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CASE STUDY

Epistaxis and systemic disease

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KEYWORDS
Epistaxis;
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Abstract
We report the case of a 77-year-old man who presented nasal obstruction sensation and epistaxis. Otorhinolaryngologic examination revealed occupation of the left nasal passage and the left maxillary sinus by an inflammatory tumour, the biopsy results of which were inconclusive. While diagnostic tests were being carried out, the patient presented a severe systemic condition consisting mainly of anemia, acute renal failure, and cavitated diffuse bilateral lung infiltrates. In the light of the results of anti-neutrophilic cytoplasmic antibodies and renal biopsy, Wegener's granulomatosis was diagnosed and treatment for the disease was instituted, with a favourable response. Finally, clinical manifestations of Wegener's granulomatosis are reviewed, with special emphasis on otolaryngologic complaints.

PALABRAS CLAVE
Epistaxis;
Enfermedad sistémica;
Granulomatosis de Wegener

Epistaxis y enfermedad sistémica

Resumen
Varón de 77 años que comenzó con un cuadro de sensación de obstrucción nasal y epistaxis. Los exámenes otorrinolaringológicos mostraron una ocupación de la fosa nasal y el seno maxilar izquierdos por una tumoración inflamatoria, cuya biopsia no resultó diagnóstica. Mientras se estudiaban dichos síntomas, el paciente presentó un grave cuadro sistémico, fundamentalmente anemia, insuficiencia renal aguda e infiltrados alveolointersticiales bilaterales pulmonares cavitados. Con base en la positividad de los anticuerpos anticitoplasma de neutrófilo y el resultado de la biopsia renal, se estableció el diagnóstico de granulomatosis de Wegener y se inició su tratamiento, con buena respuesta clínica. Finalmente se comentan las manifestaciones clínicas de la granulomatosis de Wegener y se hace especial hincapié en los síntomas otorrinolaríngeos de la enfermedad.

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Case study

A 77-year-old male with a history of smoking, deep vein thrombosis of the left lower extremity, intermittent claudication, benign prostatic hypertrophy, and operation of the right hip for osteoarthritis, with placement of joint prosthesis, in routine treatment with clopidogrel and dutasteride. The patient initially came to the otolaryngology service due to a feeling of blockage in the nasal passages and bloody mucus issue from them. In this service, computed tomography found an occupation of the left maxillary sinus (Figure 1) and a partial occupation of the left nostril (Figure 2), and rhinoscopy revealed an inflammatory tumour in the left nostril. The biopsy of the latter was reported as consistent with squamous papilloma. In the basic analyses, liver and kidney functions were normal, but a slight normocytic anemia was detected, so the patient was referred to the internal medicine service.

In this service, the patient presented progressive deterioration of his general condition, epistaxis, and mild hemoptysis. Physical examination was normal except for bilateral basal crackles on lung auscultation. A new analysis showed several alterations, among which were: hemoglobin, 10 g/dL with normal average corpuscular volume; erythrocyte sedimentation rate, 120mm in the first hour; creatinine, 11 mg/dL; urea, 276 mg/dL; and neutrophil cytoplasmic antibodies (cytoplasmic pattern), positive upon 1/20 title. Urinalysis showed nephrotic-range proteinuria and microhematuria. The electrocardiogram showed a right branch blockage. The chest radiograph (Figure 3) displayed bilateral interstitial-alveolar infiltrates and chest CT scan (Figure 4) showed the same infiltrates with multiple cavitations present in them. Abdominal ultrasound was normal. Renal biopsy revealed focal segmental glomerulonephritis, without immune deposits.

