CT of Thoracoabdominal Involvement in Erdheim-Chester Disease

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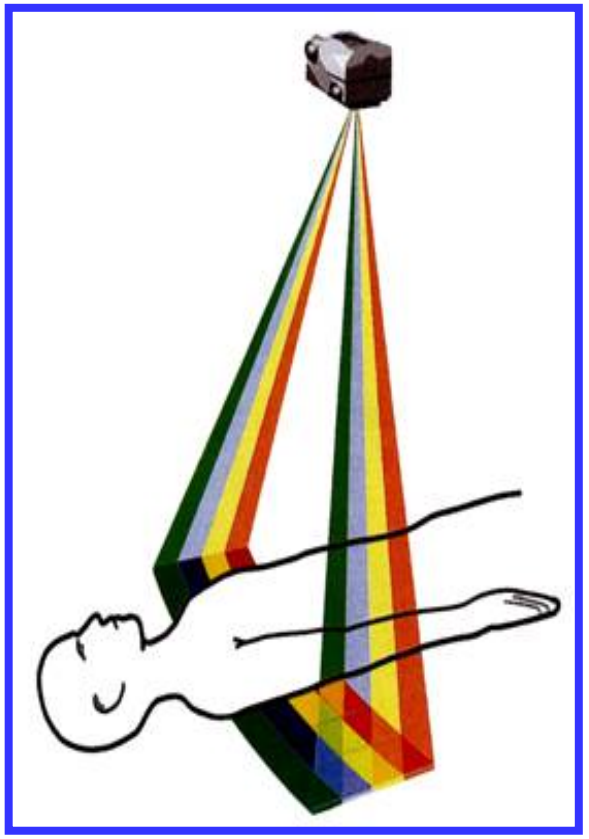
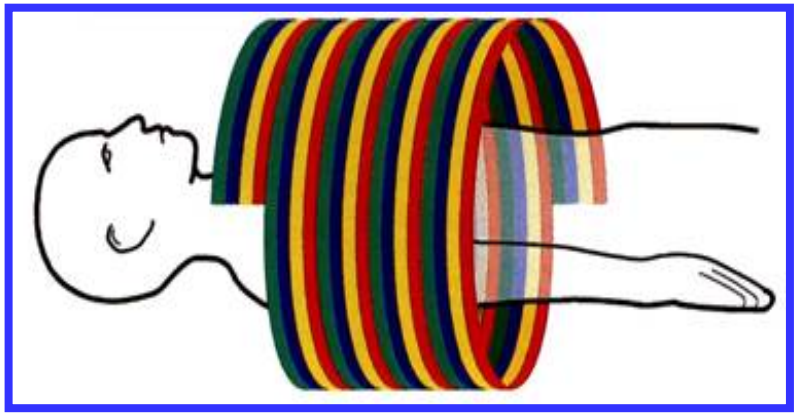
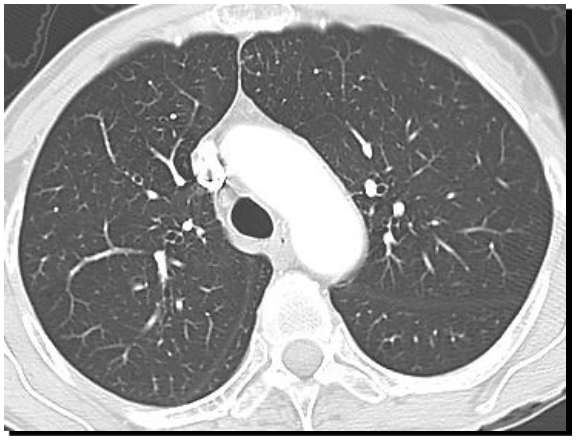
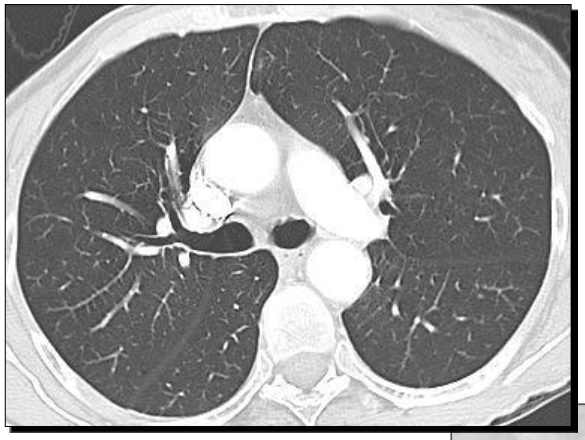




Rydberg et al. Radiographics 2000; 20:1787-1806

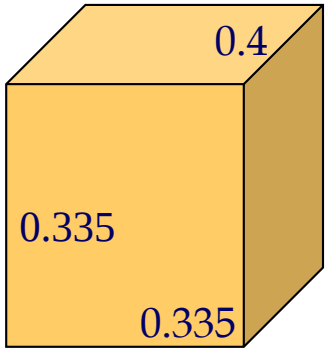
Volumetric Thin Collimation MDCT

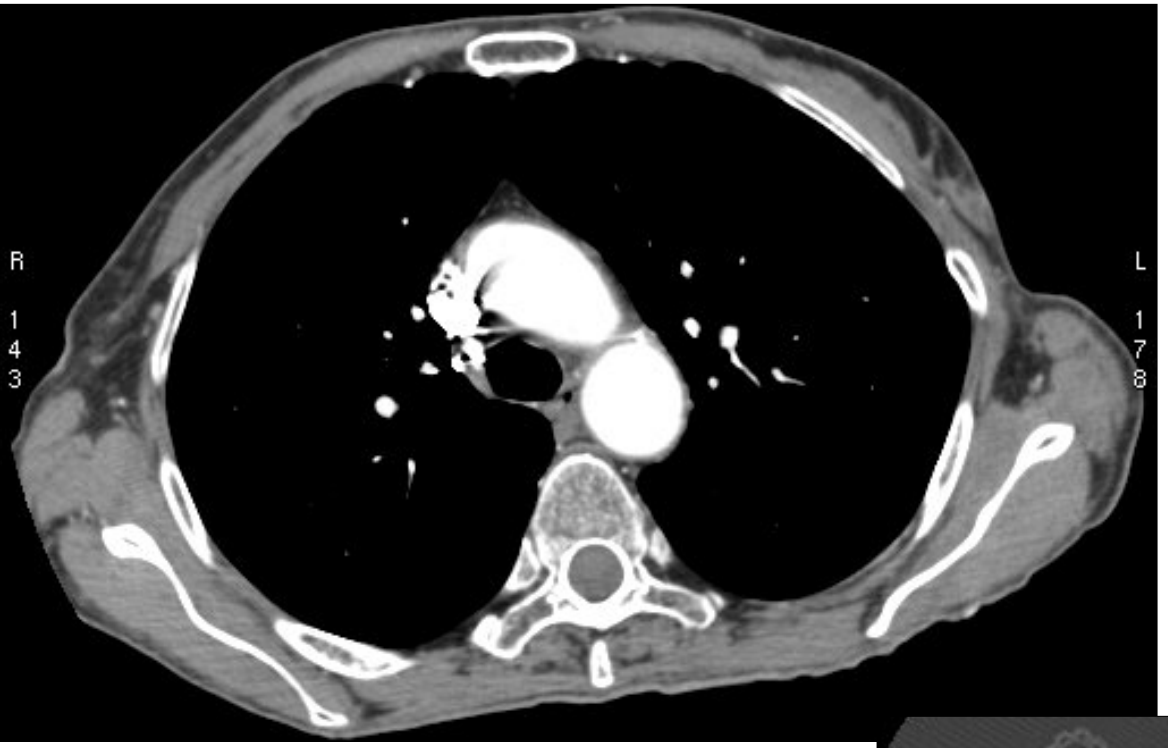




0.625 mm detector size
Axial thickness 0.8 mm
Reconst. increment 0.6 mm
High frequency algorithm
512 ² matrix
325 FOV

