Calcified cauliflower ear in relapsing polychondritis

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DESCRIPTION

A 59-year-old man with chronic obstructive pulmonary disease (COPD) presented with dyspnoea for 3 days. Physical examination revealed diffuse rhonchi over chest, and very hard, thick, deformed auricles except for the ear lobules, which were soft (figure 1A). Patient reported remote history of episodic pain and redness of his ears and nose, which improved with over-the-counter pain medicines. A radiograph of the ears showed calcification of the auricular cartilages (figure 1B, white arrow). A complete blood count showed neutrophilic leucocytosis of $13.1 \times 10^3/\mu\text{L}$ and chronic normocytic anaemia of 12.3 g/dL. Inflammatory markers including erythrocyte sedimentation rate and C reactive protein, and renal and liver function tests were normal. Prior work-up was negative for anti-nuclear and antineutrophil cytoplasmic antibodies. A clinical diagnosis of relapsing polychondritis (RP) was made based on the reported history and the Michet criteria.¹ Review of a prior lung function test ruled out any tracheobronchial pathology from RP. His dyspnoea improved with standard treatment for COPD exacerbation. No specific treatment for RP was initiated since he did not have symptoms suggestive of acute chondritis.

RP is a rare multisystemic connective tissue disorder characterised by recurrent, progressive inflammation of cartilaginous tissue.¹ Recurrent inflammation leads to cartilage destruction, and eventual fibrosis, sometimes with calcifications. The diagnosis is essentially clinical based on the

McAdam, Damiani or Michet criteria.¹ There is no pathognomonic laboratory finding. The most common presentation is auricular chondritis seen in 90% of patients.² Trauma and infection of the pinna can mimic RP. Auricular chondritis in RP is typically bilateral and spares the lobules.¹ With repeated attacks, the ear can become nodular and, in severe cases, floppy and deformed due to cartilage destruction. Microcalcific deposits can occur in the ear (as in our patient).³ There is a mean delay of about 3 years from first presentation to diagnosis of RP.³ There is no standard treatment for RP.¹ Non-steroidal anti-inflammatory drugs (NSAIDs) can help in limited disease like diffuse scleritis, episcleritis, non-severe forms of auricular and nasal chondritis and arthritis. Glucocorticoids are preferred when the disease is resistant to NSAIDs or is very severe. Immunosuppressants are useful in steroid-resistant disease or as steroid-sparing agents.² Increasing awareness about common clinical features, especially auricular involvement, can help in early diagnosis.

Learning points

- ▶ Relapsing polychondritis can have devastating complications and hence early diagnosis is crucial.
- ▶ Prompt identification of auricular signs can help in early diagnosis.

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Figure 1 (A) Deformed hard auricles except for the ear lobules, which were soft. (B) Radiograph of the ear showing calcification of the auricular cartilages.

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