

Otorrinolaringología

Tema 27. Manifestaciones otorrinolaringológicas en la granulomatosis de Wegener



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Treatment of upper airway problems in Wegener Granulomatosis

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General aspects

- Unfrequent Autoimmune disease.
- Unknown etiology.
- Main characteristics: granulomatous inflammation and anti-neutrophil anticytoplasmic antibodies (ANCA).
- Wide clinical spectrum (local-systemic).

Epidemiología

- **Incidence rate: 6-12/million in UK, Germany y Norway:**
- *Holle et al. Rheum Dis Clin N Am 2010;36:507-26.*
- **South of Europe: less frequent. 2,95/million/year:**
- *Gonzalez Gay et al. Arthritis Rheum 2003;43:388.-93.*
- **South of Sweden: prevalence (1-1-2003): 16/100000:**
- *Mohammad et al. Rheumatology 2007;46:1329-37.*
- **USA: prevalence of 3/100000:**
- *Cotch et al. Arthritis Rheum 1996;39:87-92.*

Epidemiología

- **Age at diagnosis**: average 55 años (wide range).
- **In adults**: man = woman.
- **In children**: girls/boys 4:1:
- *Akikusa et al. Arthritis Rheumatism 2007;57:837-844.*

Clasificación

- **Local disease**: only upper or inferior respiratory tract, without signs of systemic vasculitis.

- **Systemic disease**: early, generalized and severe.

- **Staphylococcus aureus:**

- WG: nasal colonization in 72%.
- Chronic sinusitis: 28%.
- Healthy people related to the health system: 25%.
- Rheumatoid arthritis: 46%.
- Hospital workers: 58%.
- *Laudien et al. Clin exp Rheumatol 2010;28:51-5.*

Pronóstico

- **Lack of ENT involvement is a sign of bad prognosis:**
- *Bligny et al. Arthritis Rheum 2004;51:83-91.*

Frequency of upper clinical manifestations in WG

- **72,3-99% of patients with WG have ENT manifestations:**
 - Reinhold-Keller et al. *Arthritis Rheum* 2000;43:1021-32.
 - Mcdonald et al. *ANCA-associated vasculitis* 1993;309-13.

ENT physical exam



Manifestaciones otológicas (19-61%)

- Otitis media with effusion.
- COM.
- Acute otitis media.
- Granulomatous inflammation of the middle ear.
- Sensorineural hearing loss and vertigo.
- External otitis, Auricular chondritis.
- Facial palsy (uni or bilateral).
- **Palsy of other cranial nerves: IX, X, XI:**
- *Takagi et al. Laryngoscope 2002;112:1684-90.*

Otitis media with effusion

- Eustachian tube stenosis.

 • **Otitis serosa.**

Oral manifestations

- Unfrequent.
- Strawberry gingival hyperplasia.
- Deep oral ulcers of the tongue, cheeks gums and palate.
- **Hiperthrophy of submandibular and/or parotid glands:**
 - *Specks et al. Arch Otolaryngol Head Neck 1991;117:218-223.*

Sinonasal manifestations

- Bleeding.
- Crusts.
- Purulent discharge.
- Nasal pain.
- Anosmia.
- **Nasal congestion:**



- Cannady SB et al. *Laryngoscope* 2009;119:757-61.

Sinonasal exam



Rinoscopy

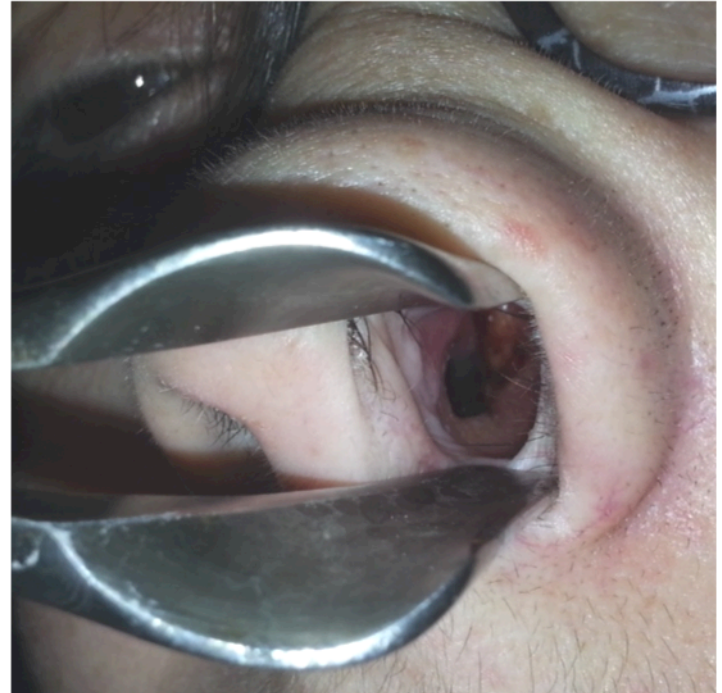


Nasal endoscopy

Nasosinusual clinical manifestations

- Vasculitis of the Kiesselbach area vessels.
- Cartilage necrosis.
- **Septal perforation:**

- Cannady SB et al. *Laryngoscope* 2009;119:757-61.