120 Kv
80-160 mAs



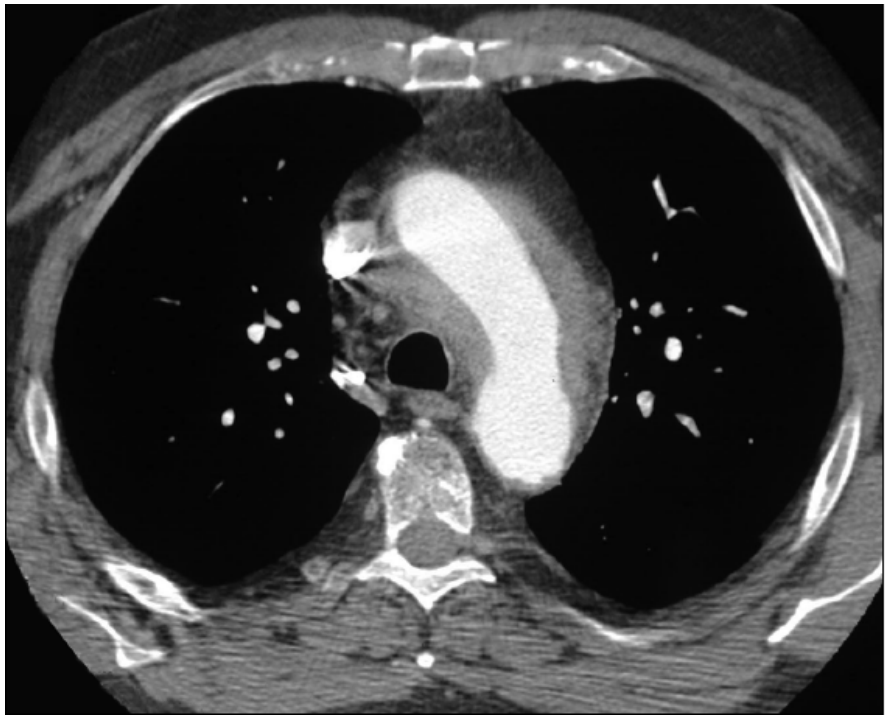


Smooth reconstruction algorithm
Mediaitinal window settings (40 HU / 400 HU)

High resolution reconstruction algorithm
Lung window settings (- 6OO HU / 1600 HU)

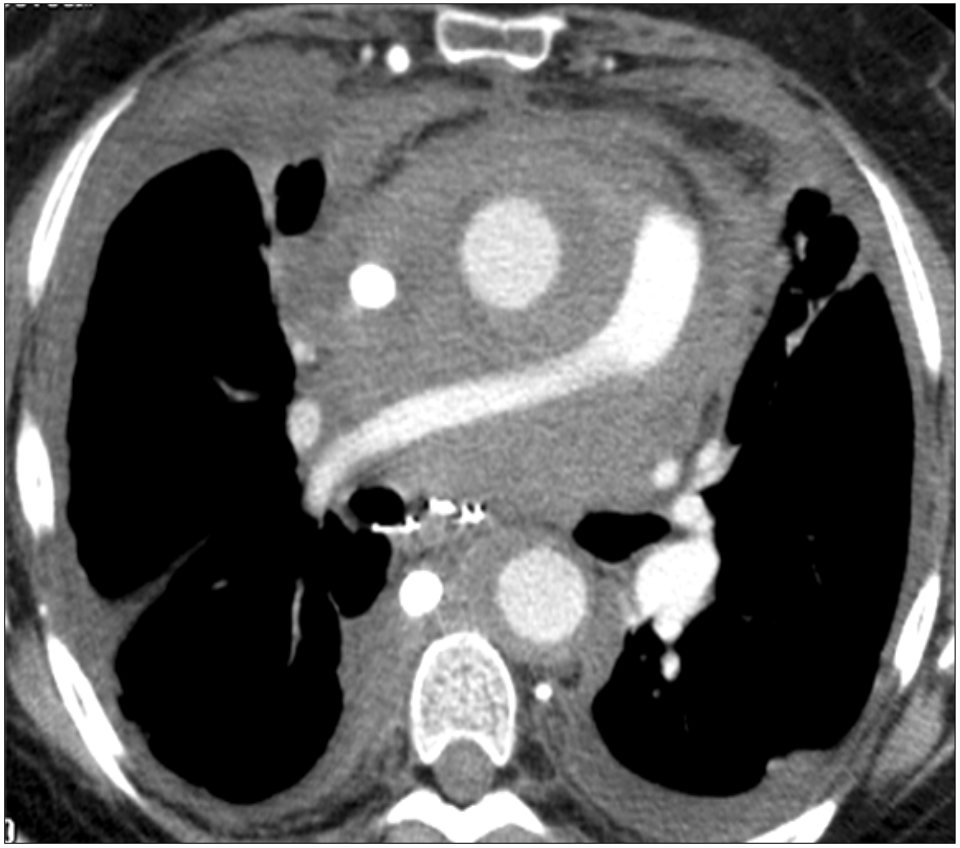
Periaortic infiltration (coated aorta) extending from aortic arch to abdominal aorta

Periaortic sheating extends upward to proximal portion of supraaortic trunks and inferiorly involves proximal portion of intercostal arteries

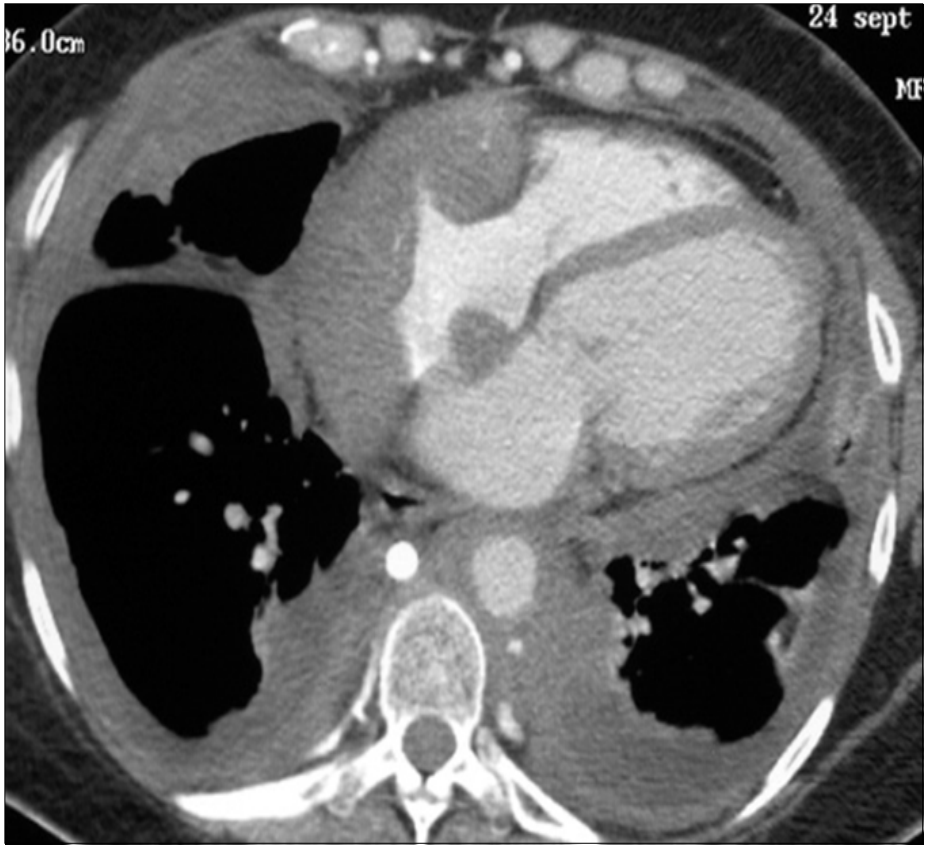


Mediastinal infiltration with a pseudotumoral appearance

Diffuse mediastinal infiltration with narrowing of the superior vena cava and right pulmonary artery

