Eur Respir J, 1994, 7, 2252–2254
DOI: 10.1183/09031936.94.07122252
Printed in UK - all rights reserved

European Respiratory Journal
ISSN 0903 - 1936

CASE REPORT

Unusual manifestations of giant cell arteritis: pulmonary nodules, cough, conjunctivitis and otitis with deafness

T. Zenone*, P-J. Souquet*, C. Bohas*, D. Vital Durand**, J-P. Bernard*

Unusual manifestations of giant cell arteritis: pulmonary nodules, cough, conjunctivitis and otitis with deafness. T. Zenone, P-J. Souquet, C. Bohas, D. Vital Durand, J-P. Bernard. ©ERS Journals Ltd 1994.

ABSTRACT: The major manifestations of giant cell arteritis have been well described. Pulmonary manifestations, however, are rare.

We report the case of a 75 year old woman with temporal arteritis, presenting with atypical manifestations, *i.e.* nodular pulmonary lesions, dry cough, rhinitis, conjunctivitis, and otitis with hearing loss.

We conclude that overlapping features of giant cell arteritis and Wegener's granulomatosis occur in some patients.

Eur Respir J., 1994, 7, 2252–2254.

*Service de Pneumologie et ** Service de Médicine Interne, Centre Hospitalier Lyon-Sud, Pierre-Bénite, France.

Correspondence: P.J. Souquet, Service de Pneumologie, Centre Hospitalier Lyon-Sud, 69495 Pierre-Bénite Cedex, France.

Keywords: Conjunctivitis, deafness, giant cell arteritis, pulmonary nodules

Received: January 28 1994
Accepted for publication June 12 1994

Giant cell arteritis (GCA), which predominantly affects elderly patients, does not always present with the classic manifestations of headache, jaw claudication, blindness, fever and polymyalgia rheumatica.

Case report

A 75 year old woman was admitted to the hospital with a one month history of fatigue, fever, dry cough, dyspnoea and chest pain. She also complained of bilateral conjunctivitis, and otitis with hearing loss. She had lost 5 kg over a one month period. Routine laboratory examination results were normal, with the exception of an erythrocyte sedimentation rate (ESR) of 58 $mm \cdot h^{-1}$ (Westergren). X-ray films of the chest (fig. 1) revealed multiple nodules in both lungs. A computed tomography (CT) scan of the thorax (fig. 2) showed parenchymal nodular opacities without cavitation. Pulmonary function tests were normal, including transfer coefficient and blood gases. Bronchoscopy revealed no abnormalities. In bronchoalveolar lavage, cell count was normal (total 43×10^9 $cells \cdot l^{-1}$; with 88% macrophages, 10% lymphocytes and 2% neutrophils). Cytological examination of bronchial aspirate disclosed no malignant tumour cells, and culture was negative for *mycobacterium tuberculosis*. Needle aspiration of a lung lesion revealed no malignant tumour cells. The patient was

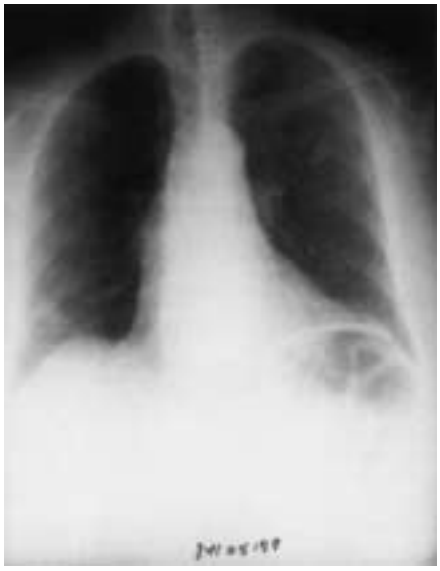

Fig. 1. – Patient's chest radiograph showing multiple pulmonary opacities.

