Erdheim Chester disease (ECD) and ENT manifestations

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ECD Medical Symposium
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ECD involvement

Brain

Eyes (Xanthelasma)

Abdomen (Hairy kidney)

Heart (atrial tumor)

Vessel (Coated aorta)

Long bones (Distal femur and proximal tibia Osteoclerosis)
Clinical study

- Few studies in the literature: Only case reports
- Pitié-Salpêtrière Hospital
  - 121 patients with ECD
  - Retrospective study

→ 62% ENT signs
(clinical and/or radiological)
Clinical ENT manifestations

- In 31 % of cases
- Ear
  - Hearing loss, vertigo, tinnitus, external otitis...
  - External auditory canal stenosis ++
- Nose and sinus
  - Sinusitis and rhinitis

Rhinosinusitis (mucopus at middle turbinate)
Normal middle turbinate
Normal external auditory canal
External auditory canal stenosis
Radiological ENT lesions

- CT scanner and/or MRI: 70%
- Ears
  - Inflammation
  - External auditory canal stenosis ++
- Nose and sinus
  - Inflammation
  - Pathognomonic aspect: bilateral osteosclerosis

![Radiological images of normal and pathological sinus CT scans with annotations indicating normal and pathological CT scans.](image)
ENT lesions in ECD

- ECD is misunderstood and thus under-diagnosed
- ENT lesions are frequent in ECD, just after the bone lesions
- ENT lesions are under-diagnosed because
  - Clinical lesions are not specific
  - ENT imaging is not done systematically

- In case of pluri-organ lesions → ECD ?
- In case of ECD suspicion → Systematic ENT imaging
  → Nasal biopsy in case of nasal lesions
- In order to know the real prevalence of the ENT symptoms and signs
  → Questionnaire
Questionnaire

- In French or English
- For **every patients**, with or without ENT signs

How to find it

- By e-mail  
  [Ecdquestionnaire@gmail.com](mailto:Ecdquestionnaire@gmail.com)
- Directly at the end of this session
- Filled questionnaires, in the box, or sent by e-mail

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<thead>
<tr>
<th>Question</th>
<th>Yes</th>
<th>No</th>
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<tbody>
<tr>
<td>Have you ever consulted an ENT?</td>
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<td>If Yes, why and where?</td>
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<td>Have you ever had runny nose over a month?</td>
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<td>Have you ever had front head or cheekbone pain?</td>
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<td>Do you have current nosebleeds?</td>
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<td>Have you ever had more than 2 sinusitis?</td>
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<td>Have you ever had nose surgery?</td>
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<td>If Yes, why and where?</td>
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<td>Have you sometimes scratching or running ears?</td>
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<tr>
<td>Do you have a hearing loss or tinnitus?</td>
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<td>Have you ever had ear surgery?</td>
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<tr>
<td>If Yes, why and where?</td>
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Thank you for your time and answers.
Thank you for your attention and...

... for your answers to the questionnaire

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