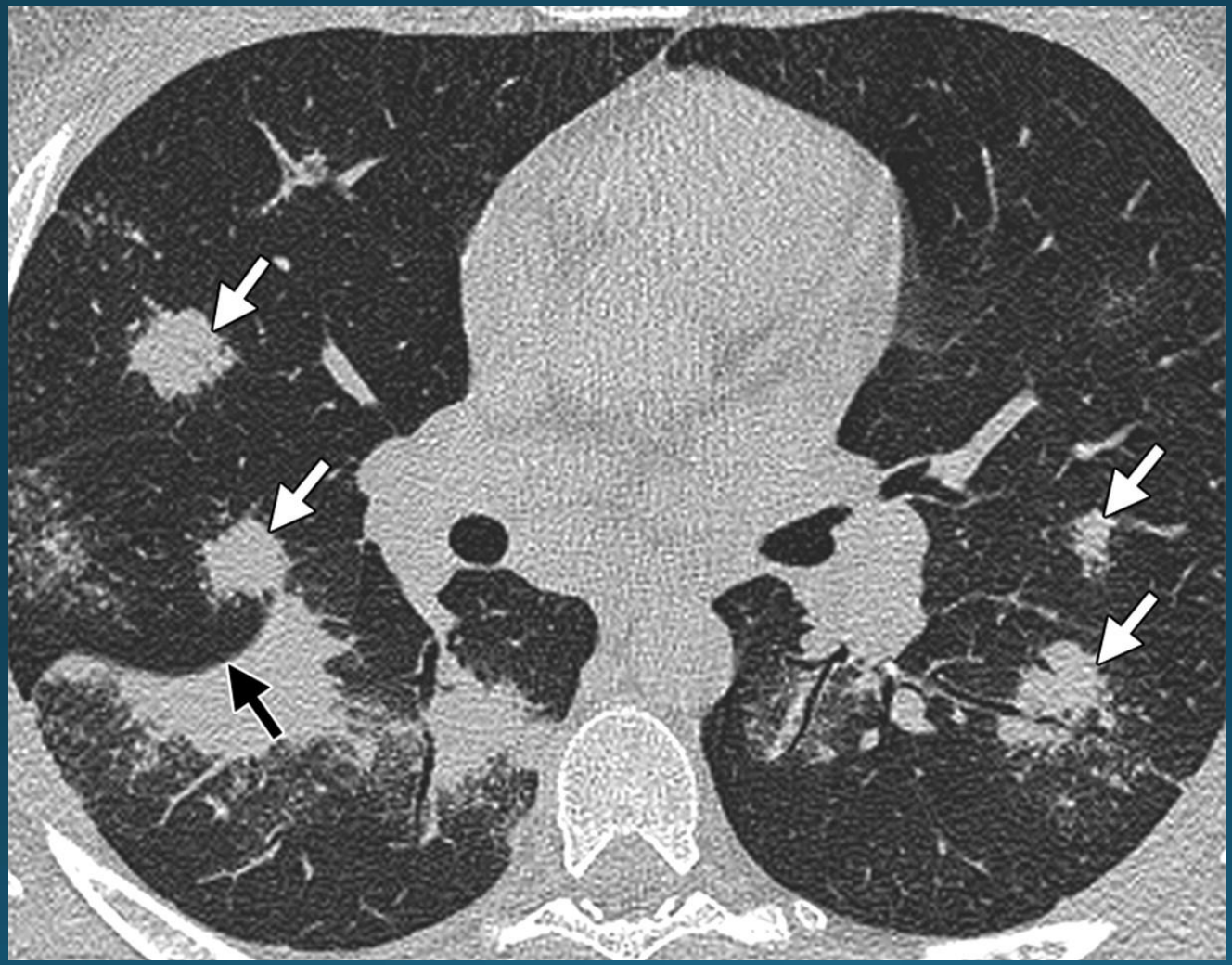

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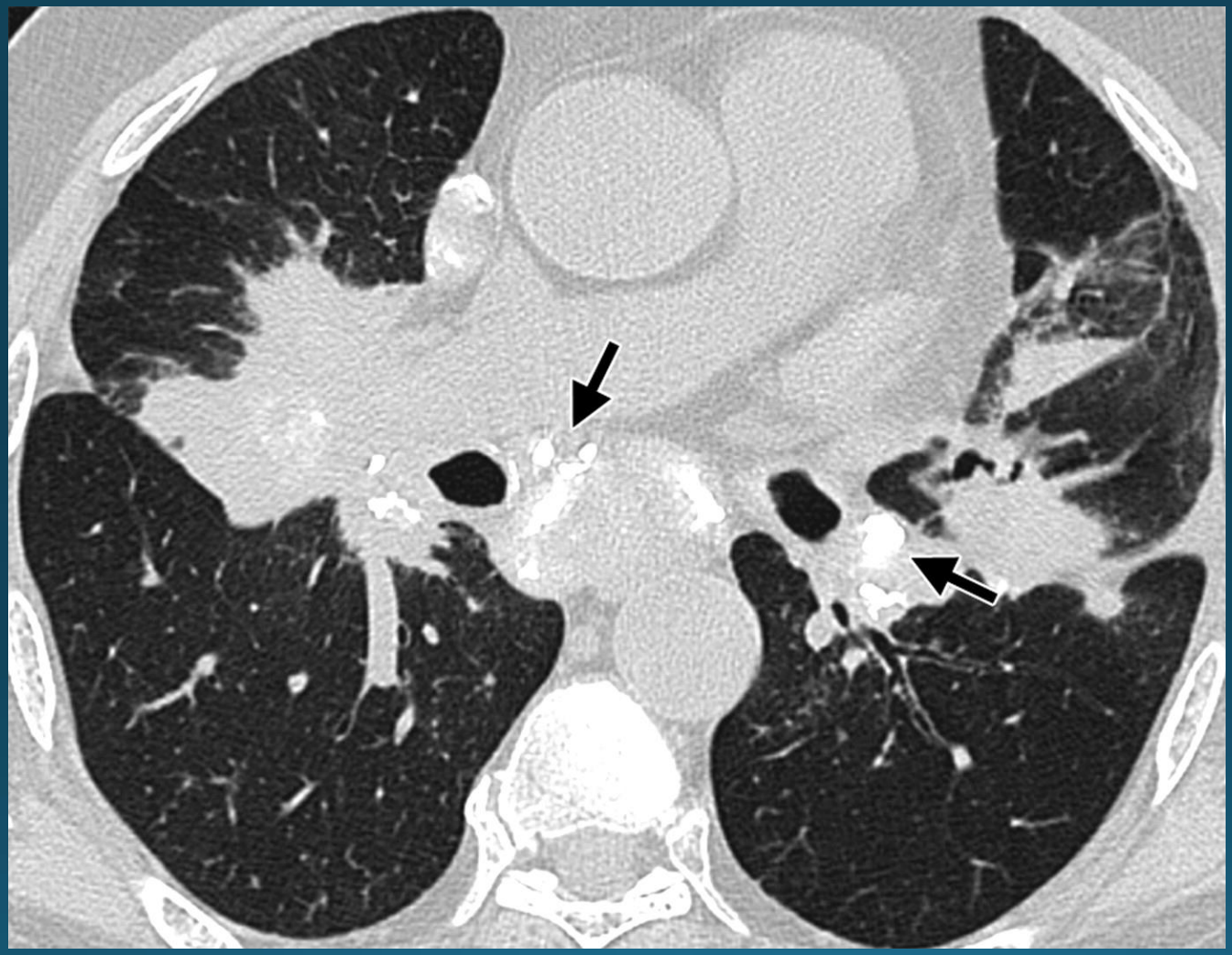