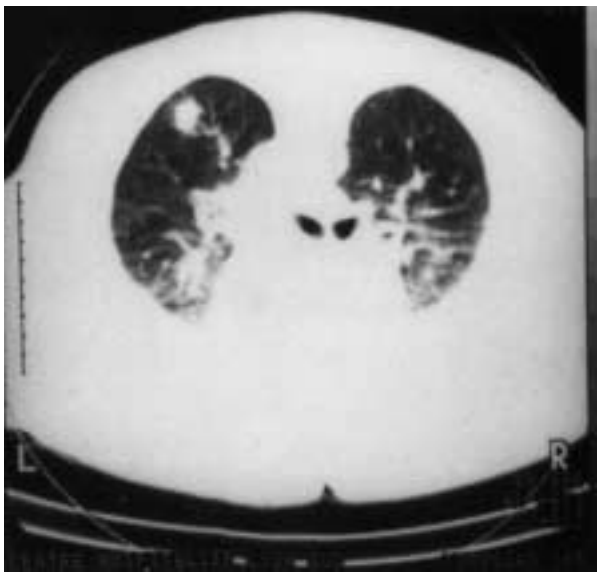

Fig . 2. – Chest computed tomography with irregular opacities in the upper lobes.

discharged from hospital receiving 10 mg·day^{-1} prednisone with no definite diagnosis.

Three weeks later, the patient was readmitted to the hospital because of persistent fatigue, fever, muscle weakness, muscle pain in both arms and legs, and temporal and frontal headaches, but without cough or dyspnoea. She complained of conjunctivitis with eye pain, otitis and moderate clear nasal discharge (rhinitis without sinusitis). Temporal arteries were normal to palpation. The ESR was 68 mm·h^{-1}.

Laboratory data were still normal, including evaluation of renal function. Immune complexes were positive. A test for rheumatoid factor was positive in a titre of 1/40 (latex) and 1/8 (Waaler-Rose). Radiologically, the opacities in the chest were still apparent. A ventilation and perfusion lung scan showed bilateral ventilation and perfusion defects in the region of the upper lobe. A radiograph of the sinuses and CT scan of the brain and sinuses were unremarkable. Examination by an otolaryngologist revealed no nasal mucosal ulceration, a bilateral serous otitis media and a perforation of the right tympanum. Audiometry revealed a bilateral conduction and sensorineural defect. A nasal biopsy showed a chronic inflammation but no necrotizing granulomatous vasculitis in favour of Wegener's granulomatosis (WG). Examination by an ophthalmologist revealed a bilateral conjunctivitis. A conjunctival biopsy was normal. An electromyogram showed a mild neuropathy. A muscle biopsy was normal (no angiitis in favour of polyarteritis nodosa).

Because of the prominent symptoms of headaches and muscle pain, suggestive of polymyalgia rheumatica, a temporal artery biopsy was performed. Intimal fibrosis, inflammatory (lymphocytic and mononuclear) infiltration of the vessel wall, disruption of the internal elastic lamina, and dense perivascular fibrosis with proliferation of capillaries were compatible with GCA). The patient was discharged from the hospital receiving prednisone 60 mg·day^{-1}. The patient's condition improved rapidly.

At the follow-up evaluation, the patient was asymptomatic. The ESR was 5 mm·h^{-1}. The tympanum was normal and her hearing had improved markedly, as was documented on repeated audiograms. Chest roentgenography showed a favourable course. The prednisone dose was slowly tapered over the subsequent 6 months. Upon reduction of the prednisone dose to 27.5 mg·day^{-1}, the patient had recurrence of her previous symptoms with jaw claudication. However, X-ray films of the chest showed marked improvement, with almost complete resolution of lesions. Response to reinstitution of full-dose steroids was rapid.

One year later, the only persistent symptom was a clear nasal discharge without sinusitis. Radiograph and CT scan of the thorax and sinuses were still normal. During the succeeding several months, the patient was maintained on methylprednisolone, at a dose of 8 mg·day^{-1} and felt well. Six months later, she experienced rapid loss of vision in her right eye. Ophthalmoscopy was in favour of a retinal artery occlusion. X-ray films of the chest were still normal. Parenteral methylprednisolone treatment, 120 mg·day^{-1}, was reinstituted. The central scotoma resolved but without amelioration of visual acuity. During the succeeding 8 months, the patient was maintained on methylprednisolone, at a dose of 30 mg·day^{-1}, the ESR ranged between 12–25 mm·h^{-1} and she was symptom free.

Discussion

The clinical manifestations of our patient, the fact that symptoms and ESR responded rapidly to corticosteroid treatment with relapses when the steroid dose was reduced, the histopathological characteristic of the temporal artery, were consistent with the diagnosis of GCA. Our patient met the classification criteria of GCA as proposed by the American College of Rheumatology [1].