Nasosinusal clinical manifestations

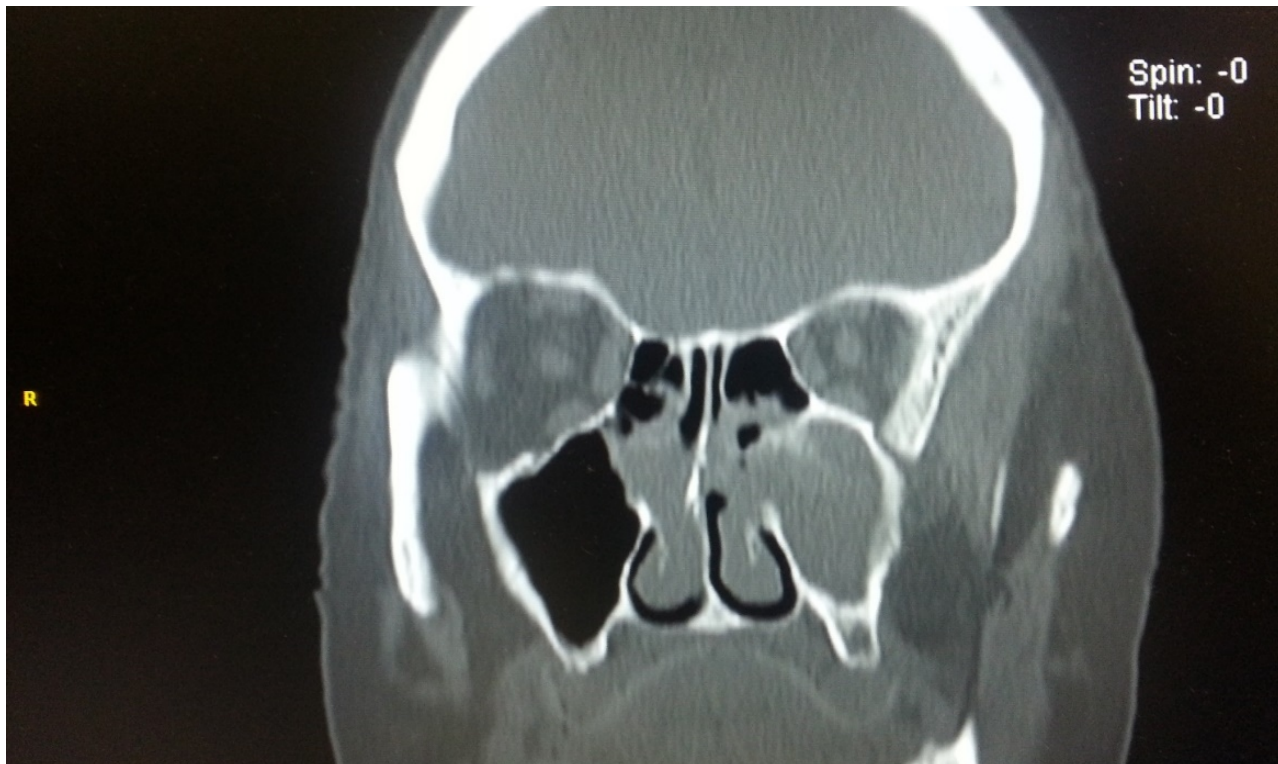
- **Saddle nose:**

- *Congdon et al. Laryngoscope 2002;112:731-7.*



Sinonasal clinical manifestations

Inflammation → edema → sinus ostium stenosis → sinusitis.



Orbital tumor

- 15% of the patients with WG (2% as the first manifestation). Unilateral.
- Ocular pain, proptosis, diplopia...
- Local inflammation or sinus infiltration o nasofaríngeal.



Orbital tumor

- **Nasolacrimal disease (8%):**
- *Kwan et al. Br J Ophthalmol 2000;84:329-31.*

Subglottic stenosis

- Most frequent laryngeal/tracheal manifestation.
- Mild to severe disease (it could be letal).
- First manifestation (1-6%) or during follow up.
- Scar fibrosis (follow up is independent to WG or treatment) or active disease.

Subglottic stenosis

- Frequency: 8,5-50%.
- **Clinical trial with etanercept**: 8,3% at diagnosis and 17,8% at the end of the study:
 - *Stone et al. Arthritis Rheum 2003;48:2299-309.*

Estenosis subglótica

- More prevalence in women.
- Average age at presentation: 26 años.
- More frequent when the disease diagnosis is made before 20 years old.
- **28-44% is associated with bronchial stenosis:**
 - *Summers RM. Chest 2002;121:242-250.*

Estenosis subglótica

- **First manifestation**: eritematous disease and ulcerations. Circumferential. In most of the cases asymptomatic or very few clinical manifestations.
- **Without proper diagnosis**: scar fibrosis and secondary disnea.
- **Frequently confused with asma.**

Subglottic stenosis

- **Sometimes the only clinical manifestations of the disease:**
diagnosis by biopsy or ANCA+:
 - *Guerrero et al. J Clin Rheumatol 2001;7:91-6.*

Other clinical test

- CT scan.
- MRI.
- Flow/volumen curves.

