Granulomatosis with polyangiitis

- **“Wegener's granulomatosis”**
- Both ocular and orbital, have been reported in 40-50% of GPA patients and can occur in either the classic or limited form of the disease.
- Ophthalmologic disease occasionally can be the initial clinical manifestation in the limited form of granulomatosis with polyangiitis.
- Multi-system systemic necrotizing non-caseating granulomatous vasculitis affecting small to medium-sized arteries, capillaries and veins.

Granulomatosis with polyangiitis

- Orbital granulomas can be characterized as
  - contiguous: extension of granulomatous disease of the paranasal sinuses
  - focal: arising primarily within the orbit
- Complications include optic nerve compression with subsequent atrophy and visual loss.
- Extension of the disease to the cranial nerves traversing the cavernous sinus may manifest as Tolosa-Hunt syndrome with painful opthalmoplegia.

Imaging

- Diffuse inflammatory infiltrate that molds to the orbital contour
- proptosis
- usually associated with adjacent paranasal sinus or nasal disease

GPA