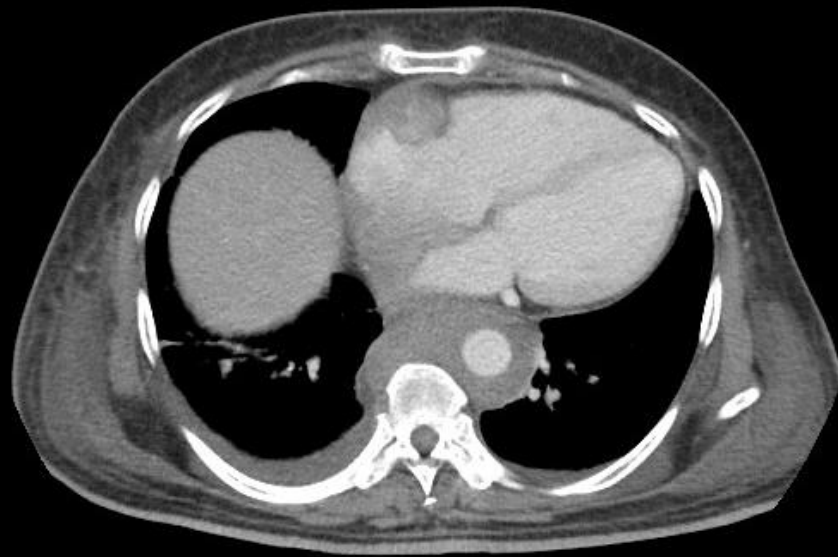


Case of the day : Case 4 - GU

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Thailand



Case of the day : GU



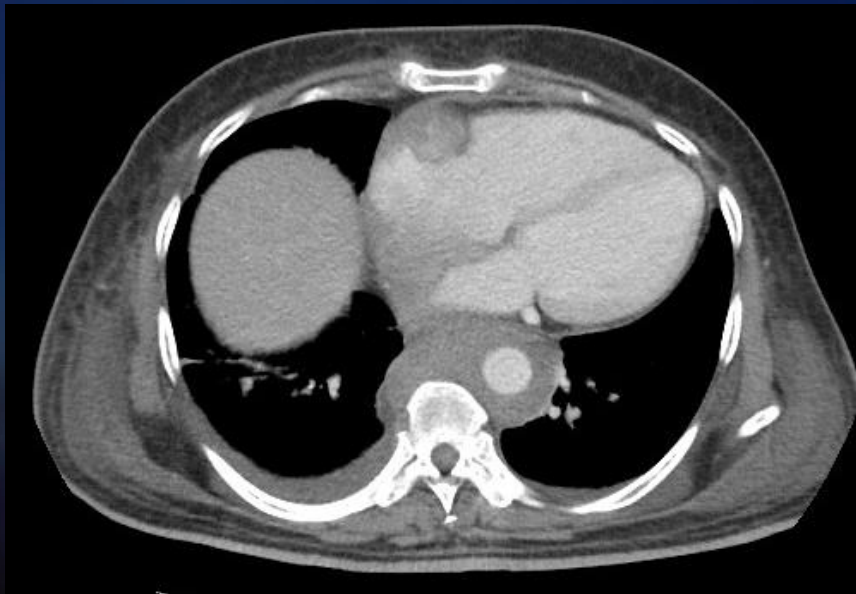
What is the most likely diagnosis?

- A. Retroperitoneal fibrosis
- B. Erdheim-Chester disease**
- C. Lymphoma
- D. Retroperitoneal sarcoma
- E. Castleman disease



Findings

- CT imaging of the chest and abdomen revealed extensive enhancing soft-tissue encasement of the aorta and kidneys



Diagnosis

- Erdheim-Chester disease (ECD)



Point of learning : ECD

- ECD is a non-Langerhans cell histiocytosis characterized by multiorgan xanthomatous infiltration
- Orbital and central nervous system involvement are common findings
- Retroperitoneal infiltration is present in one-third of patients and manifests similar to other cases of retroperitoneal fibrosis
- Cardiovascular involvement typically manifests as circumferential infiltration of the thoracic and abdominal aorta
- The prognosis of patients with ECD is poor. The 5-year survival rate was initially reported as 41%, but it has increased to 68% after interferon alpha administration in one series. Steroid therapy has not been shown to affect survival