Diferential diagnosis of subglottic stenosis

- Relapsing polychondritis.
- Sarcoidosis.
- Amiloidosis.
- ...
-
- Idiopatic.

Tracheal stenosis

- Similar characteristics to subglottic.

Tracheal stenosis: differential diagnosis

- Sarcoidosis.
- Behcet disease.
- **Difuse involvement**: relapsing polichondritis, osteochondroplastic tracheopathy, amiloidosis, papillomatosis.
- Endotracheal intubation.
- Postinfectious stenosis.
- Crohn disease.

Treatment

- Surgical treatment can reduce the morbidity of ENT manifestations.
- **But be careful:** proper selection of candidates to surgery. Patients who doesn't get a good response to inmunosupresor treatment.
- **Surgical treatment:** best results in remmision phase.

Treatment of middle ear disease

- **Middle ear**: ¡be careful with surgery!:
 - Most patients only need medical treatment. Good response to immunosupresor treatment.
 - Surgical treatment: biopsy (very few times with a positive result) or aftermath.
- *Takagi et al. Laryngoscope 2002;112:1684-90.*
- Transtympanic tubes.
- Timpanoplasty/mastoidectomy.

Ear disease treatment

- **Inner ear**: sensorineural hearing loss:
medical/hearing aids/cochlear implant:
- *Abou-Elhmd et al. J Laryngol Otol 1996;110:958.*



Sinonasal disease treatment

- **Nasal crusts**: irrigations with saline.
- Oral or topical Antibiotics. Fatty creams.
- Severe cases: ent cleaning with care.
- **Surgery**: in active phase, only biopsy (> 5 mm).



Sinonsal surgical treatment in WG

- **Sinusitis**: endoscopic surgery in selected patients.



Sinonasal surgical treatment

- **Septal perforation**: it is better to avoid reconstructive surgery.



- **Long term remission patients:**
 - Calvarial bone (temporal). With an L shape:
 - *Taha et al. Arch Facial Plast Surg 2008;10:305-11.*
 - Rib cartilage.
 - Iliac crest.
 - Auricular cartilage:
 - *Congdon D et al. Laryngoscope 2002;112:731-7.*

Treatment of the orbital tumor

- **Medical treatment**. Corticosteroids/ciclofosfamida, rituximab.
- **Surgery**: orbital decompression (severe pain, proptosis or nerve compression of the optical nerve).
 - *Fechner FP et al. Laryngoscope 2002;112:1945-50.*

Sinonasal treatment

- **Nasolacrimal disease**: dacriocistorinostomy. Risk of infection or necrosis. Not active phase with a clean nose without crust and infection.

Treatment of subglottic stenosis

- Observation/medical/tracheostomy and surgery to increase the aerial path.
- **Acute obstruction**: temporal o permanent tracheostomy (6-80%):
 - *Langford et al. Arthritis Rheum 1996;39:1754-60*
 - *Alaani et al. J Laryngol Otol 2004;118:786-90.*

Treatment of subglottic stenosis

- 20-26% have a good response to systemic medical treatment.
- **74-80% need surgery:**
 - *Langford et al. Arthritis Rheum 1996;39:1754-60*
 - *Schokkenbroek et al. Eur Arch Otorhinolaryngol 2008;265:549-55.*

Treatment of subglottic stenosis

- Not active disease.
- Dilatation + local corticosteroid injections.
- Laser surgery (CO² o Nd:YAG).
- ¿Stents?
- Laryngotracheoplasty.

Dilatation + Corticosteroids

- **Endoscopic treatment**: unpredictable results.
- Dilatation with a tracheoscope of Groningen.

Laser surgery

- With or without corticosteroids.
- Variable results.
- **Repeated treatment:**
 - *Shvero et al. Yonsei Med J. 2007;31;48:748-53.*

Histologic diagnosis of WG with head and head biopsy

- Sinonasal biopsy with local anesthesia.



Head and neck biopsy

- Paranasal sinuses (50%).
- Nasal fossa (20%-53%).
- Middle ear.
- **Subglottis:**
 - *Tsuzuki K et al. Auris Nasus Larynx 2009;36:64-70.*
 - *Maguchi et al. Auris Nasus Larynx 2001;28:S53-8.*

Summary

- Systematic ENT examination in patients with diagnosis or suspicion o WG.
- In the presence of active disease: physical exam and biopsy (at the edge of active disease).
- Surgical treatment only in patients without active disease to treat the aftermaths.