GIANT CELL ARTERITIS
(TEMPORAL ARTERITIS, HORTON'S DISEASE, CRANIAL ARTERITIS)

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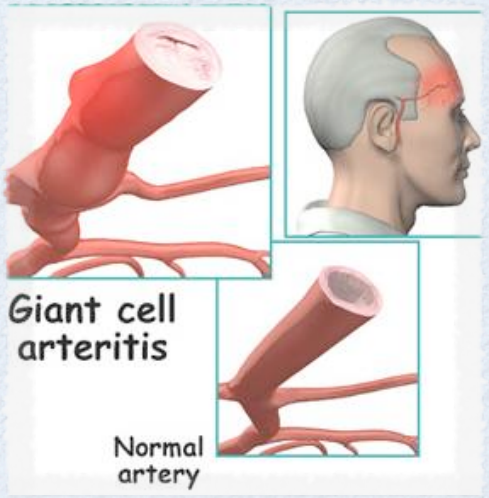
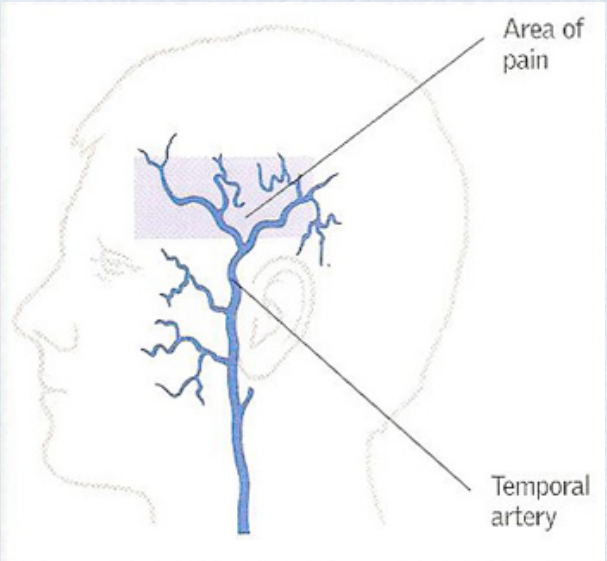
GIANT CELL ARTERITIS

- GCA, the commonest of all the vasculitides, is a chronic vasculitis of large and medium- sized blood vessels (Smeeth et al. 2006).
- Patients with GCA have damage to the blood vessel wall, which is primarily the result of cellular immune mechanisms.
- The inflammatory response results in the formation of multinucleated giant cells that represent the histological hallmark of GCA.



GIANT CELL ARTERITIS

- GCA mainly affects the cranial branches of arteries arising from the aortic arch and respects the intracranial arteries, which lack internal elastic lamina.
- The superficial temporal artery is involved in almost all patients, i.e., the term ”temporal arteritis”.



EPIDEMIOLOGY

- The incidence of GCA is 3 per 100000 per year in Rochester, Minnesota (Hauser KA et al. 1971) and 9 per 100000 per year in Göteborg, Sweden (Bengtsson BA et al. 1982).
- GCA is rare in Asians and African Americans and is most common in persons of British or Scandinavian heritage (Maida E et al. 1987).
- GCA was found in 1,7% of 889 postmortem examinations (Ainsworth RW et al. 1961).
- The incidence of GCA rises dramatically with increasing age after age 50, from 17,4 per 100000 (Hutchinson J. 1890) to 22 per 100000 (Ross Russel RW 1959).
- The mean age at diagnosis is about 70 years (Hoffman GS et al. 2002).
- GCA is 2- 4 times more common in F than in M (Palm E 1958).
- An association with human leukocyte antigen (HLA) DR4 antigen was suggested (Nordborg E et al. 2003).

THE IHS DIAGNOSTIC CRITERIA FOR GCA (ICHD- II)

- A. One or more of the following:

  1. Swollen and tender scalp artery (usually superficial temporal artery)

  2. Elevated red blood cell sedimentation rate

  3. Disappearance of headache within 48 hours of steroid therapy

- B. Temporal artery biopsy demonstrating GCA.
- C. Headache as a new symptom or of a new type occurs in close temporal relation to oncet of GCA.

SYMPTOMS - 1

- **Headache** is the most common presenting symptom (33%) and is a prominent symptom in 70% of the patients (Salvarani et al. 1987; Solomon and Cappa 1987; Gonzalez- Gay et al. 2005).
- The headache quality is described as throbbing, generalized, and continuous.
- Any new-onset headache in a patient aged >50 with an **elevated erythrocyte sedimentation rate** has to be considered as a GCA until the contrary is proven (Dasgupta et al. 2010).
- The most important feature is that the headache is either a new finding in a patient without a history of headaches or a new headache type in a patient with a history of chronic headaches.
- Approximately 5% of patients experience **visual scintillations,** suggestive of a migraine aura (Caselli et al., 1988a; Campbell and Caselli, 1991).