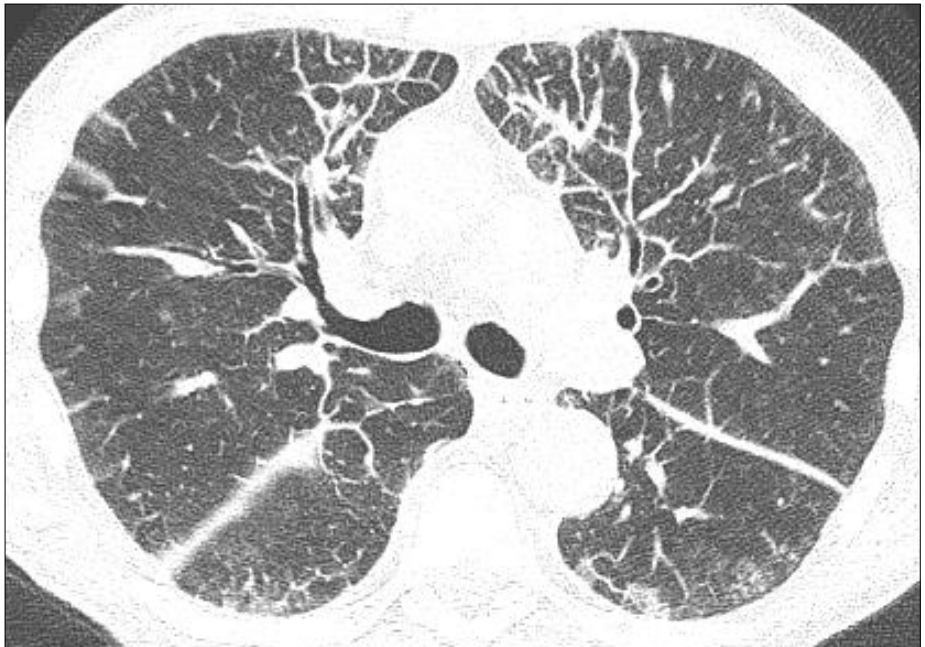


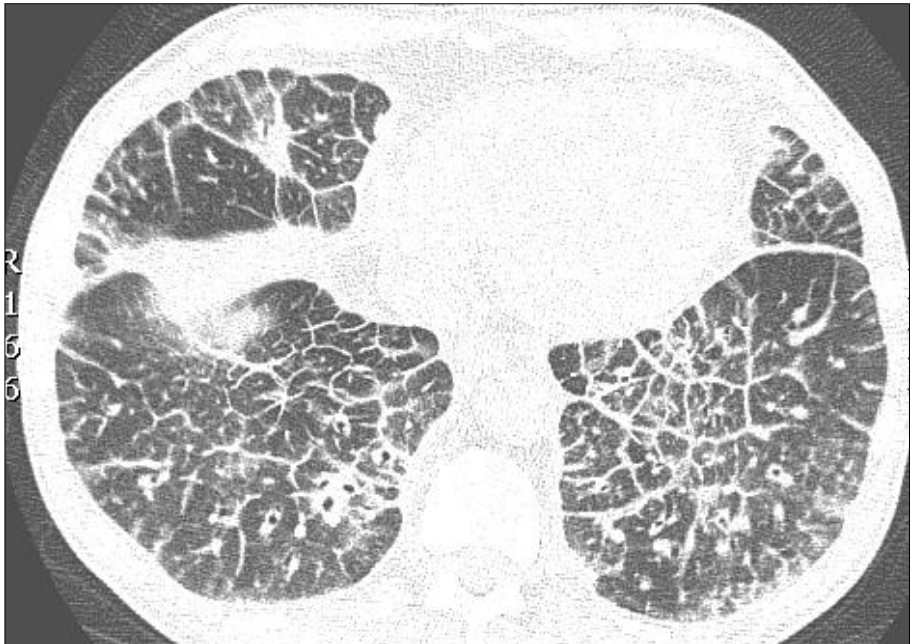
Bilateral pleural thickening and effusions
Infiltration of the right atrium wall and right coronary sulcus



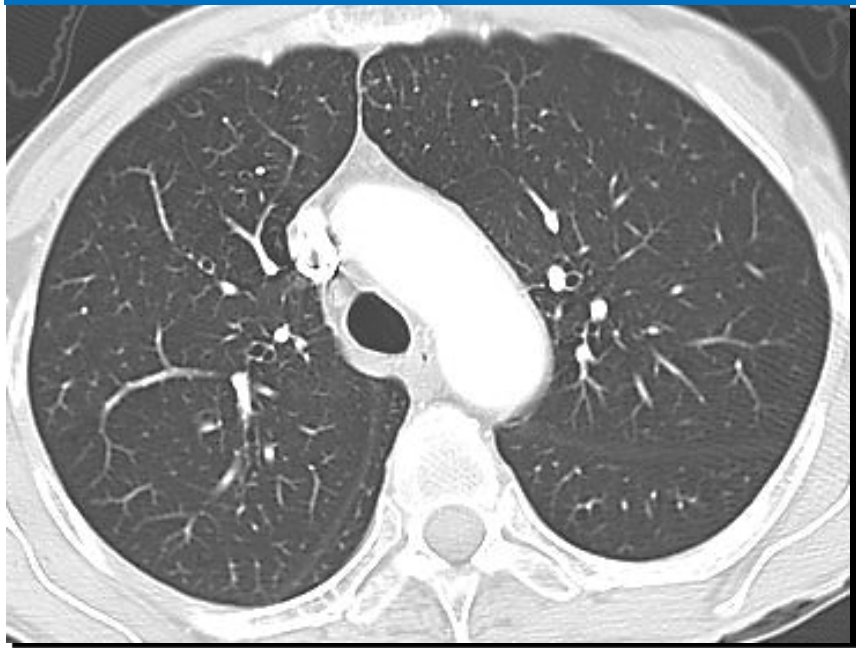
Bilateral pleural thickening and effusions