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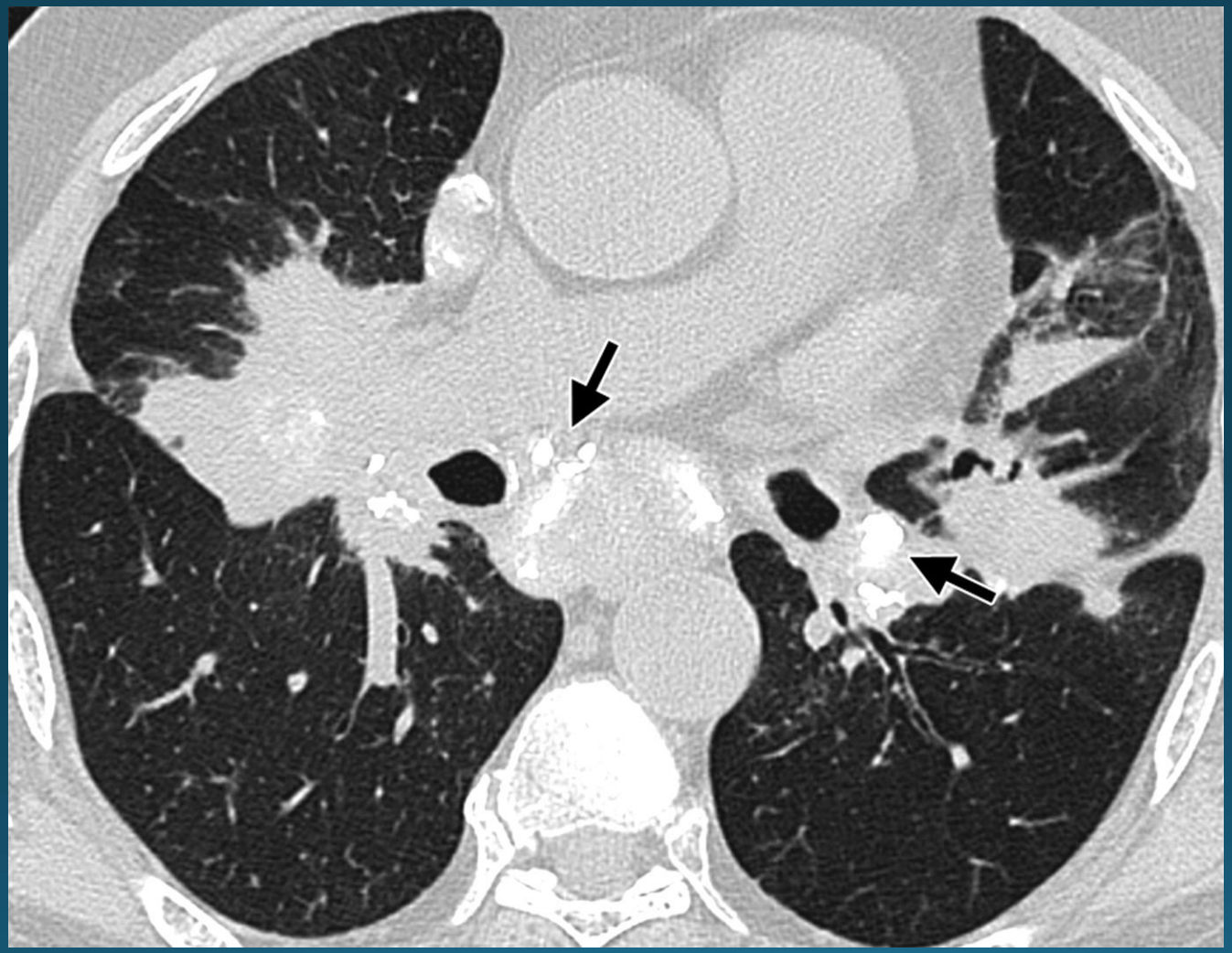
Axial high-resolution CT scan (pulmonary parenchymal window) shows bilateral enlargement and peripheral calcification of mediastinal and hilar lymph nodes (arrows).



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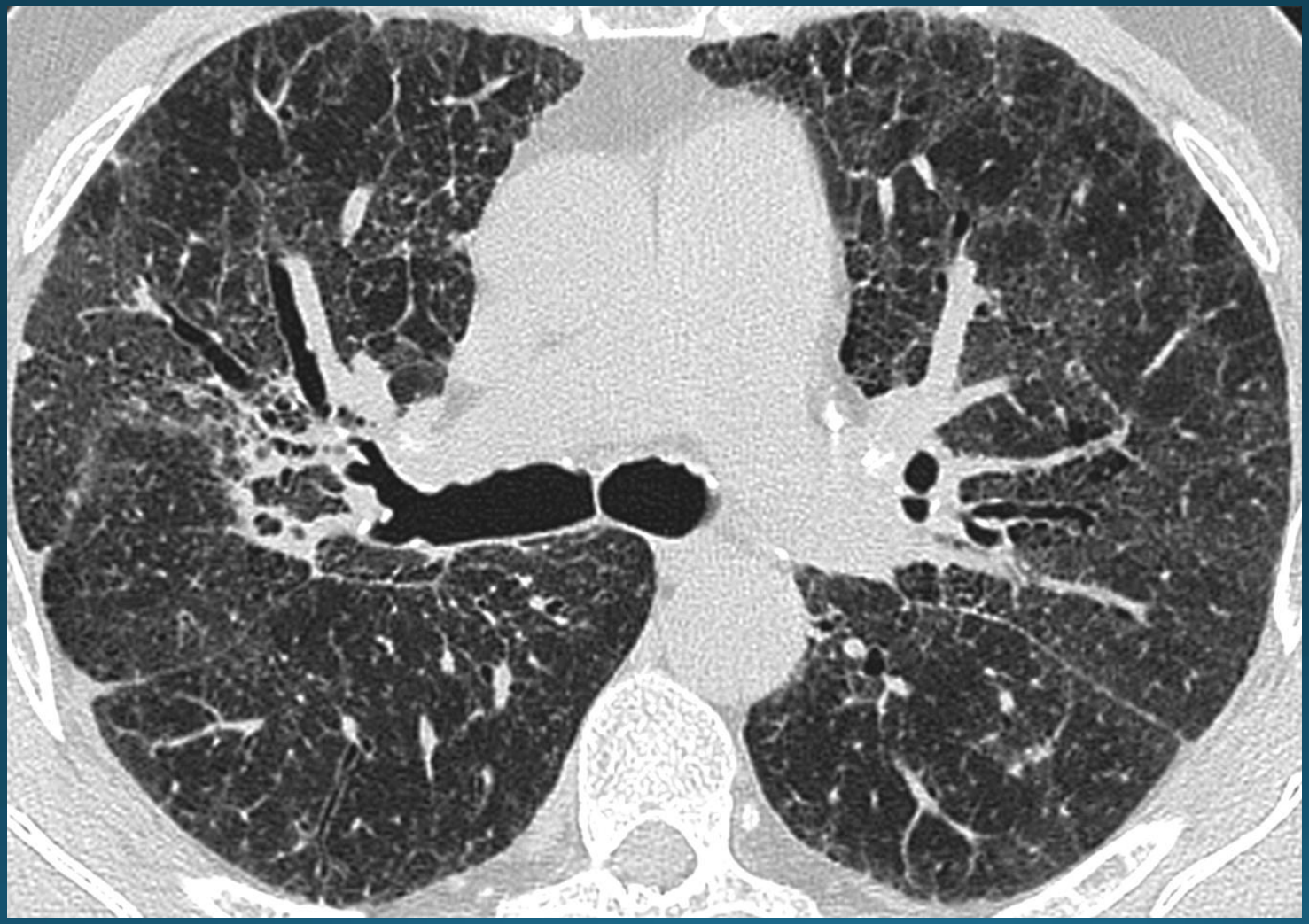
RadioGraphics

Ground-glass opacities in pulmonary sarcoidosis.



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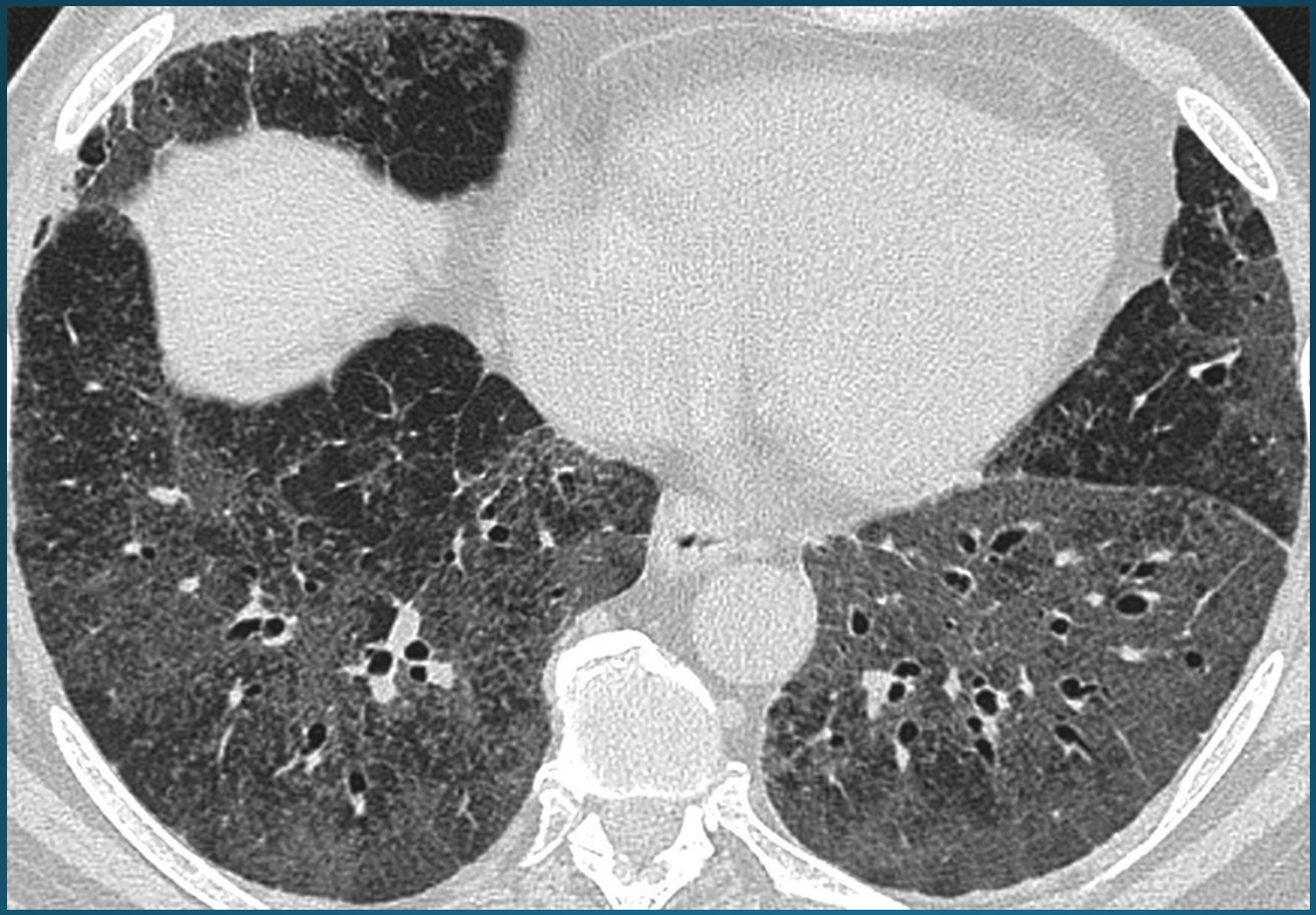