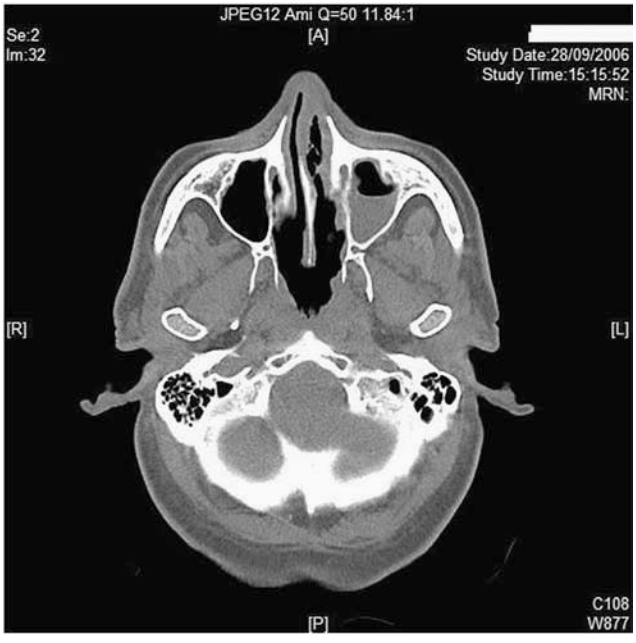

Figure 1 CT scan showing partial occupation of the left maxillary sinus.

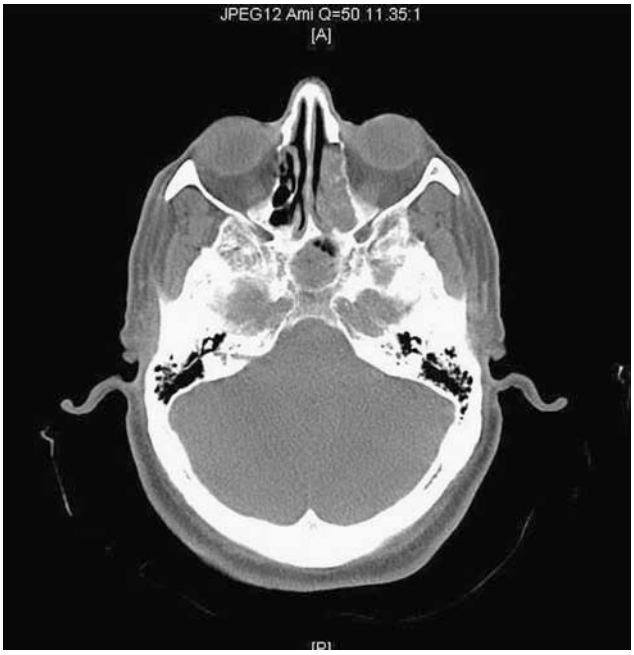

Figure 2 CT scan showing partial occupation of the left nostril.

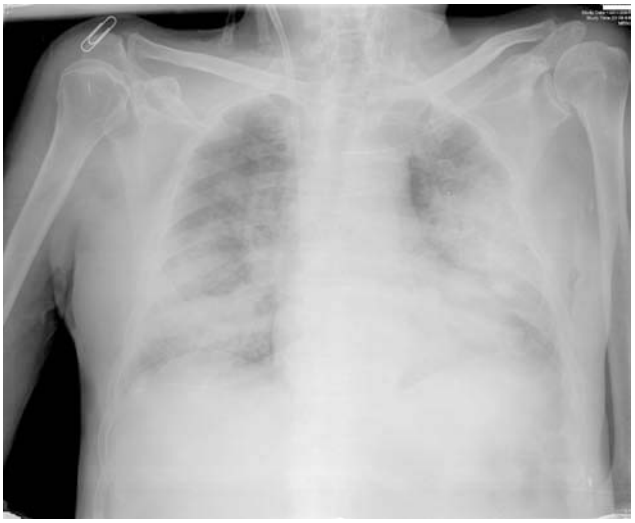

Figure 3 Chest radiograph showing bilateral interstitial-alveolar infiltrates.

Evolution

All symptoms improved initially with hemodialysis and massive doses of glucocorticoids. Then treatment with cyclophosphamide, intermediate doses of glucocorticoids, and trimethoprim-sulfamethoxazole was commenced. The treatment resulted in further improvement, so hemodialysis was not needed any longer. The pulmonary infiltrates virtually disappeared from control chest radiographs.