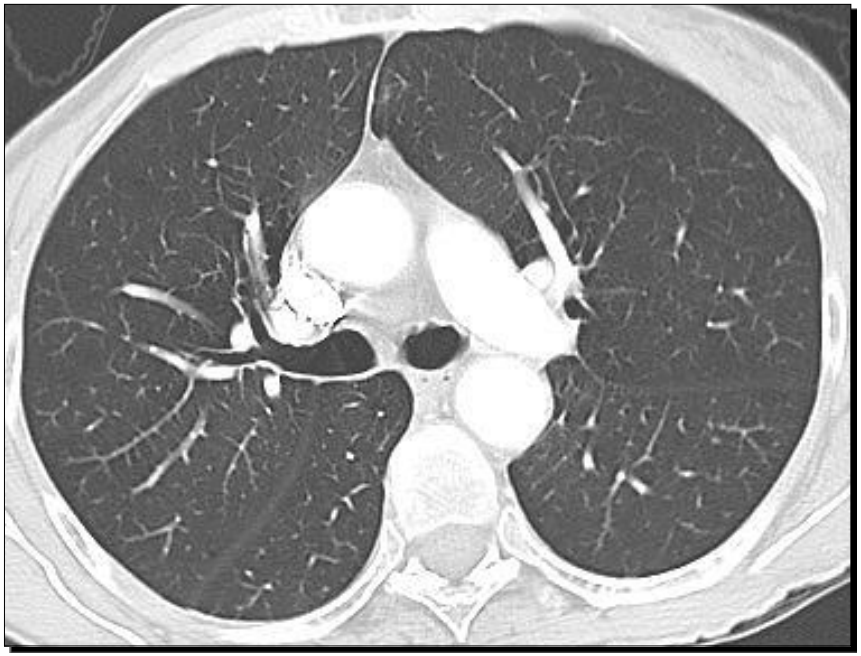
Bilateral smooth thickening of the interlobular septa
Thickening of the fissures