SYMPTOMS -II

- **Occipitonuchal pain** may result from vasculitic involvement of the occipital arteries (Caselli et al., 1988).
- **Jaw claudication** occurs in approximately 40% of patients and is the initial symptom in roughly 4% (Caselli et al. 1988).
- **Tongue claudication** occurs in approximately 4% of patients and is rarely the initial symptom (Caselli et al., 1988).
- Approximately 15% of patients with CGA have **carotidynia** (Caselli et al., 1988).
- One or more **systemic manifestations**, including fever, malaise, fatigue, anorexia, and weight loss, are present in most patients (Salvarani et al. 1987; Solomon and Cappa 1987; Gonzalez- Gay et al. 2005).

SYMPTOMS - III

- Irreversible partial or complete **visual loss** in one or in both eyes occurs in less than 20% of patients.
- **Diplopia** occurs in roughly 2% (Caselli et al., 1988a) to 14% (Hollenhorst et al., 1960) of patients with GCA.
- About 4% of patients have **TIA or stroke** at some point during the course of GCA (Caselli et al. 1988b).
- **Aortic arch syndrome** occurs in about 10-15% of patients with GCA, presenting with claudication of the arms and rarely the legs, and absent or decreased pulses in the neck or arms.
- About 14% of all patients have **neuropathies,** including mononeuropathies and peripheral polyneuropathies of the upper or lower extremities (Caselli et al. 1988a).

Laboratory investigations and imaging

- The American College of Rheumatology classification criteria for GCA include an **ESR** of 50 mm/h or more.
- Temporal artery **biopsy** remains the gold standard for investigation (Robb-Nicholson et al. 1988; Mukhtyar et al. 2009). Biopsies should show interruption of the internal elastic membrane with infiltration of mononuclear cells in the arterial wall.
- Ultrasonography can show a typical hypoechoic "halo" around affected temporal arteries (representing **vessel wall edema,** as well as arterial stenosis and occlusion) (Karassa et al. 2005; Schmidt 2007).
- **3T MRI** using intravenous contrast may show increased vessel wall thickness and edema, with increased mural enhancement post contrast and luminal stenosis (Bley et al. 2005).

TREATMENT -1

- The mainstay of therapy remains **high-dose corticosteroids**, which should be initiated immediately when clinical suspicion of GCA is raised (Mukhtyar et al. 2009; Dasgupta et al. 2010).
- Uncomplicated GCA (no jaw claudication or visual disturbance): 40-60 mg prednisolone daily (Kyle 1991).
- Evolving visual loss or amaurosis fugax (complicated GCA): 500 mg to 1 g of IV methylprednisolone for 3 days before oral corticosteroids (Chevalet et al. 2000; Mazlumzadeh et al. 2006)
- Established visual loss: 60 mg prednisolone daily to protect the controlateral eye (Cgan et al. 2001; Foroozan et al. 2003)
- Symptoms of GCA typically respond rapidly to this treatment, followed by resolution of the inflammatory response.

TREATMENT - II

- The initial dose of 40- 60 mg prednisolone has to be continued until symptoms and laboratory abnormalities resolve (at least 3- 4 weeks).
- Then a slow and gradual decrease is considered wth a reduction by 10 mg every 2 weeks to 20 mg; then by 2,5 mg every 2- 4 weeks to 10 mg; and then by 1 mg every 1- 2 months provided there is no relapse.
- Although corticosteroids can be discontinued within 1- 2 years in most cases, some patients will require long-term low-dose therapy.
- Patients should be closely monitored for evidence of relapse, disease-related complications, and glucocorticosteroid- related complications.
- Experience using conventional disease- modifying drugs such as methotrexate is mixed, and biological therapies require further evaluation for their steroid- sparing potential (Borg and Dasgupta 2009).

DIFFERENTIAL DIAGNOSIS OF HEADACHE IN THE ELDERLY PATIENT

- GCA
- Brain tumor
- Carcinomatous meningitis
- Aneurysm
- Monoclonal gammopathies
- Polyclonal hyperglobulinemias
- Connective- tissue diseases
- Leukemias
- Lymphomas
- Carcinomas
- Sarcomas

Thanks !

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