GCA is a vasculitis that can involve vessels other than the carotids and their branches. Dry cough is now a well-recognized feature of this disease [2]. However, few cases of GCA associated with lung involvement have been published [3–5]. Lung involvement in patients with GCA is a difficult diagnosis to establish without an open lung biopsy; and caution must be used in the interpretation of this manifestation in our case, since there was no histological proof of the real nature of pulmonary nodules. However, the disappearance of the pulmonary nodules with corticosteroid therapy was an argument against other causes (metastases and infection). Pleural effusions are also rare in GCA, nine reports having been published in the literature, the histological studies of the pleura were nonspecific [6].

Deafness which resolved with steroids has been described in some cases [7]. Our case is somewhat unusual, in that otitis and conjunctivitis in GCA have not previously been described. The prompt alleviation of otitis and conjunctivitis with corticosteroid therapy, the recurrence and resolution as the disease waxed and waned are strong evidence that the GCA was the underlying cause. Exactly how arteritis caused otitis is not clear. Conjunctivitis may be the result of an incomplete anterior segment ischaemia.

These manifestations in the present case report are also suggestive of Wegener's granulomatosis, and the possibility of "limited" Wegener's granulomatosis can not be completely excluded [8]. Necrotizing granulomatous vasculitis is frequently not identified on nasal biopsy, even in overt Wegener's granulomatosis. In our case, antineutrophil cytoplasmic antibodies have not been measured. However, our patient was treated with corticosteroid treatment only; Wegener's granulomatosis often does not respond to prednisone alone and usually requires immunosuppressive agents, specifically cyclophosphamide [8, 9]. Cases of Wegener's granulomatosis associated with clinical signs suggestive of GCA have also been reported in the literature [9–11]. Whether such occurrences signify co-existence of two separate disorders or diverse clinical manifestations of a single underlying vasculitis remains unknown [11]. Other vasculitis may involve the temporal artery, but

clinical features usually allow discrimination [12]. In our case, leucocytoclasis and fibrinoid necrosis were absent on temporal artery biopsy.

Our case illustrates the fact that although certain clinical syndromes have been attributed to specific type of systemic vasculitis, considerable overlap occurs.

References

1. Hunder GG, Bloch DA, Michel BA, *et al*. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. *Arthritis Rheum* 1990: 33: 1121–1128.
2. Larson TS, Hall S, Hepper NGG, Hunder GG. Respiratory tract symptoms as a clue to giant cell arteritis. *Ann Intern Med* 1984; 101: 594–597.
3. Kramer MR, Melzer E, Nesher G, Sonnenblick M. Pulmonary manifestations of temporal arteritis. *Eur J Respir Dis* 1987; 71: 403–433.
4. Karam GH, Fulmer JD. Giant cell arteritis presenting as interstitial lung disease. *Chest* 1982; 82: 781–784.
5. Bradley JD, Pinals RS, Blumenfeld HB, Poston WM. Giant cell arteritis with pulmonary nodules. *Am J Med* 1984; 77: 135–140.
6. Romero S, Vela P, Padilla I, Rosas J, Martin C, Aranda I. Pleural effusion as manifestations of temporal arteritis. *Thorax* 1992; 47: 398–399.
7. Wolfovitz E, Levy Y, Brook JG. Sudden deafness in a patient with temporal arteritis. *J Rheumatol* 1987; 14: 384–385.
8. Leavitt RY, Fauci AS. Pulmonary vasculitis. *Am Rev Respir Dis* 1986; 134: 149–166.
9. Small P, Brisson ML. Wegener's granulomatosis presenting as temporal arteritis. *Arthritis Rheum* 1991; 34: 220–223.
10. Vermeulen JP, Mahowald ML. A case of Wegener's granulomatosis presenting with jaw claudication. *J Rheumatol* 1984; 11: 707–709.
11. Nishino H, De Remee RA, Rubino FA, Parisi JE. Wegener's granulomatosis associated with vasculitis of the temporal artery: report of five cases. *Mayo Clin Proc* 1993; 68: 115–121.
12. Genereau T, Herson S. Les artérites temporales. Eléments du diagnostic différentiel de la maladie de Horton. *Ann Med Interne (Paris)* 1993; 144: 198–211.