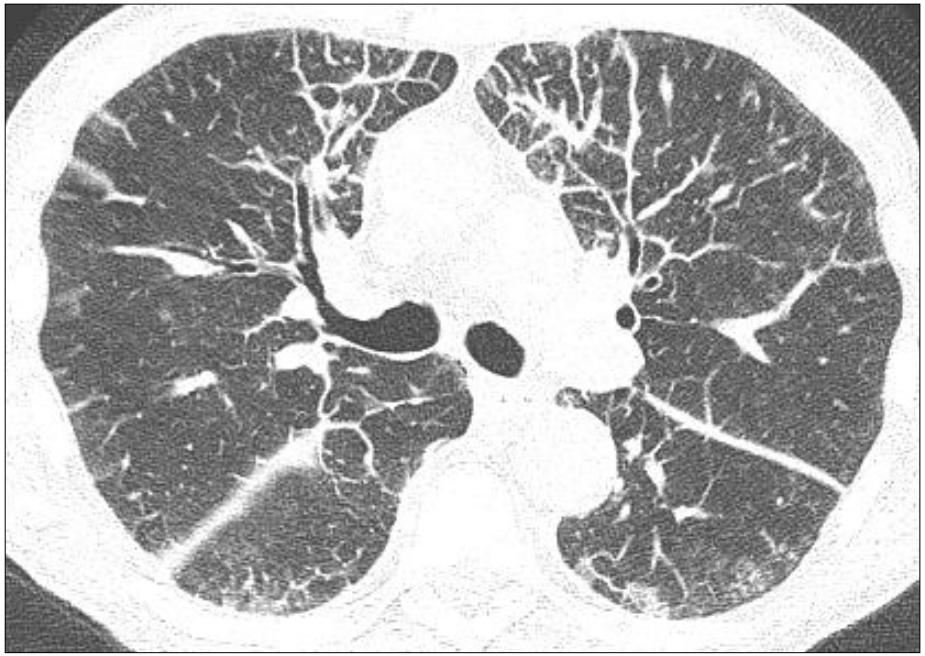


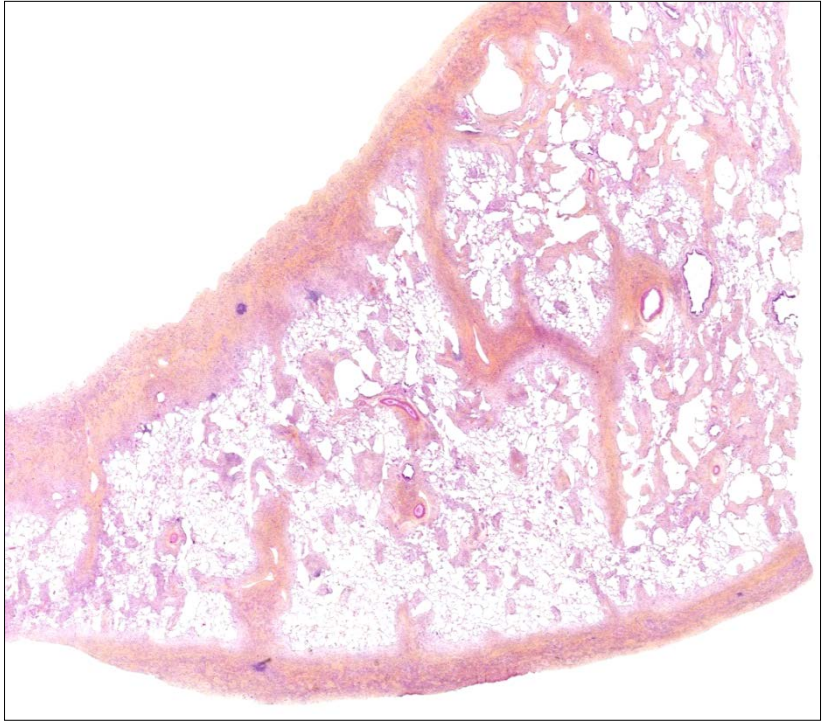
Normal Appearance

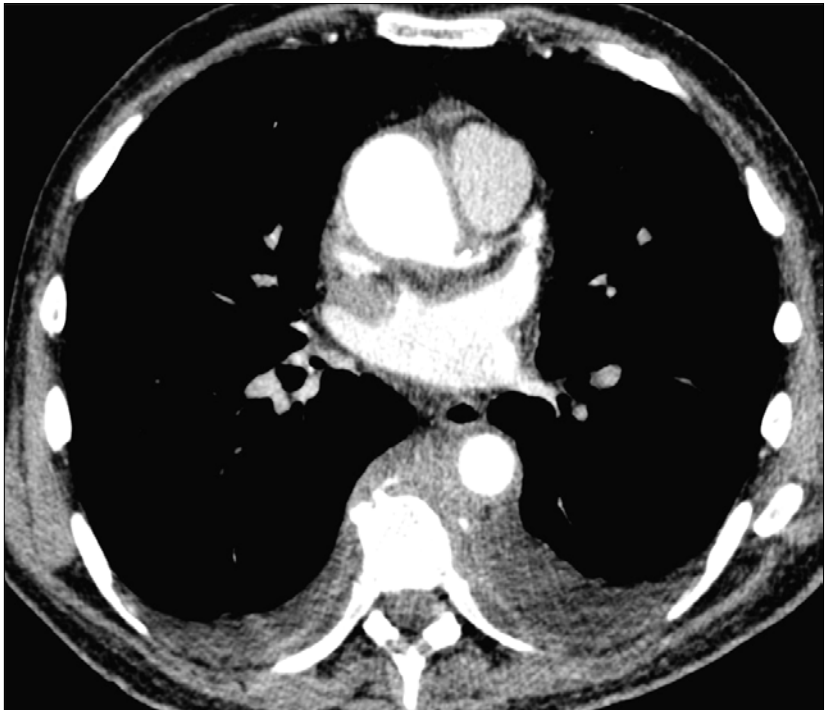




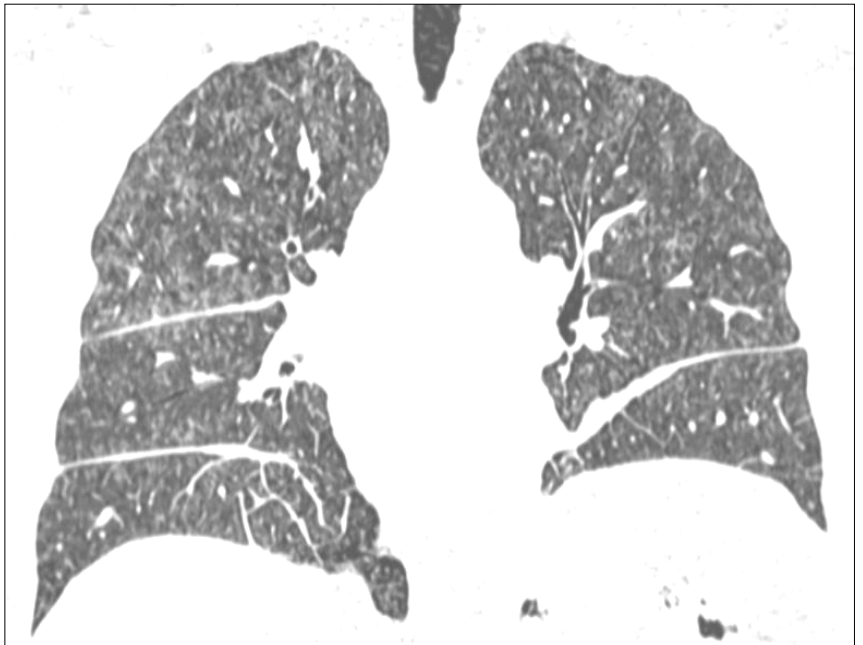
Erdheim Chester Disease



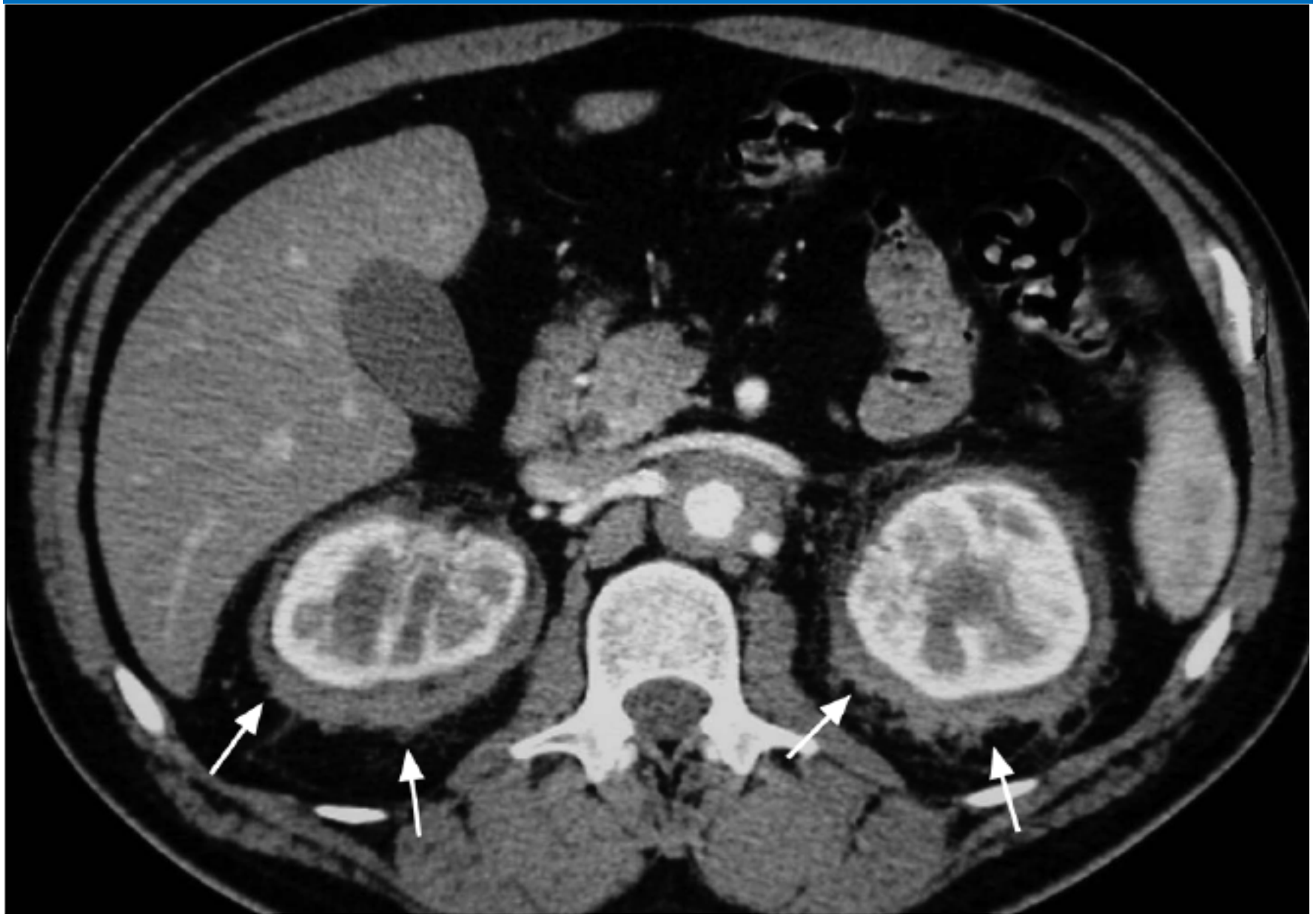




Small nodular opacities and GGO / Thickening of the fissures



Bilateral and symmetric perirenal infiltration with irregular bands (hairy appearance) / Circumferential sheating of aorta

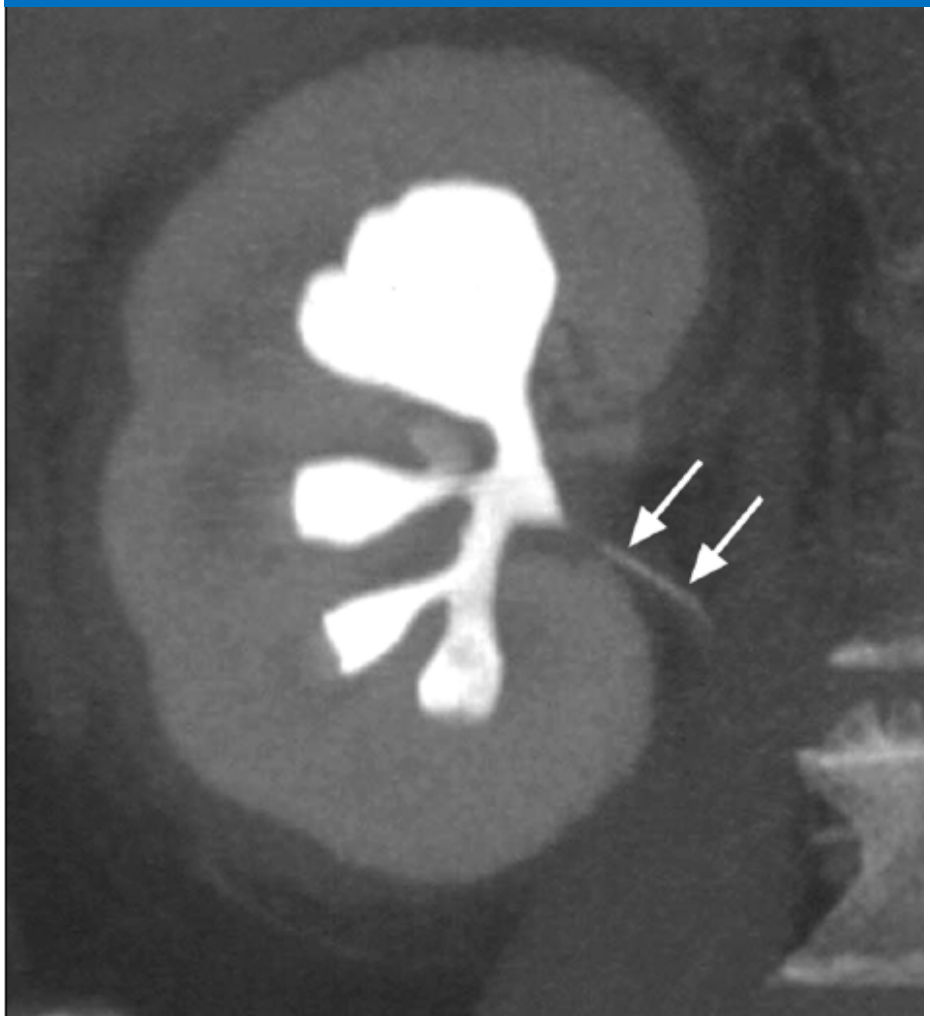


Bilateral and symmetric perirenal infiltration with irregular bands (hairy appearance)

