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PHOTO QUIZ

Floppy ears and tracheal wall narrowing

F. Azeredo[1], A. Severo[1], G. Zanetti[2], E. Marchiori[2]*

[1]Department of Radiology, Fluminense Federal University, Rio de Janeiro, Brazil, [2]Department of Radiology, Federal University of Rio de Janeiro, Rio de Janeiro, Brazil, *corresponding author: tel.: +55 24 22492777, fax: 55(21)26299017, e-mail: edmarchiori@gmail.com

CASE REPORT

A 64-year-old woman presented at our instution with a one-month history of polyarthritis, along with left hearing impairment, blurred vision, progressive dyspnoea, and dry cough. The symptoms had started 18 years previously, when she presented with a sudden episode of pain and swelling in both ears that spared the lobules. At that time, she was diagnosed with infectious perichondritis and treated with antibiotics, without response. She also developed recurrent episodes of intermittent polyarthritis, as well as repeated episodes of pain, swelling, and heat in the nose. Serological findings were negative. The patient also reported an episode of subglottic stenosis five years after the onset of symptoms.

On physical examination, a saddle nose deformity and floppy, deformed ears were noted.

Figure 1. *Floppy, deformed ears were noted*

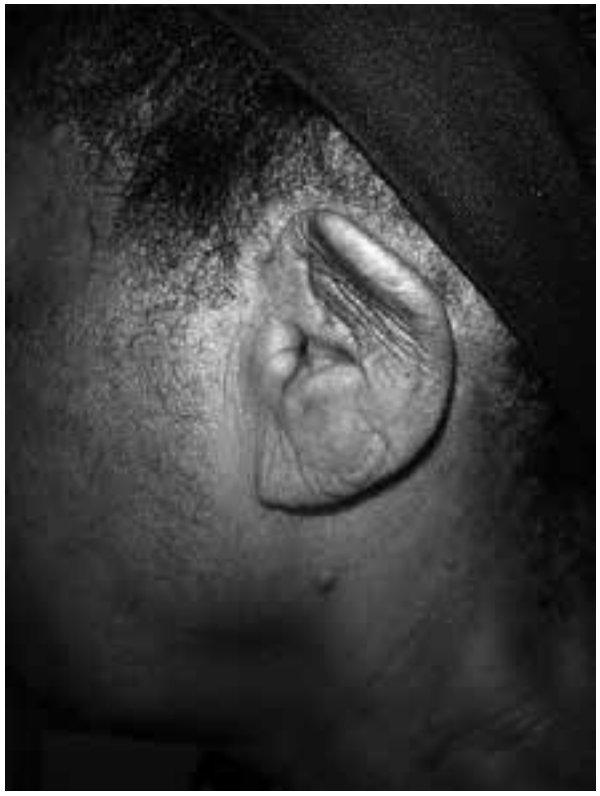
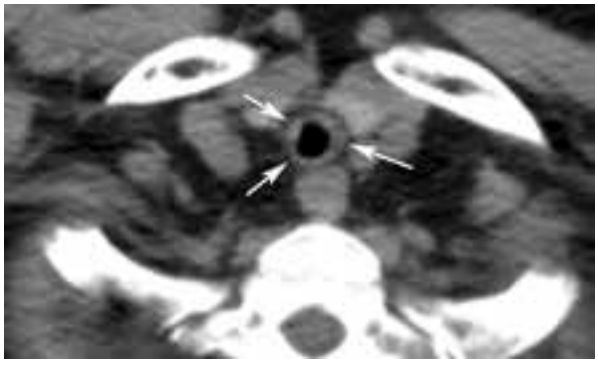


Figure 2. *Chest computed tomography revealed smooth tracheal and main bronchial wall thickening that spared the posterior membranous wall, with narrowing of its lumen. No significant parenchymal findings were observed*



WHAT IS YOUR DIAGNOSIS?

See page 421 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 417)

FLOPPY EARS AND TRACHEAL WALL NARROWING

DIAGNOSIS

The diagnosis of relapsing polychondritis was established on a clinical basis, according to the criteria of nasal, auricular, and laryngotracheal chondritis and seronegative inflammatory arthritis.[1] A biopsy was not necessary for the diagnosis.[2]

Relapsing polychondritis is characterised by recurrent, potentially severe, episodes of inflammation in cartilaginous tissues, including the elastic cartilage of the ears and nose, the hyaline cartilage of peripheral joints, the fibrocartilage at axial sites, and the cartilage in the tracheobronchial tree. It may also affect other proteoglycan-rich structures, such as the eye, heart, blood vessels, and inner ear.[3]

External ear pain is the most common symptom of relapsing polychondritis, and is usually an isolated presenting symptom. Almost invariably, this condition is misdiagnosed as infectious perichondritis of the ear; however, relapsing polychondritis typically spares the lobule, in contrast to infectious processes. With repeated attacks, the ear can become nodular and, in severe cases, floppy and deformed as the cartilaginous support is lost.[3] Nasal chondritis is painful, affects the distal part of the nasal septum, and through recurrent episodes leads to a saddle nose deformity.[2]

Arthritis is the second most common symptom of relapsing polychondritis. Intermittent, migratory, asymmetric, seronegative, and usually nonerosive poly- or oligoarthritis, lasting weeks to months, is the most frequently encountered pattern. Ocular symptoms occur in approximately 60% of patients.[3] Airway involvement is common and sometimes severe in patients with relapsing polychondritis. Airway symptoms include progressive dyspnoea, cough, stridor, hoarseness, and chest discomfort,[4] which can be explained by destruction and fibrosis of the laryngeal and tracheal cartilaginous rings, creating luminal collapse, and also by airway narrowing due to inflammation and cicatricial fibrosis.[3,4] Cardiovascular involvement is the second most frequent cause of death in these patients.[1] Clinicians should be aware of the existence of relapsing polychondritis, as the disease can be managed with medication to reduce the frequency, duration, and severity of flare-ups.

REFERENCES

1. Michet CJ Jr, McKenna CH, Luthra HS, O'Fallon WM. Relapsing polychondritis. Survival and predictive role of early disease manifestations. Ann Intern Med. 1986;104:74-8.
2. Kent P, Michet C, Luthra H. Relapsing polycondritis: Curr Opin Rheum. 2004;16:56-61.
3. Trentan D, Christine H. Relapsing Polycondritis. An Intern Med. 1998;129:114-22.
4. Ernst A, Rafeq S, Boiselle P, et al. Relapsing polychondritis and airway involvement. Chest. 2009;135:1024-30.