Mod Rheumatol (2005) 15:126–129
DOI 10.1007/s10165-004-0375-5

CASE REPORT

Makiho Sekiuchi · Kimimasa Nakabayashi
Tomohumi Marumo · Yoshihiro Arimura
Toshihiko Nagasawa · Akira Yamada · Yasunori Fujioka

Concurrent occurrence of allergic granulomatous angiitis and temporal arteritis: a case report and review of the literature

Received: September 1, 2004 / Accepted: December 3, 2004

Abstract Allergic granulomatous angiitis (AGA) is a disease entity that was first distinguished from classical polyarteritis nodosa by Churg and Strauss in 1951, and is characterized by the clinical features of allergic rhinitis or bronchial asthma, eosinophilia, and vasculitis. Allergic granulomatous angiitis has been described to mainly involve small vessels. We herein describe a case of Churg–Strauss syndrome which demonstrated the clinical and laboratory findings supporting a diagnosis of AGA and was also associated with the clinical and pathological findings for temporal arteritis, which was characterized by eosinophil infiltration and granuloma formation of the temporal artery (middle-sized vessel).

Key words Allergic granulomatous angiitis (AGA) · Churg–Strauss syndrome · Juvenile temporal arteritis with hyper-eosinophilia · Temporal arteritis

Introduction

Vasculitic syndrome is a disease group that demonstrates various clinical features produced by the inflammation and recovery processes of involved blood vessels. Although the pathogenesis of vasculitides is generally considered to involve the immunological processes, the exact mechanism of this syndrome has yet to be clarified. The classification of vasculitis is very difficult and it remains confusing, because some cases may show an overlap of the clinical features of different vasculitides that may have atypical symptoms for each disease. Although various classifications to define vasculitides have so far been used, the classification established at the Chapel Hill conference in 1992 is now widely accepted.[1] Nevertheless, cases continue to be reported that cannot be strictly put into one disease group based on this classification. Since we had a case which demonstrated both AGA[2,3] and temporal arteritis[4] clinically and pathologically, we describe our findings of this case regarding its place among the other similar reported cases.

M. Sekiuchi · K. Nakabayashi (✉) · T. Marumo · Y. Arimura · T. Nagasawa · A. Yamada
First Department of Internal Medicine, Kyorin University School of Medicine, 6-20-2 Shinkawa, Mitaka 181-8611, Japan
Tel. +81-422-47-5511; Fax +81-422-42-9607
e-mail: kimimasa@kyorin-u.ac.jp

Y. Fujioka
Department of Pathology, Kyorin University School of Medicine, Mitaka, Japan

Case report

A 59-year-old woman, who had been followed for bronchial asthma by her family doctor for over 18 years and had been treated by a β-stimulant, theophylline, and small doses of steroid, but not by any antileukotriene drugs, came to our hospital in March 2002 complaining of a transient loss of vision, bilateral headaches, jaw claudication on chewing, facial edema, numbness of the extremities, myalgia, and gait disturbance.

On admission, on May 14, 2002, her body temperature was 36.7°C and her blood pressure was 118/62 mmHg. She had lost 4 kg of weight in the previous month. Her face was slightly edematous and her jaw muscle was hard. She also had nasal congestion, although she had not noticed it. Both superficial temporal arteries were palpable and tortuous, but no murmurs were audible and they were swollen with tenderness. The pulses of the femoral, popliteal, dorsalis pedis, and posterior tibial arteries were also satisfactorily palpable. Wheezing was audible in her chest. She felt numbness in her right thumb and on the ulnar side of both forearms, and she lost muscle strength to a level of MMT3/5 in her left forearm to the fingertips and in both lower extremities. Moreover, the tendon reflexes of her left biceps, bilateral triceps, and right Achilles had also decreased. These signs indicated mononeuritis multiplex.

The results of abnormal laboratory tests were as follows: platelet count 414000/mm^3, white blood count 20000/mm^3 with 63% eosinophils (12600/mm^3), Westergren erythro-

cyte sedimentation rate (ESR) 35 mm/1 h, C-reactive protein 1.3 mg/dl, IgE 855 IU/ml, myeloperoxidase-antineutrophil cytoplasmic antibody (MPO-ANCA) 115 EU, rheumatoid factor 67 IU/ml, rheumatoid arthritis hemagglutinin test 1:160, eosinophil cationic protein 209 μg/l (normal <14.7), interleukin (IL)-4 16.4 pg/ml (normal <6.0), IL-6 13.3 pg/ml (normal <4.0), and transforming growth factor β1 64.8 ng/ml (normal 1.56–3.24). The antinuclear antibody, proteinase-3-ANCA, serum electrolytes, blood urea nitrogen, creatinine, liver function, and urinalysis findings were normal. Chest computed tomography showed a small nodule in segment 3 in the right lung. No thickness, narrowing, or dilation of the aorta was seen which thus suggested inflammatory processes in the vessel wall by magnetic resonance arteriography (MRA) and other diagnostic imaging modalities. The neck ultrasound test suggested only the sclerotic process at the bifurcation of the right cervical artery. Computed tomography, magnetic resonance imaging, and MRA for the brain and sinuses revealed lacunar infarctions and chronic sinusitis. A nerve conduction study in the motor and the sensory nerve of the right forearm was delayed and suggested axonal degeneration.