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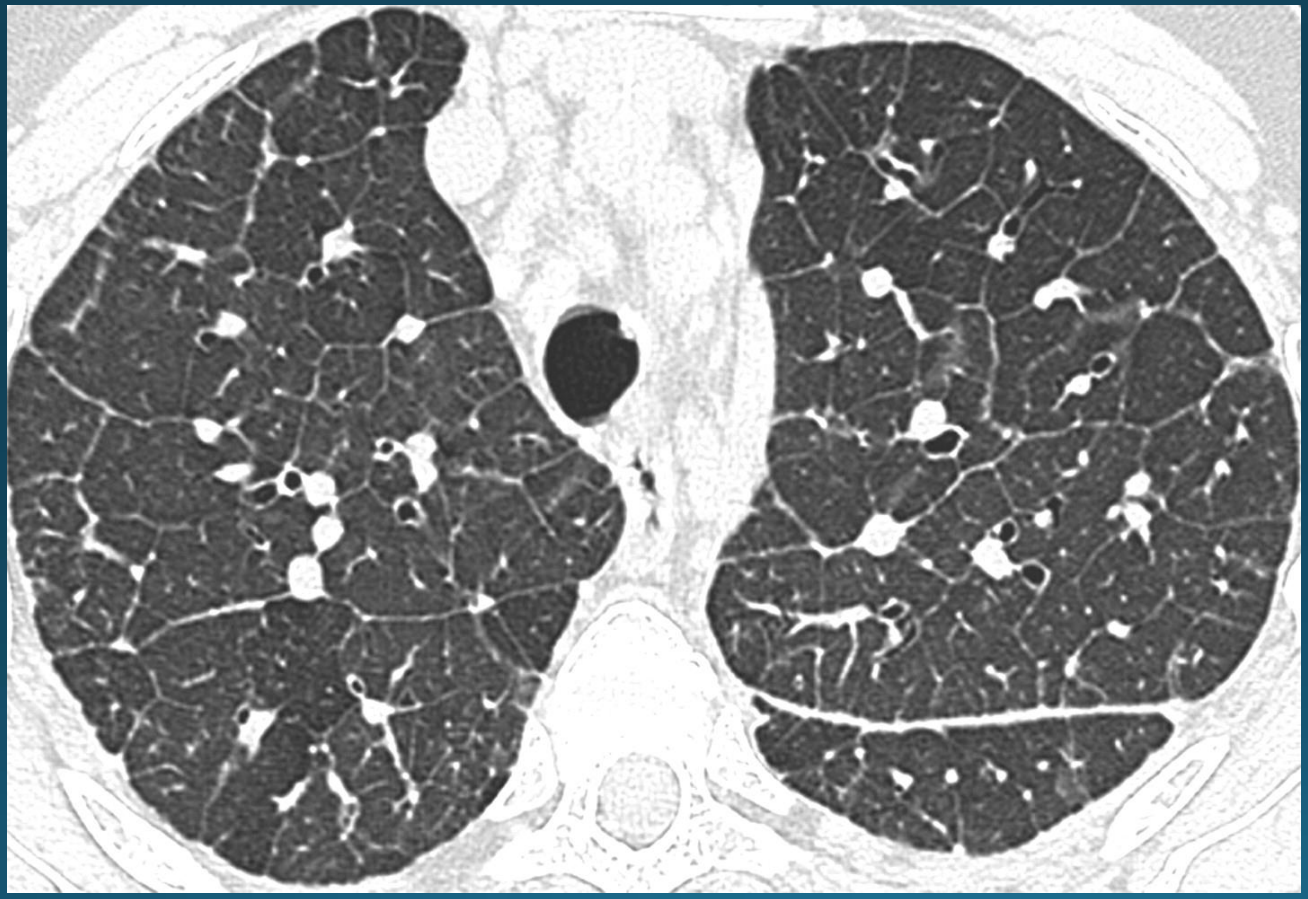
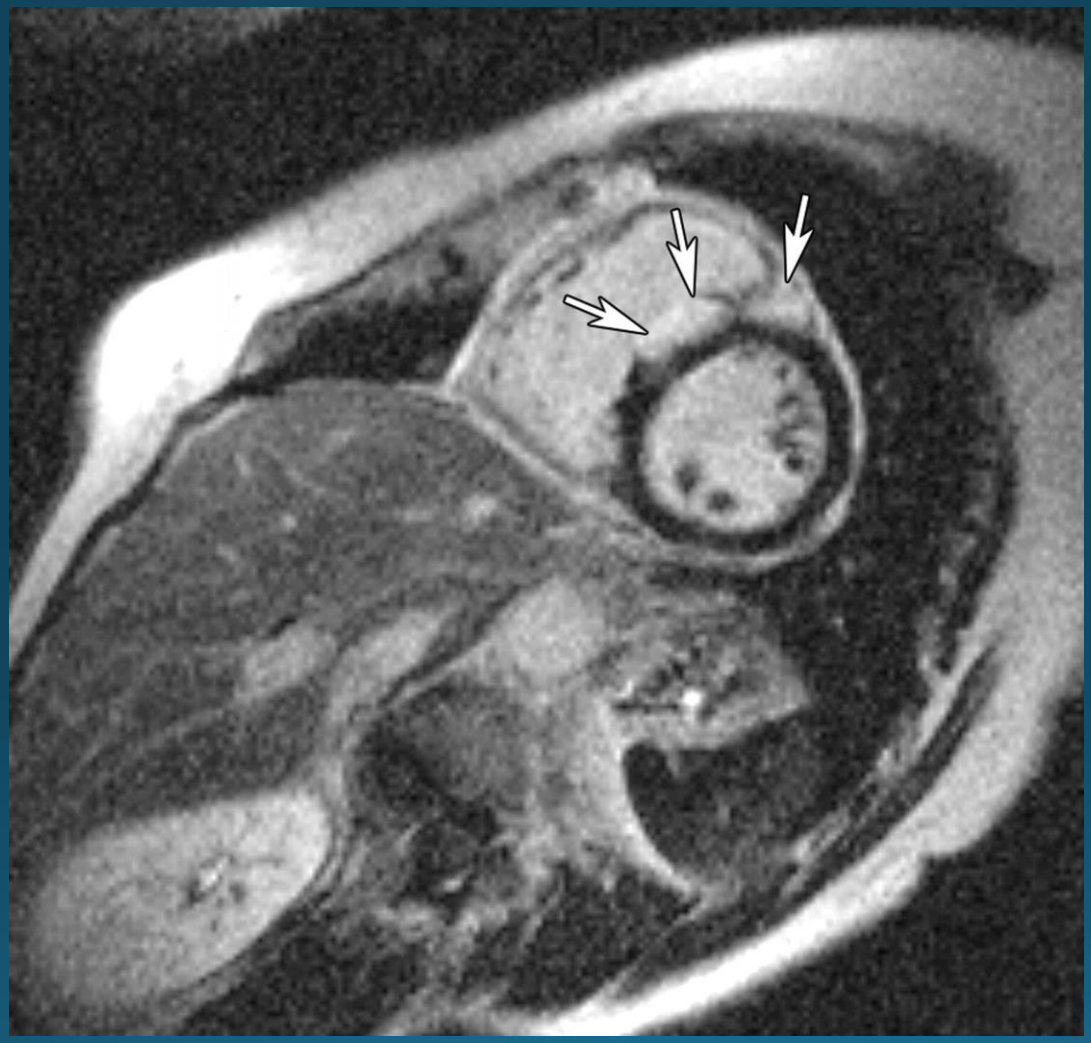
Axial high-resolution CT scan shows mediastinal lymph node enlargement and a reticular pattern produced by nodularity and thickening of interlobular septa, pleural surfaces, and fissures, features that are seen in lymphangitic carcinomatosis as well as.