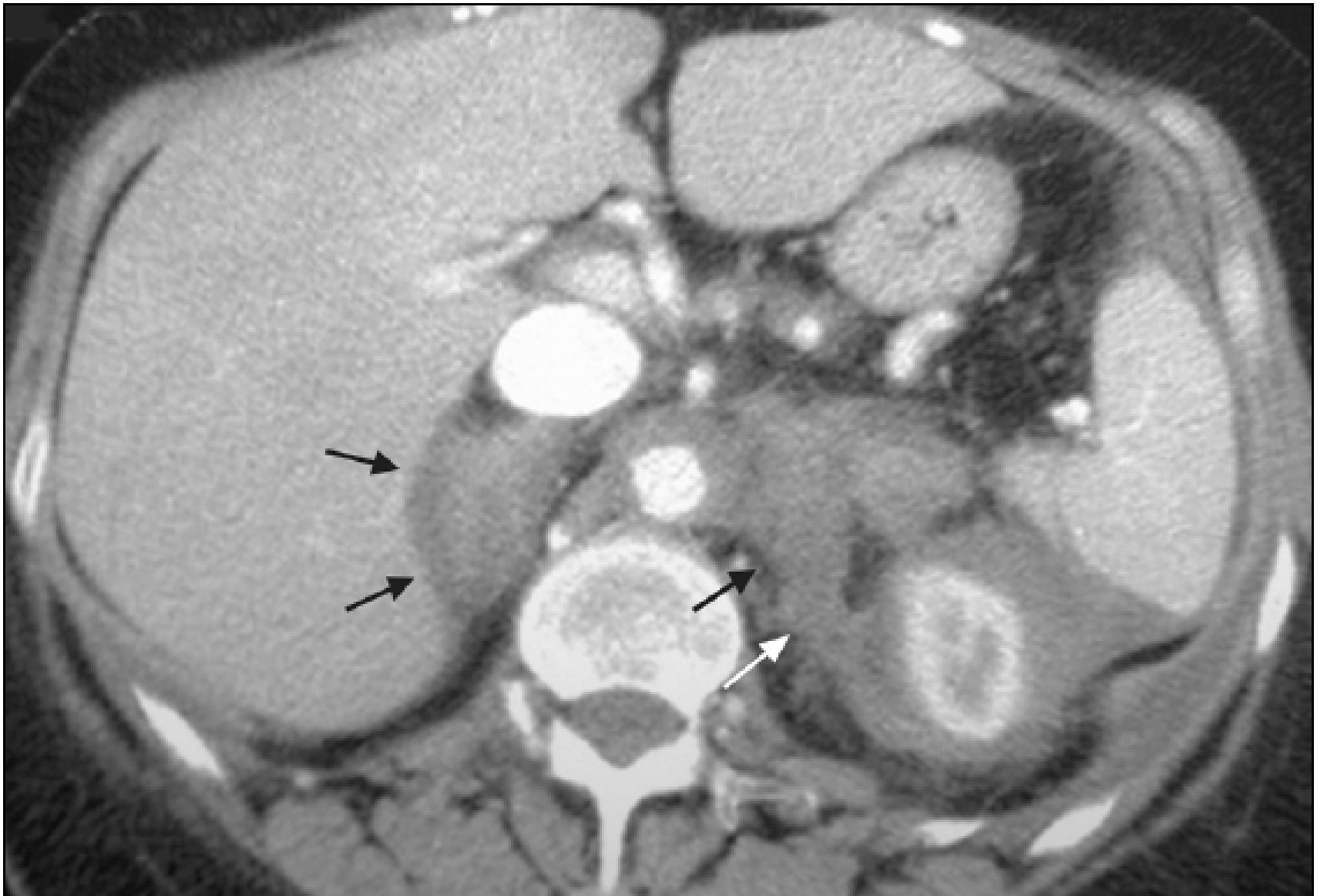
ECD confirmed by perirenal biopsy
Distal portion of left renal artery and superior mesenteric artery infiltrated and sheated

Bilateral asymptomatic perirenal infiltration extending and expending sinus of left kidney
Homogeneous circumferential periaortic infitration

ECD diagnosis obtained from perirenal biopsy
Bilateral pelvocalicectasis due to obstruction of upper portion of ureters / Perirenal infiltration



Extension of perirenal infiltration into adrenal fossae and surrounding adrenal gland; left perirenal infiltration extending into anterior pararenal space



CT and MR Imaging Findings of Cerebral, Facial, and Orbital Involvement in Erdheim-Chester Disease

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Cerebral, Facial, and Orbital Involvement in Erdheim-Chester Disease

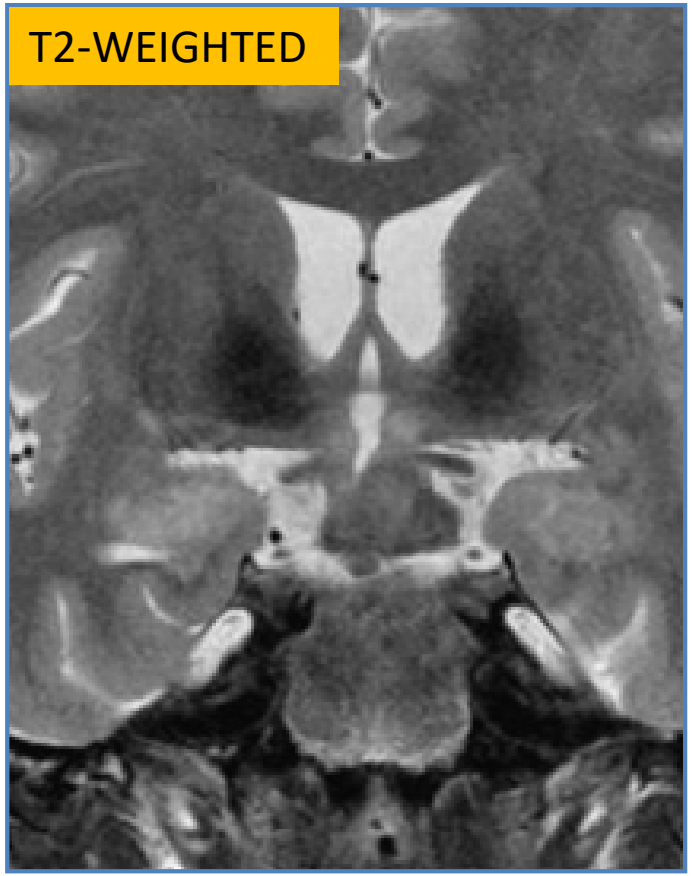
- Hypothalamic-pituitary axis involvement
- Meningeal Lesions
- Intraaxial Lesions
- Vascular Involvement
- Orbital Involvement
- Sinus and Skull Involvement