To make a histological diagnosis, a biopsy of the left temporal artery was performed (Fig. 1). The biopsy specimen revealed lumen narrowing, an interrupted internal elastic lamina, granuloma formation and fibrinoid necrosis in the tunica media, and prominent eosinophil infiltrate in the entire tunicae. The patient was diagnosed to have allergic granulomatous angiitis associated with temporal arteritis, which will be described in more detail in the discussion.

Our patient thus began to receive treatment with 40 mg/day oral prednisolone. After the treatment, her condition improved quickly except for the numbness in both forearms; no loss of vision recurred, and a nodule in the right lung disappeared completely. Although she experienced bleeding from the small intestine 5 weeks after the treatment and required a blood transfusion, her symptoms improved after the administration of oral prednisolone and fasting therapy. The further hospital course was uneventful and she was discharged about 10 weeks later.

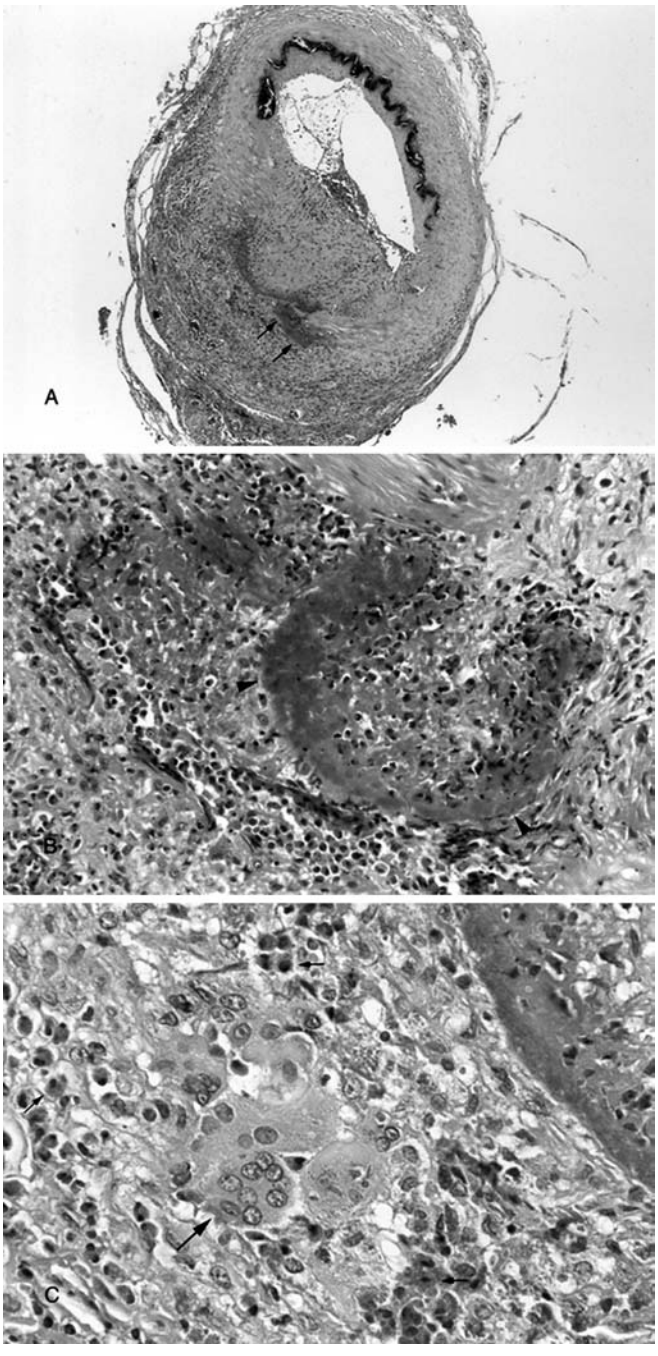

Fig. 1. **A** Temporal artery with an interrupted internal elastica lamina and dense infiltrates of inflammatory cells are observed in the tunicae, and some necrotic lesions between the tunica interna and media are also seen (*arrows*) (hematoxylin–eosin [H&E] stain, ×40). **B** Necrotic lesion between the tunica interna and media (*arrowheads*) and a marked infiltration of eosinophils (H&E, ×200). **C** Granuloma formation with giant cells (*large arrow*) and eosinophils in the tunica media (*small arrow*) (H&E, ×400)