Heart

- In 25% of patients, often conductive system
- Myocardial involvement → 5%
- MR :
  - Increased signal intensity on T2
  - Enhancement on contrast-enhanced T1
  - Limited MR use due to pacemaker
- Gallium-67 scintigraphy
  - low sensitivity and specificity
  - helpful in monitoring disease activity

Cardiac sarcoidosis in a 59-year-old woman with abnormal electrocardiographic findings.



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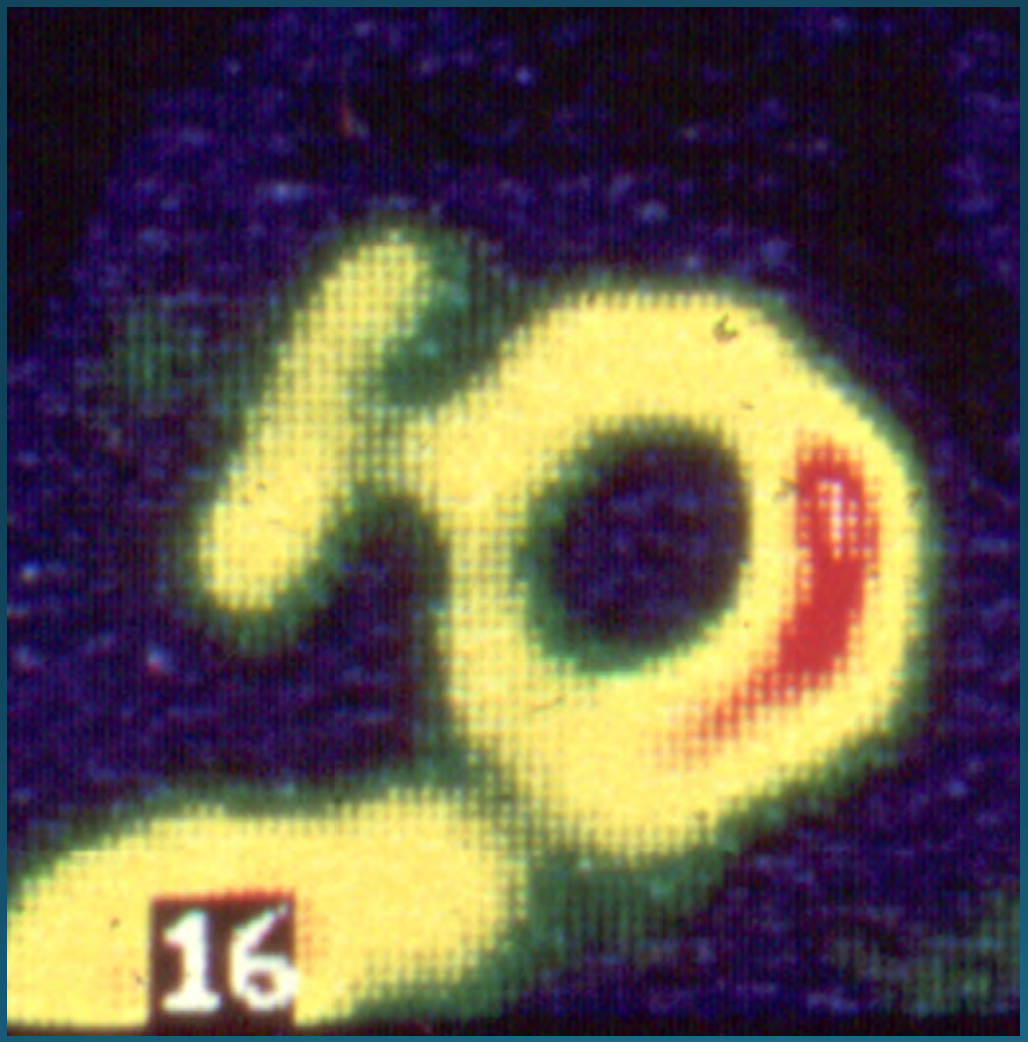
Cardiac sarcoidosis in a 60-year-old man who presented with complete atrioventricular blockage.



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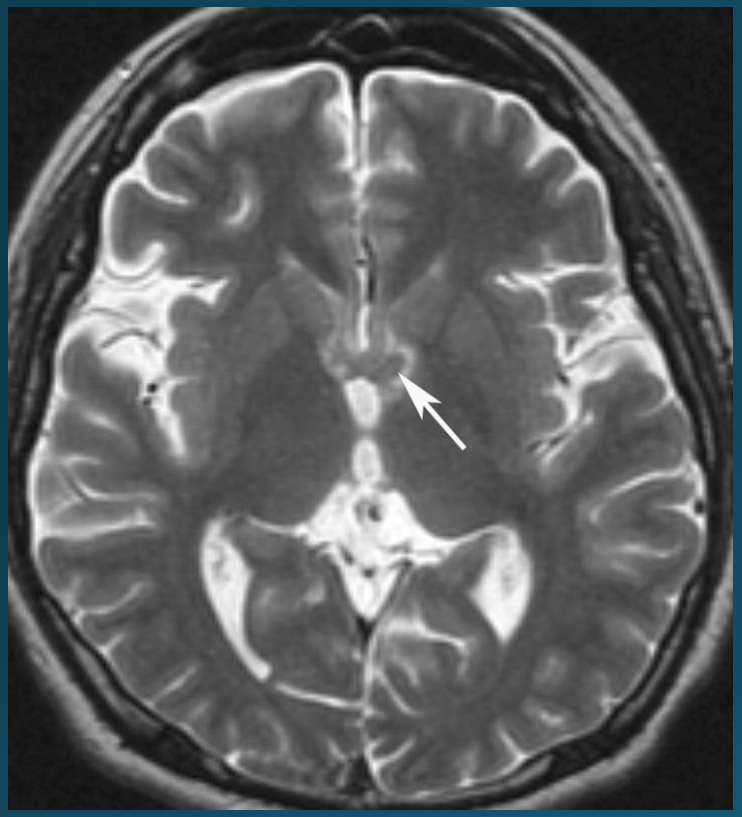
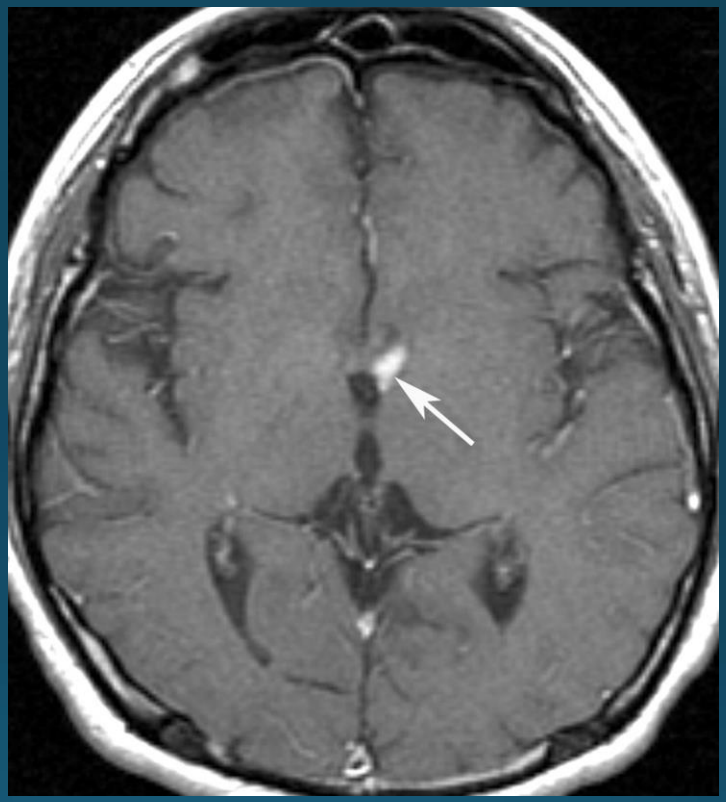
CNS

- Up to 25% of patients
- Clinically recognizable in <10%
- Manifestations and prognosis are variable
- Imaging findings mimic infectious or metastatic disease
- CSF analysis might help:
  - increased ACE titer
  - Increased CD4:CD8 ratio

Neurosarcoidosis in a 24-year-old man who presented with diabetes insipidus.