Drier. Radiology; 2010; 255: 586

Hypothalamic-pituitary axis involvement

MR images in 30-year-old man with diabetes insipidus

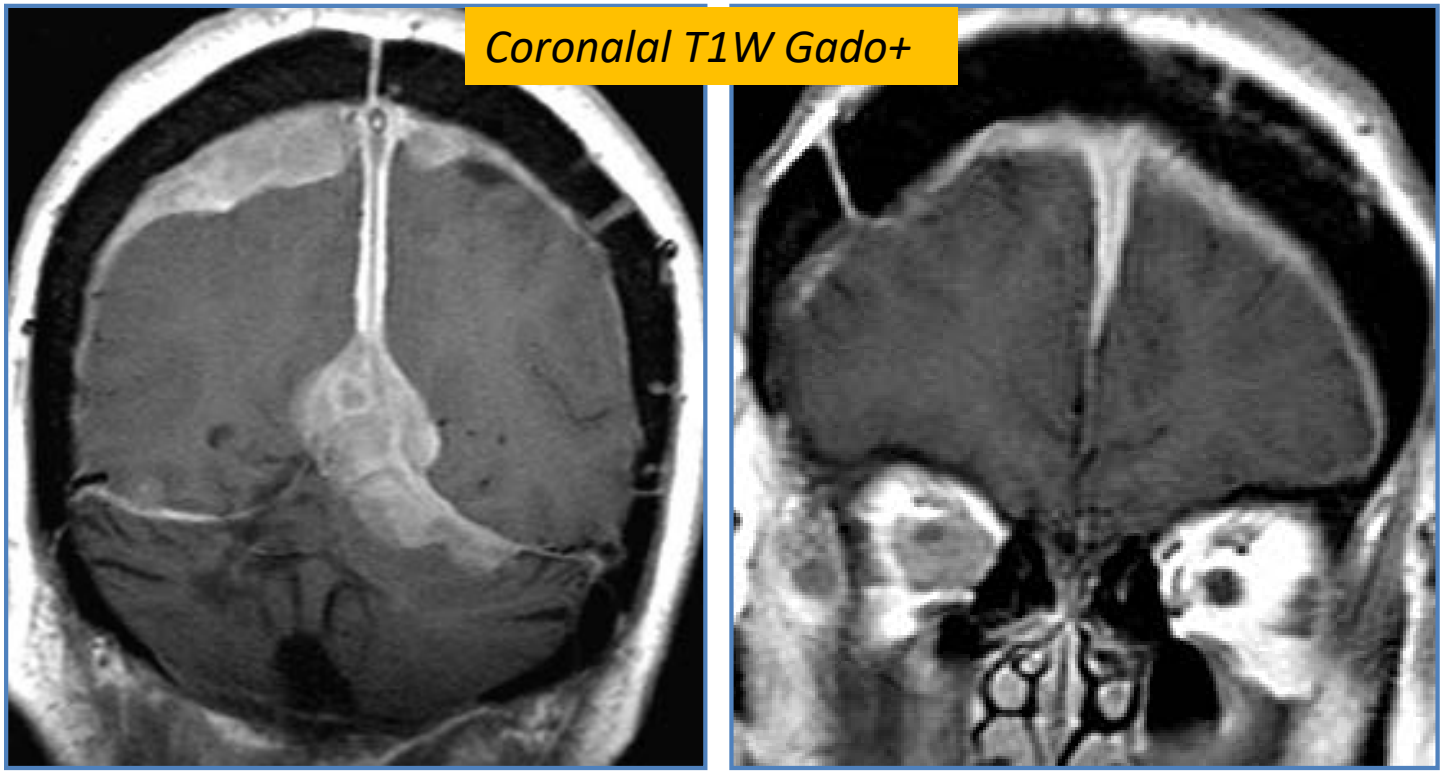
Nodular mass of indibular stalk, with homogeneous intense enhancement after gadolinium-based contrast material



Meningeal Lesions

Coronal T1-eighted gadolinium-enhanced MR images in 62-year-old woman

Multiple enancing dural masses with diffuse ehancing dural thickening and diffuse skull bone thickening
Diffuse linear dural thickening and enhancement



Meningioma-like mass lesions / Diffuse perimeningeal thickening

Intraaxial Lesions

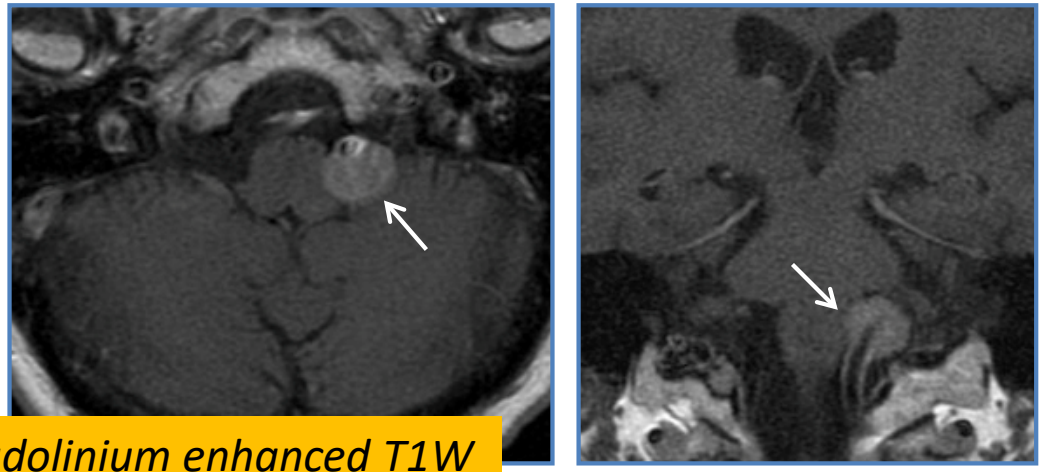
Multiple enhancing intraaxial (supratentorial or infratentorial) focal nodules or masses having isointense signal onT1W images, an iso- or hypointense signal on T2W images, and intense homogeneous enhancementon gadolinium-enhanced T1W images

Axial images in 44-year-old man with cerebellar ataxia: Symmetric hyperintense signal on T2W images in peridentate regions

Bilateral symmetric high signal intensity in the dental nucleus areas on T2W images and corresponding low signal intensiy on T1W images

