Granulomatosis with Polyangiitis (Wegener’s Granulomatosis)

Surgical Interventions for Diagnosis & Treatment

Sasan Dabiri, M.D. Assistant Professor
Department of Otorhinolaryngology – Head & Neck Surgery
Amir A’lâm hospital
Tehran University of Medical Sciences
October 2012
Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

Introduction

1931 : Klinger

1939 : Wegener: About a strange rhinogenous granulomatous with special participation of arterial system and the kidney.

1954 : Godman & Churg

1985 : ANCA discovered

1994 : CHCC new nomenclature

Friedrich Wegener
1907 - 1990
# Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

## Introduction

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Endoscopy</td>
<td>Endoscopic Sinus Surgery</td>
</tr>
<tr>
<td>Biopsy</td>
<td>Rhinoplasty</td>
</tr>
<tr>
<td>Head &amp; Neck</td>
<td>Tympanostomy Tube (VT)</td>
</tr>
<tr>
<td>Lung</td>
<td>Mastoidectomy</td>
</tr>
<tr>
<td>Kidney</td>
<td>Laryngotracheal Microsurgery</td>
</tr>
<tr>
<td>Other sites</td>
<td></td>
</tr>
</tbody>
</table>
Biopsy

- Central for diagnosis in suspected patients
  - 30% of Bx⁺ may be ANCA⁻

Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

Biopsy

• Terminology for Dx by Head & Neck Bx:
  – Diagnostic
  – Probable
  – Suggestive
  – Suspicious
  – Nonspecific

  • Granulomatous
  • Necrosis
  • Vasculitis

  • Head & Neck
  • Lung
  • Kidney
Biopsy

• Where for Bx?
  – Paranasal Sinuses: the most favorable result
  – Other Head & Neck sites: Less positive than PNS
  – Other sites: when No appropriate sites are found

➢ Nerve (sural) - Muscle - Skin - Lung - Kidney
Biopsy

• How to Bx?
  – Should include adjacent normal soft tissue (if possible)
  – Should be with less manipulation (later complications)
  – Preferably when disease is not in active phase
Biopsy

• How to Interpret?
  – Negative Predictive Value: 74% (→ high false negative)
  – Repeat Bx: when report is probable or less
  – Repeat Bx: when Negative Bx & ANCA
Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

Rhinologic Aspects

- Crusting
- Granulation
- Destruction
- Rhinorrhea
- **Nasal or Sinus Pain**
- Epistaxis
- Hyposmia

- Epiphora
- Perforated nasal septum
- Saddle-nose deformity
- Chronic Sinusitis
- Bony Sclerosis
- Orbital mass
- Orbital inflammation
Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

Rhinologic Aspects

• Septal perforation
Rhinologic Aspects

- Nasopharynx
Rhinologic Aspects

• Saddle nose deformity
Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

Rhinologic Aspects

• Rhinosinusitis
Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

Otologic Aspects

- Otitis Media with Effusion
- Chronic Suppurative Otitis Media
- Facial Nerve Palsy
- Meningitis
- Sensorineural Hearing Loss
- Vertigo
Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

Otologic Aspects

• Facial Palsy
Otologic Aspects

- Sansorineural Hearing Loss
Laryngologic Aspects

• Subglottic Stenosis
Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

Oral Aspects

• Ophthalmus ulcer
Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

Oral Aspects

- Strawberry gingiva
Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

Oral Aspects

• Palatal crust
Granulomatosis with Polyangiitis (Wegener’s) – Surgery for Dx & Rx

Treatment

- Endoscopic Sinus Surgery
- Endoscopic Orbital & Nasolacrimal Surgery
- Rhinoplasty
- Tympanostomy Tube (VT) insertion
- Mastoidectomy
- Laryngotracheal Microsurgery
Sincere Gratitude to

- Dr. Mohammadi Ardehali
- Dr. Dabir moghaddam
- Dr. Bastani nezhad
Thanks for Your Attention