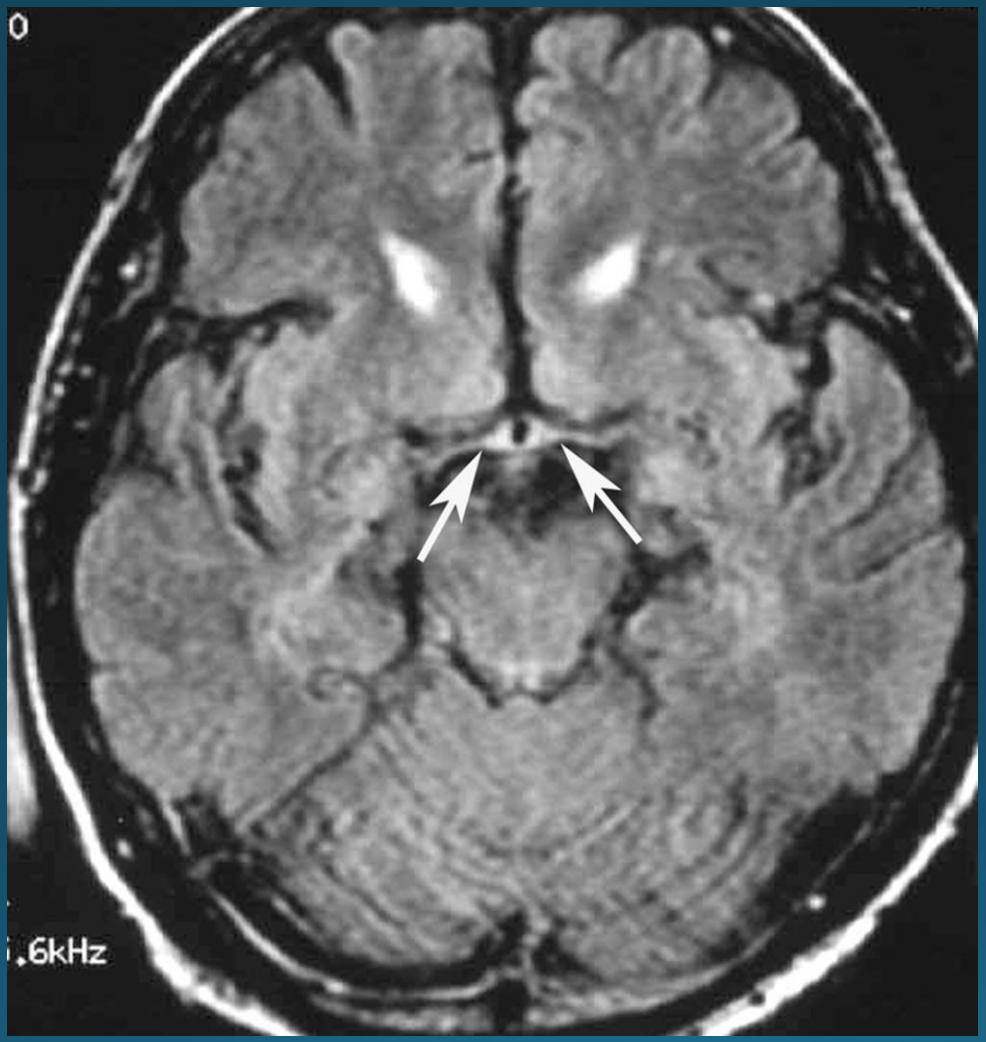




Axial T2-weighted MR image demonstrates an isointense periventricular lesion (arrow) surrounded by minimal high-signal-intensity edema.

On a contrast-enhanced T1-weighted MR image, the lesion demonstrates enhancement (arrow).

Neurosarcoidosis.



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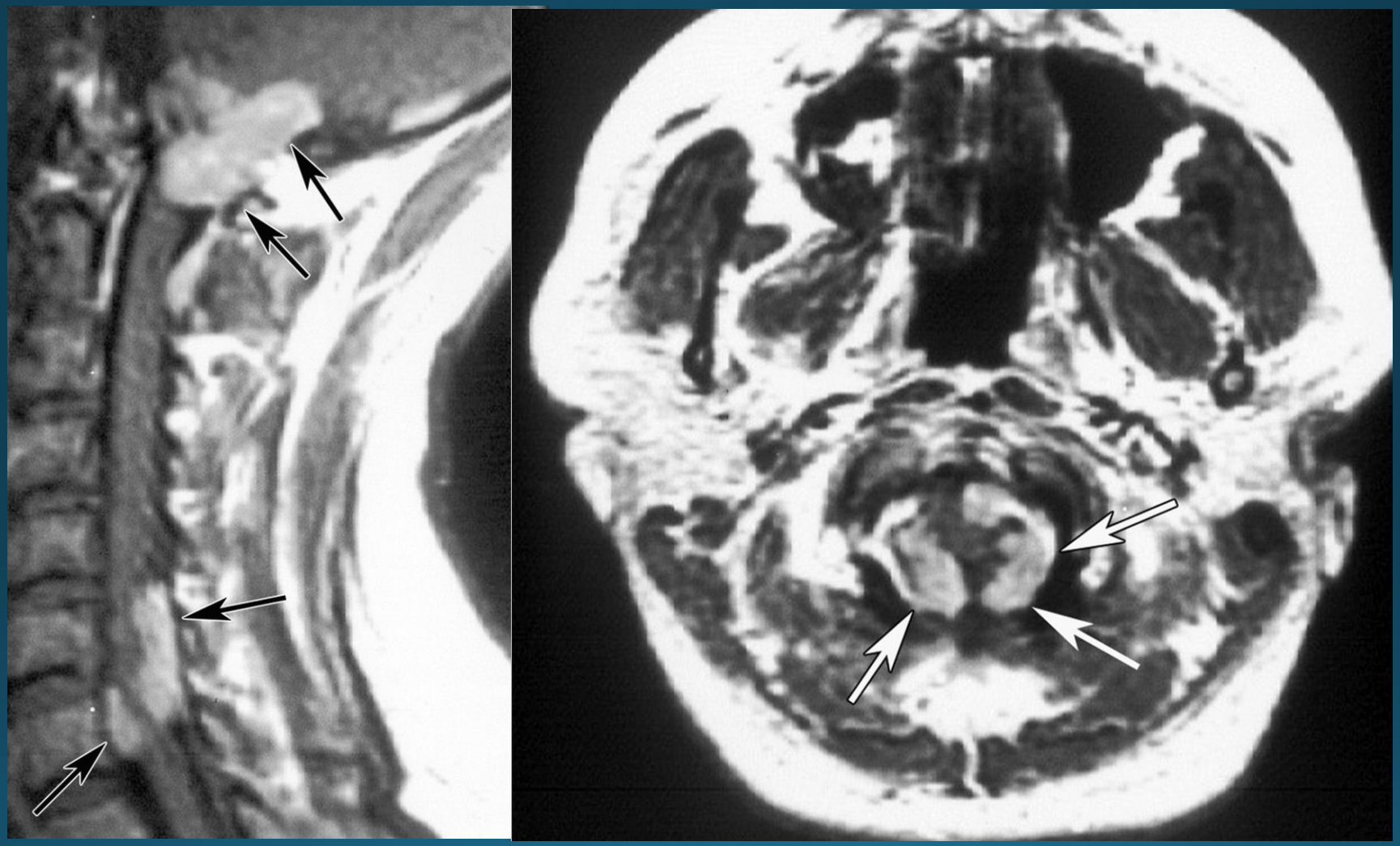
Leptomeningeal

- Commonly affecting the base of the brain → aseptic meningitis
- Contrast-enhanced T1-weighted imaging diagnostic.
- Less common → small enhancing nodules on the brain surface and in the perivascular spaces
- Brain and spinal cord involvement occur in the early stage of the disease and responds rapidly to steroid treatment.