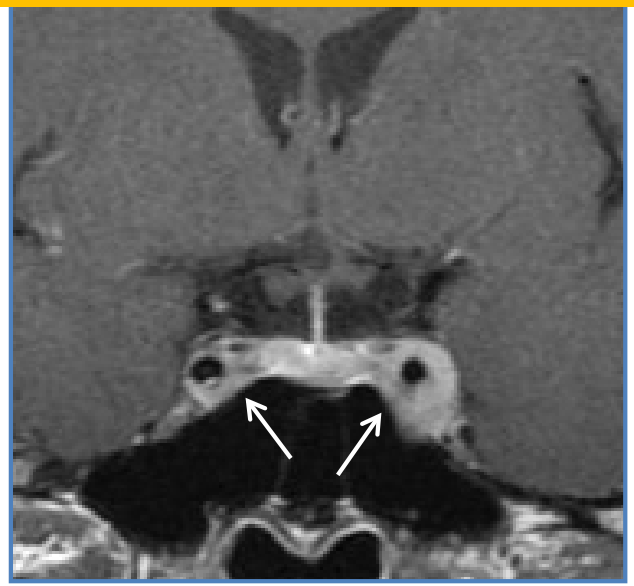
Vascular Involvement

Encasement of the left vertebral artery by a homogeneously enhancing mass



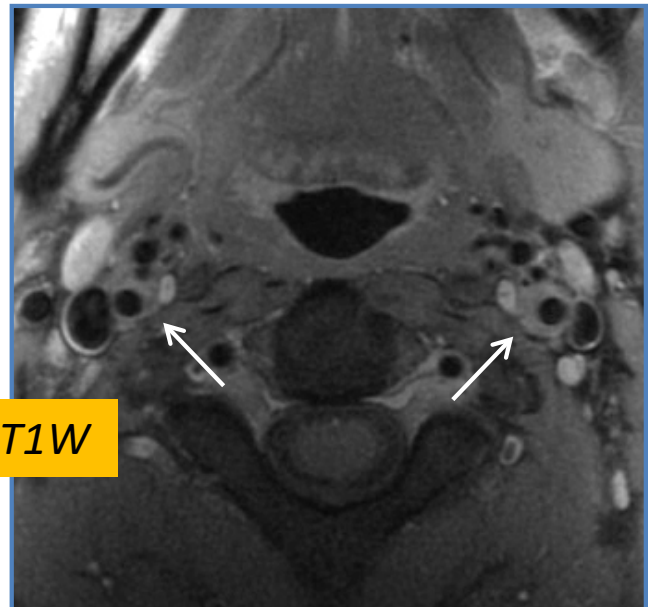
Gadolinium enhanced T1W

Coronal Gadolinium enhanced T1W



Intracranial extensionof perivascular infiltration along both internal carotid arteries

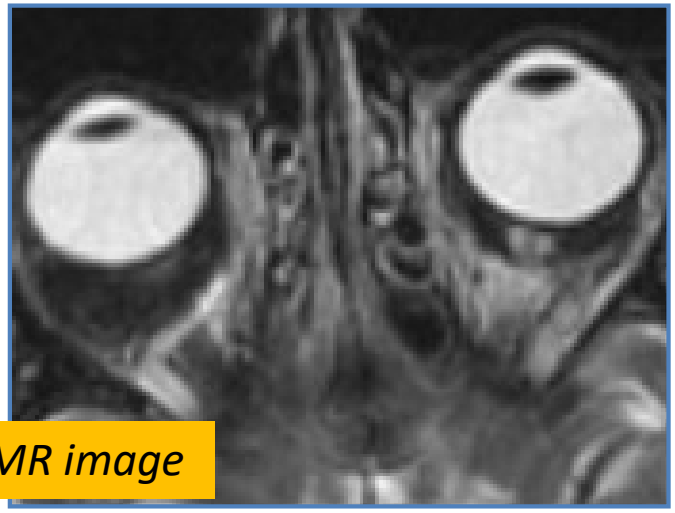
Bilateral cervical pericarotid infiltration



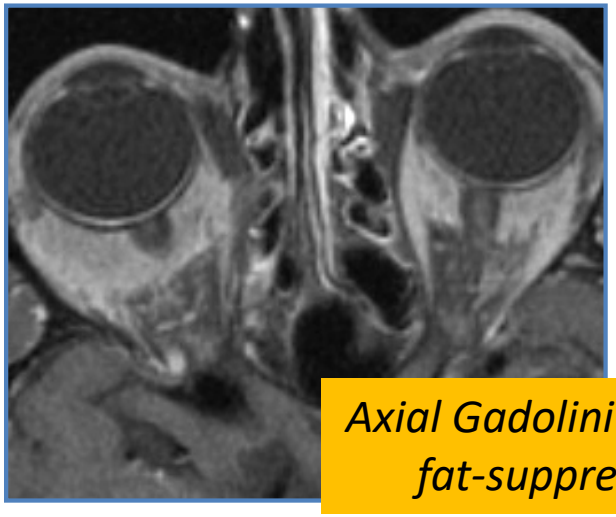
Axial Gadolinium enhanced T1W

Orbital Involvement

MR images in 40-year-old man with exophtalmos: Bilateral retro-ocular intraconal infiltration having low signal intensity and being enhancing



Axial T2W MR image

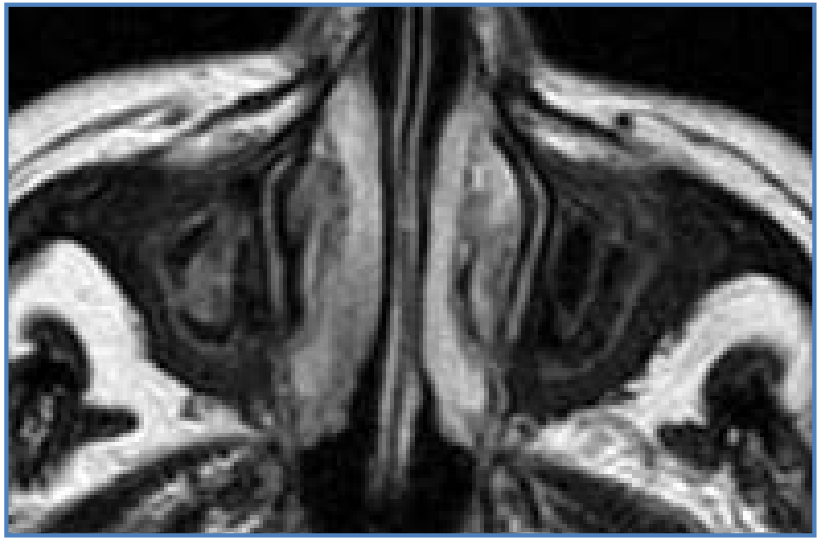


Axial Gadolinium enhanced fat-suppressed T1W

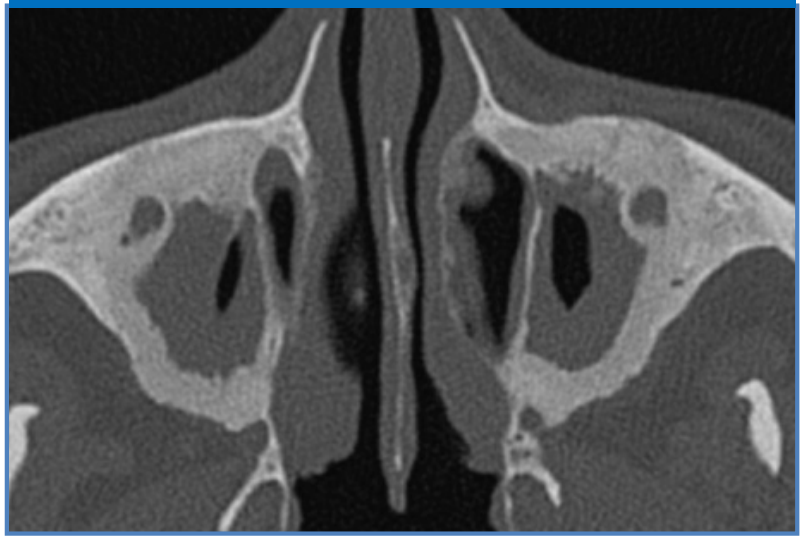
Bilateral or unilateral intraconal masses. Some large masses may extend to extraconal space

Sinus and Skull Involvement

T2-W MR: Bilateral maxillary sinus wall thickening with low signal intensity



Corresponding CT scan: Bilateral osteosclerosis



Bilateral maxillary and sphenoid sinus wall osteosclerosis with a hypointense signal on both T1 and T2–weighted MR images
Bilateral ethmoidal cells osteosclerosis