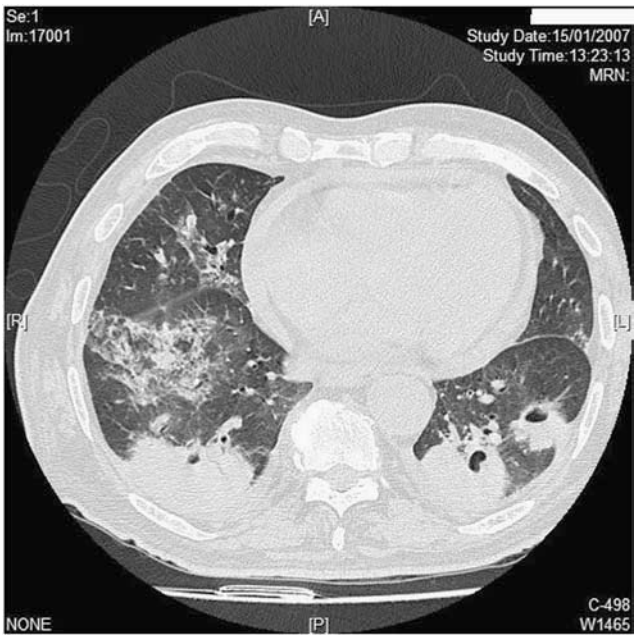

Figure 4 Chest CT scan showing pulmonary interstitial-alveolar infiltrates and multiple cavitations.

Diagnosis

Wegener's granulomatosis.

Discussion

Wegener's granulomatosis is a form of vasculitis that primarily affects the arteries and veins of small size. It is a rare process, with prevalences that vary between populations, but are generally around <°5 cases/ 100 000 inhabitants. It occurs equally in men and women and can occur at any age.[1]

Pathologically, it is characterized by the appearance of necrotizing vasculitis and formation of granulomas, both intravascular and extravascular. Glomerulonephritis typically occurs in the kidneys, without immune deposits. Clinical manifestations preferentially involve the airways, both superior and inferior, and the kidneys.[2] It is not uncommon for it to also affect the eyes, skin, and nervous system, both central and peripheral. The analyses show elevated acute phase reactants and positive anti-neutrophil cytoplasmic antibodies of anti-proteinase 3 type are characteristic, showing a cytoplasmic pattern.[3]

Diagnosis is made through typical anatomical pathology results in the biopsy of affected tissues, preferably in the lung. The presence of anti-proteinase 3 type antibodies to neutrophil cytoplasm is also useful to substantiate the diagnosis. The differential diagnosis must be made with other processes such as vasculitis, Goodpasture's syndrome, relapsing polychondritis, rhinoscleroma, various neoplasms, midline granuloma, lymphomatoid granulomatosis, and other granulomatous diseases. The treatment basically consists of the administration of cyclophosphamide and corticosteroids. The effectiveness of other drugs such as trimethoprim-sulfamethoxazole or rituximab has not yet been clearly established.[4]

The case we present highlights an aggressive form of presentation of Wegener's granulomatosis, with rapid deterioration of renal function while the patient was under study. This emphasizes the importance of diagnosing this disease as early as possible.[5]

Otologic manifestations of Wegener's granulomatosis are varied and are generally the most common form of presentation of the disease. Persistent congestion of the mucosa in the nostrils is common, as is the formation of scabs and sores, which can lead to perforation of the nasal septum; the side walls or the nasal dorsum may also be affected and destroyed, leading to the deformity of "saddle nose." Epistaxis is an omnipresent symptom. Inflammatory and destructive processes often occur in the paranasal sinuses, similar to those of the nostrils, which may eventually affect the bone structures. Inflammation and ulceration may also appear in the laryngeal and tracheal mucosa, occasionally leading to the formation of stenosis and other types of deformities; the subglottis is the region most frequently affected. Finally, various otologic conditions are also possible; among these serous otopathy, chronic otitis media and conductive or sensorineural hearing loss are the most common.[6]

Conflict of interests

The authors declare no conflict of interests.

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