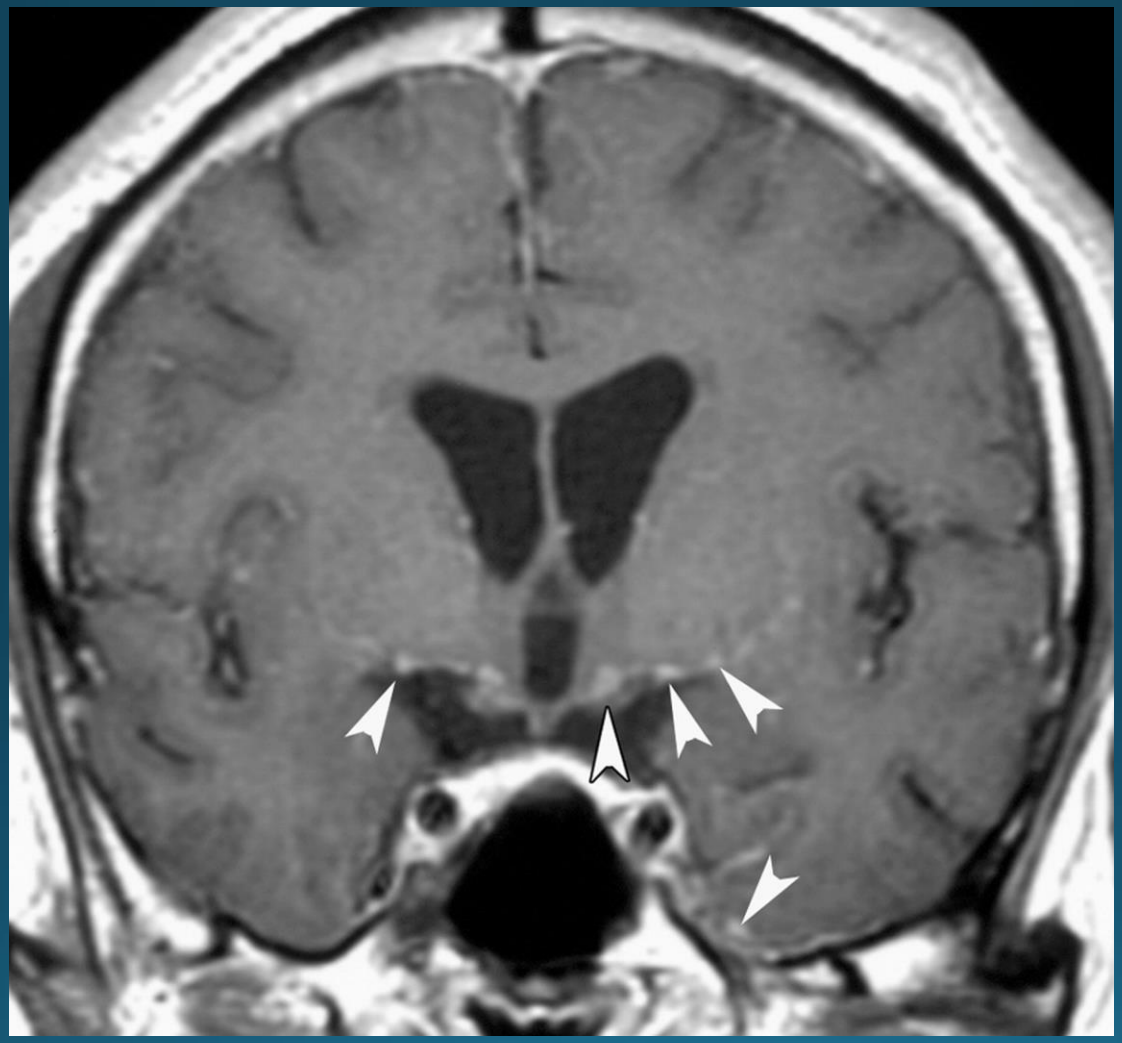
Spinal cord

- Cervical and thoracic spinal regions
- T2-weighted → intramedullary lesion with decreased signal intensity.
- Enlarged spinal cord with high signal intensity due to associated edema.
- Sarcoid granuloma → enhancement on T1

Leptomeningeal involvement in a 23-year-old woman.



Leptomeningeal involvement in a 34-year-old man who presented with bilateral facial palsy.



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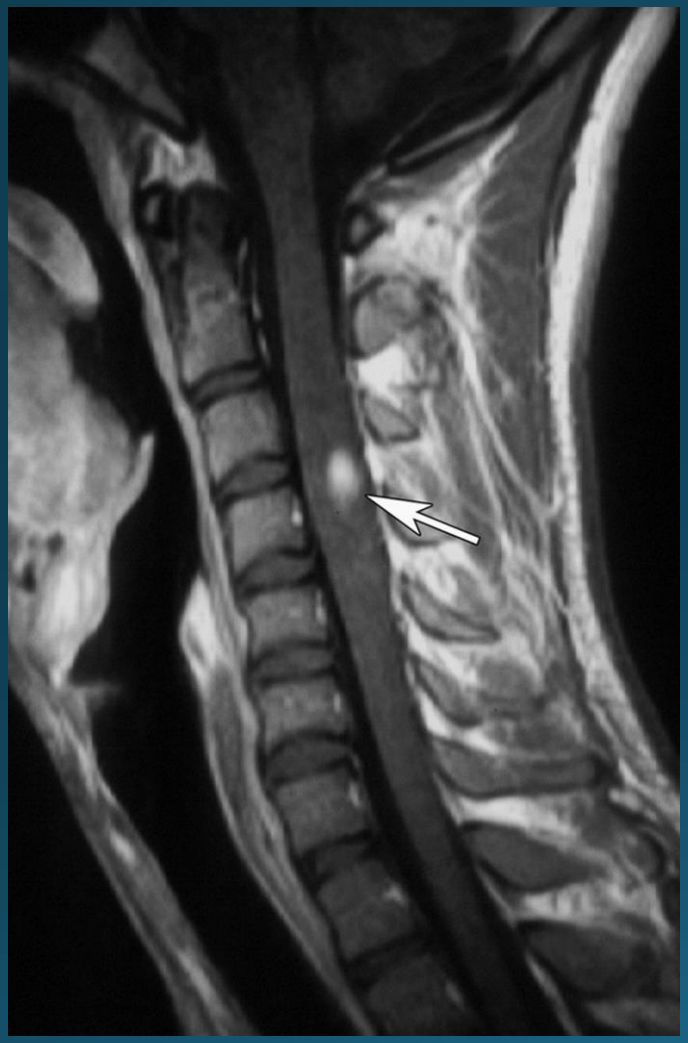
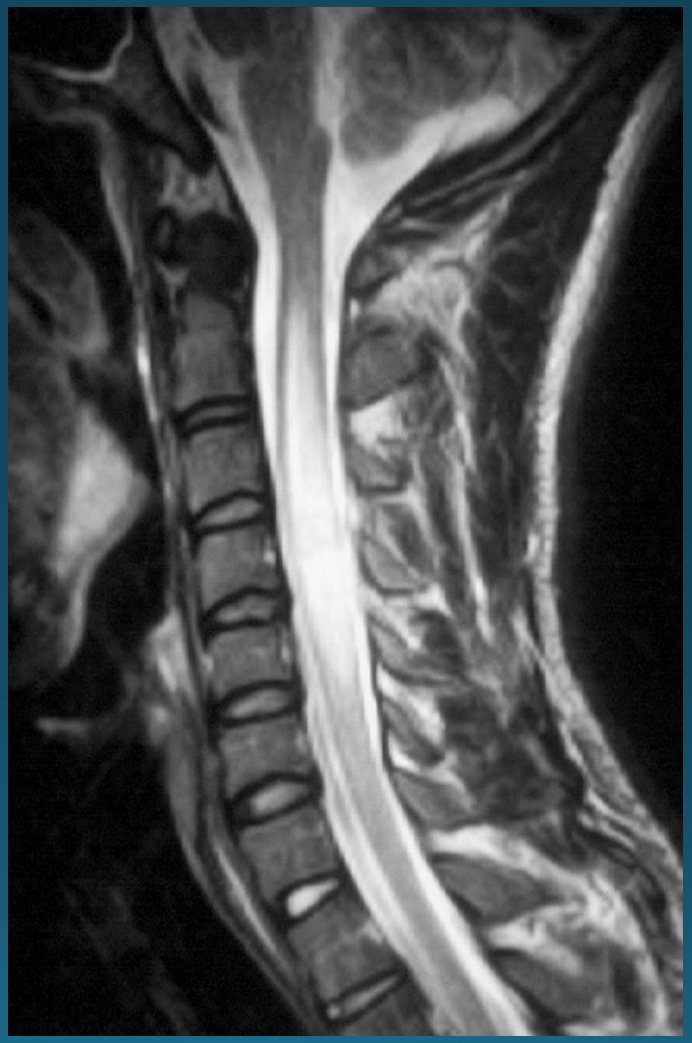
Spinal cord involvement in a 23-year-old man who presented with mild paresis of the hands.

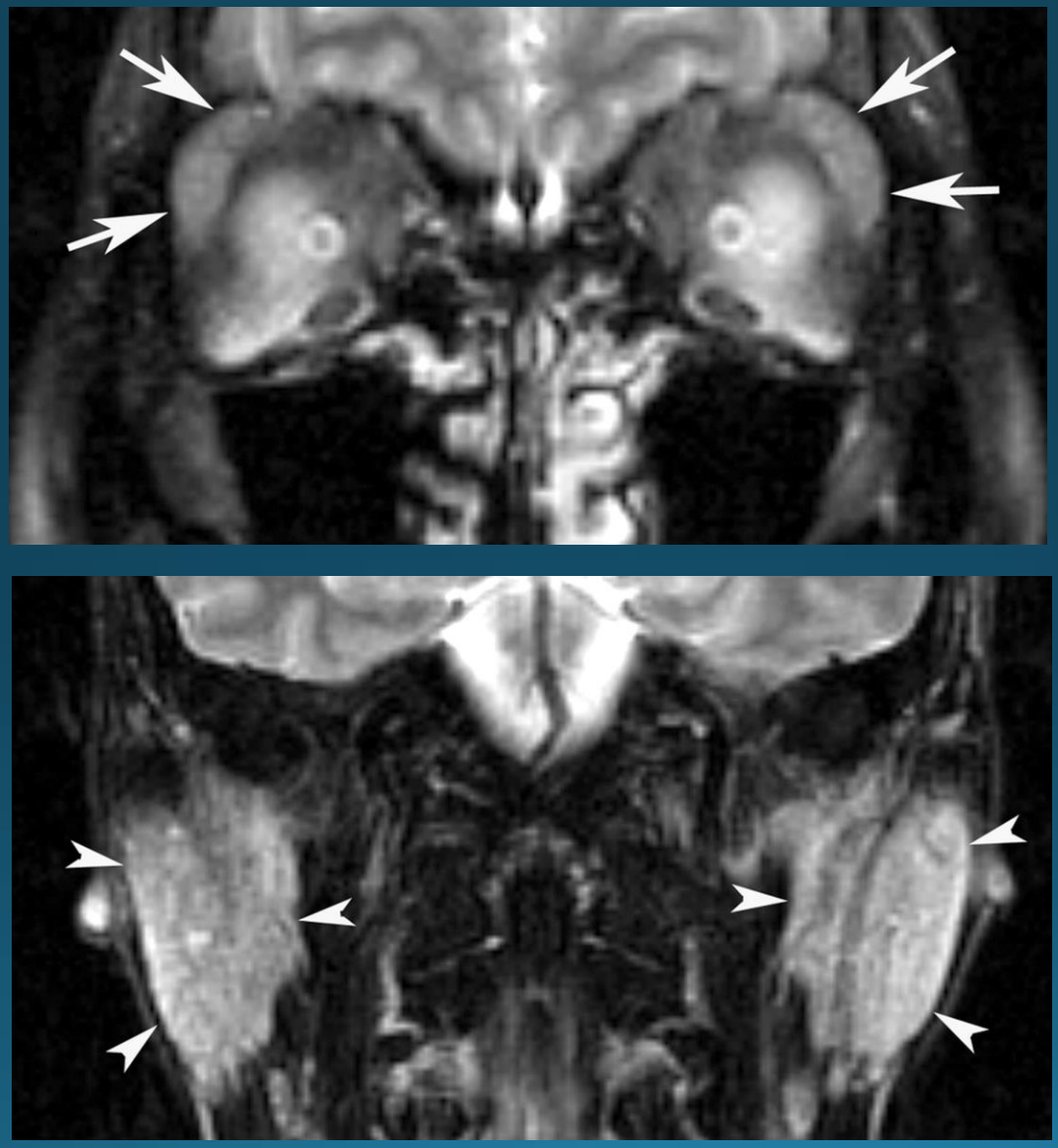




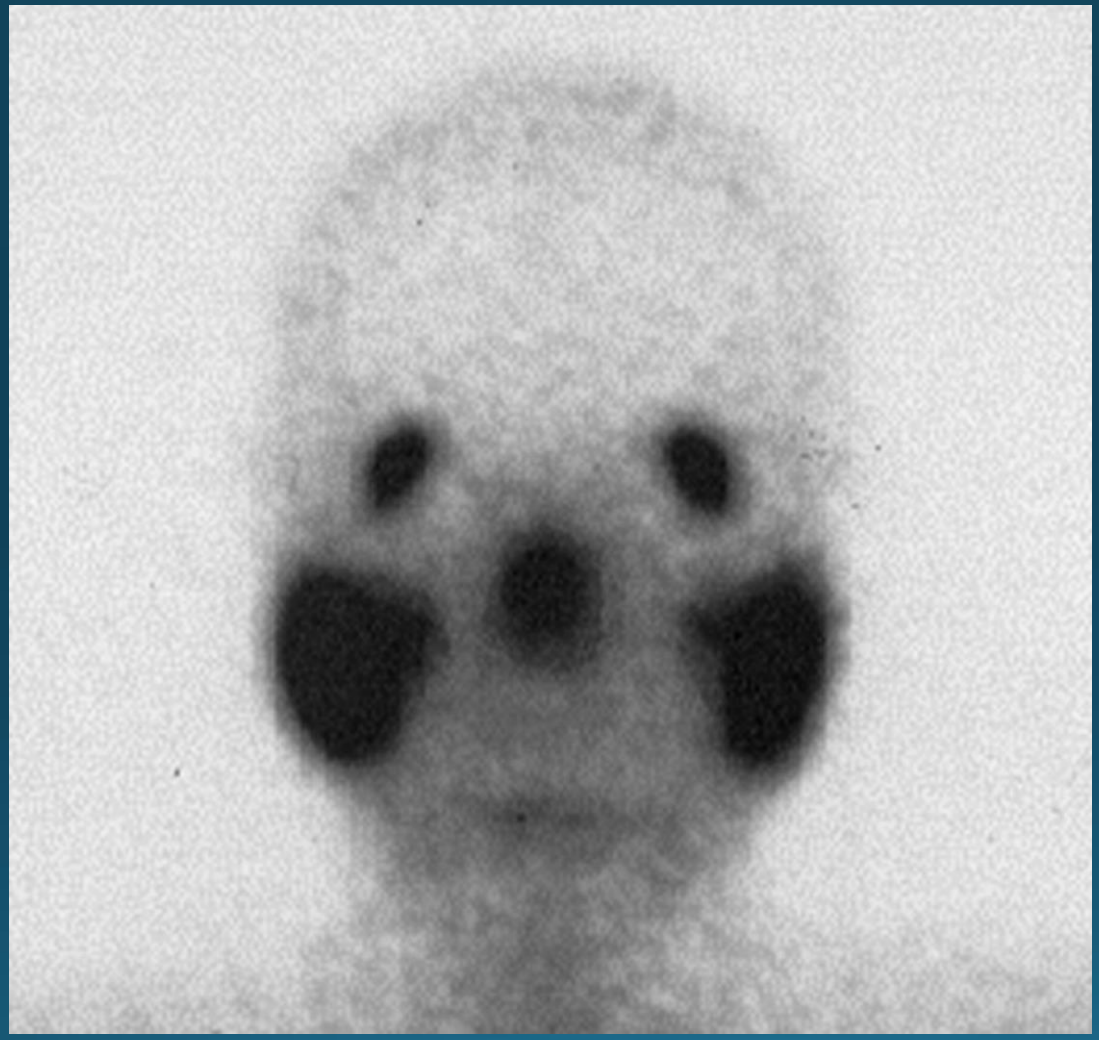
Eyes

- In up to 80% of patients
- Uveitis → the most common condition and is typically bilateral.
- Acute uveitis usually resolves spontaneously or responds to local corticosteroid therapy (eyedrops).
- Lacrimal glands involvement less common

Ocular involvement in the same patient as in Figure 1.



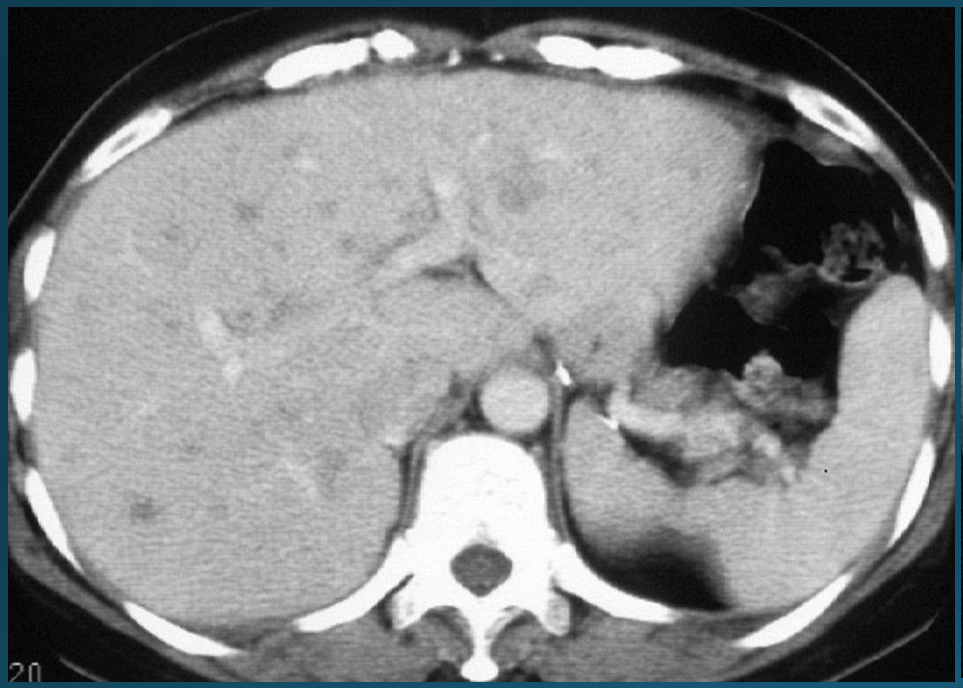
Ocular involvement in the same patient



Liver and spleen

- 50%–80% of autopsy specimens
- Dysfunction of these organs is uncommon.
- Isolated hepatosplenic sarcoidosis do not require treatment.
- Splenic nodules are larger and more common than hepatic
- Multiple nodules in hepatic sarcoidosis are easily mistaken for more common diseases, including metastases and lymphoma

Hepatosplenic involvement in a 49-year-old woman with pulmonary sarcoidosis.



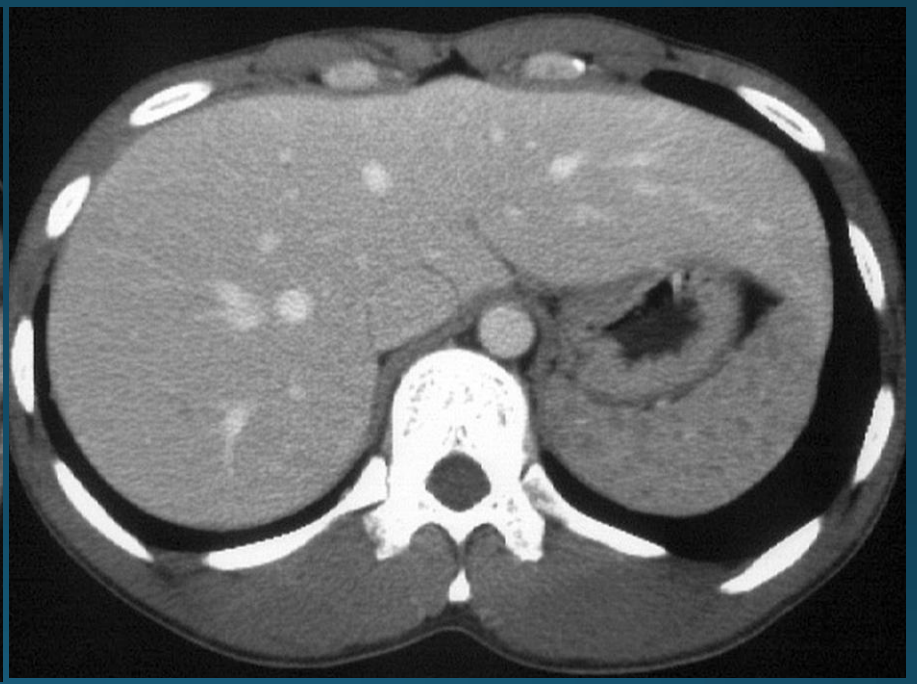
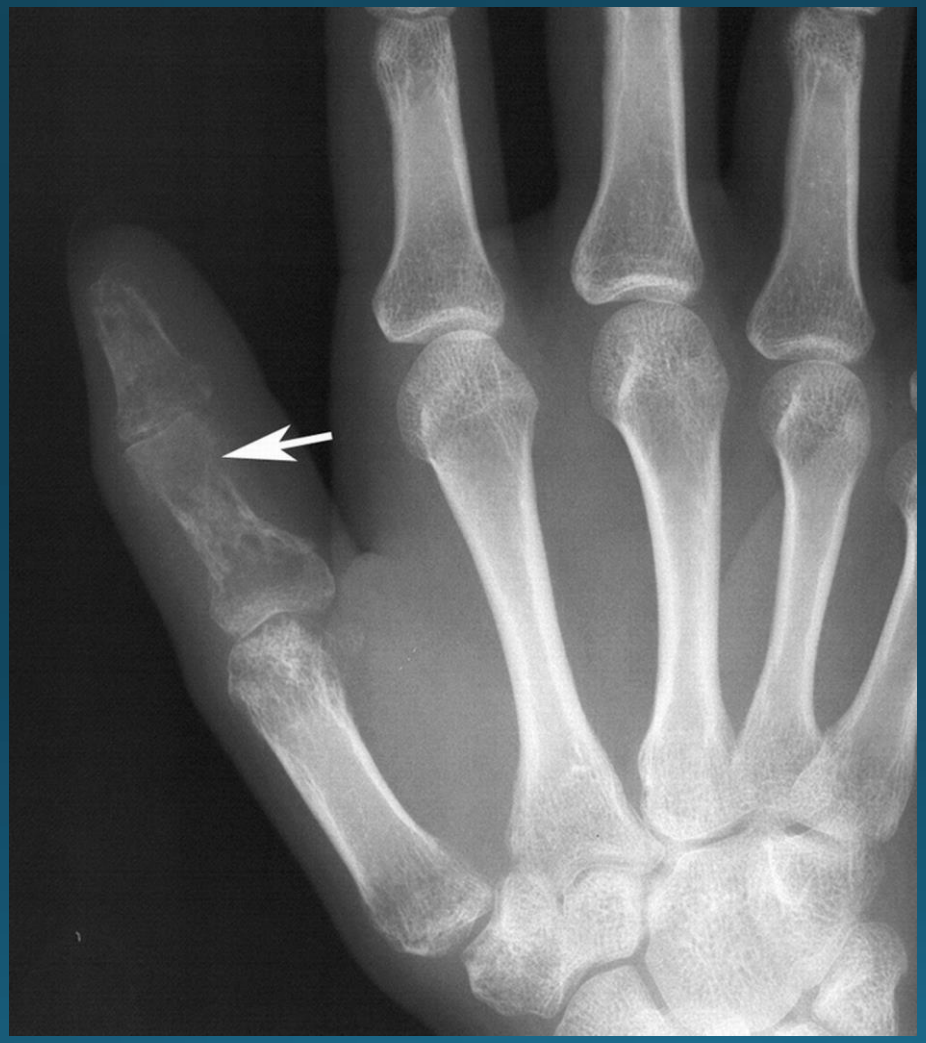


Figure 28a. Bone sarcoidosis in a 28-year-old man who presented with left thumb pain.



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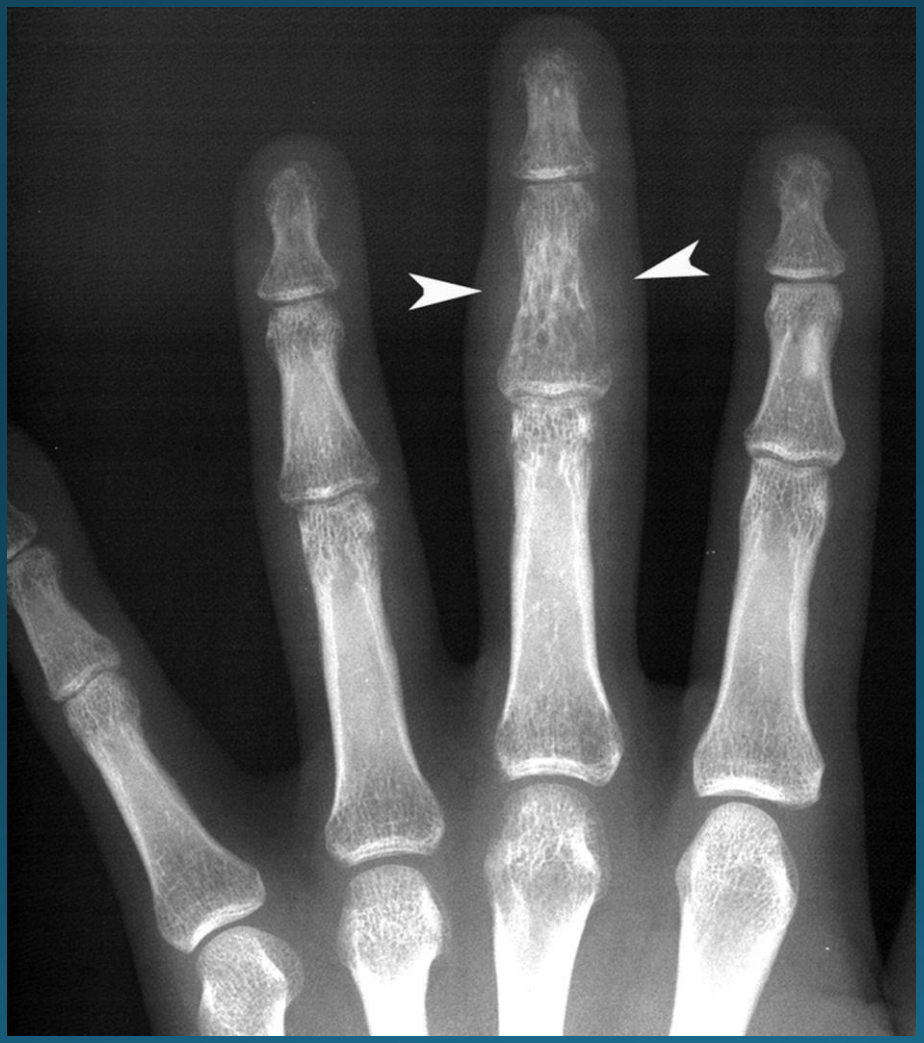
MSK

- Usually in patients with generalized disease
- Joint involvement is common
- Inflammatory arthralgia (up to 40% of cases) → knees, ankles, elbows, and wrists.
- Erythema nodosum, periarticular ankle inflammation, and mediastinal lymphadenopathy→ Löfgren syndrome→ self-limiting course with spontaneous resolution

- Skeletal involvement → phalanges in the hands and feet → 5%–10% of patients

- Radiologic features:
  - cystlike radiolucent areas
  - a lacelike honeycomb appearance
  - extensive bone erosion with pathologic fractures
  - subcutaneous soft-tissue mass or tenosynovitis may also be present. The combination of these radiologic features is virtually diagnostic.

Bone sarcoidosis in a 28-year-old man who presented with left thumb pain



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Bone sarcoidosis