Discussion

This case meets the criteria of the American College of Rheumatology (ARA) 1990 for AGA,[3] because of the existence of peripheral eosinophilia, bronchial asthma, a lung shadow, pathognomonic arteritis (tissue eosinophil infiltration), and mononeuritis multiplex. Furthermore, this case also meets the criteria for temporal arteritis,[4] including an onset at older than 50 years of age, temporal headache and tenderness on both temporal arteries of recent occurrence, elevated ESR, and pathologic findings with granuloma formation in the temporal artery. Therefore, this case can be considered to be a concurrent occurrence of AGA and temporal arteritis (TA).[5–7] However, these two diseases could also be explained by either AGA with temporal artery involvement[8,9] or TA associated with the tissue eosinophil infiltration (so-called juvenile TA with hypereosinophilia).[10–12]

To date, we have found nine cases that have been reported to demonstrate a concurrent occurrence of AGA

Table 1. Clinical manifestations in patients with concurrent occurrence of allergic granulomatous angiitis and temporal arteritis in the literature

First author[Ref.]	Bronchial asthma	Blood eosinophilia	Mononeuritis multiplex	MPO-ANCA
Conn[8]	(+)	(+)	(+)	N.T.
Adoue[14]	(+)	(+)	(+)	N.T.
Bollinger[11]	(−)	(+)	(−)	N.T.
Amato[5]	(+)	(+)	(+)	N.T.
Vidal[6]	(+)	(+)	(−)	N.T.
Lie[13]	(+)	(+)	(−)	N.T.
Serrano-Castro[9]	(+)	(+)	(−)	N.T.
Guardiola[7]	(+)	(+)	(+)	(+)
Endo[15]	(+)[a]	(+)	(+)	(−)[b]
Present case	(+)	(+)	(+)	(+)

N.T., not tested; MPO-ANCA, myeloperoxidase-antineutrophil cytoplasmic antibody
[a] Later onset than vasculitic symptoms
[b] Datum obtained at poststeroid treatment

Table 2. Published cases with the clinical features of "temporal arteritis" and "allergic granulomatous angiitis," and their pathological findings in the temporal arteries

First author[Ref.]	Age (years)	Sex	Pathological findings of temporal artery		
			Eosinophil infiltrate	Granuloma	Necrotic lesion
Conn[8]	49	F	(+)	(−)	(+)
Adoue[14]	?	?	(+)	(+)	(−)
Bollinger[11]	23	M	(±)	(+)	(+)
Amato[5]	26	M	(+)	(+)	(+)
Vidal[6]	41	M	(+)	(−)	(−)
Lie[13]	48	M	(+)	(−)	(−)
Serrano-Castro[9]	74	M	(+)	(−)	?
Guardiola[7]	77	F	(−)	(+)	(−)
Endo[15]	27	M	(+)	(−)	(−)
Present case	59	F	(+)	(+)	(+)

and TA (Tables 1 and 2).[5–9,11,13–15] Clinically, all cases had eosinophilia in the peripheral blood and eight cases except for that of Bollinger et al.[11] had a history of bronchial asthma (Table 1). Therefore, eight cases except for that of Bollinger et al. had sufficient clinical manifestations for a diagnosis of AGA to be made. However, Bollinger's patient did not show any other symptoms such as mononeuritis multiplex for AGA. Pathologically, eight cases except for that of Guardiola et al.[7] had an eosinophilic infiltration of the temporal artery at the time of biopsy (Table 2). Four of these patients had granuloma in their biopsy specimens,[5,7,11,14] but only three had necrosis in the tunica.[5,8,11] Only two out of these three cases showed both granuloma and necrosis in the biopsy specimens, and they were characterized by a relatively young onset age for TA (23 and 26 years old, repectively[5,11]). However, the ages of these patients ruled out a diagnosis of TA based on the ARA diagnostic criteria.[4] Accordingly, we think that these two cases most likely had AGA with temporal artery involvement[5,8] or juvenile TA with hypereosinophilia,[11–13] rather than a concurrent occurrence of AGA and TA[5–7] as mentioned above. However, cases with juvenile temporal arteritis as described in the literature did not demonstrate granuloma formation excepting for that of Bollinger et al.[11] As a result, it is preferable to consider these two cases to have AGA with temporal artery involvement.[5,11]

Our patient had the typical symptoms of AGA and TA simultaneously. A biopsy specimen of the temporal artery revealed a remarkable amount of eosinophil infiltrate, granuloma formation, and fibrinoid necrosis. Therefore, the present case was also characterized by the coexistence of granuloma and necrosis in the involved temporal artery. Taken together, this patient was suggested to have AGA involving the temporal artery in a manner similar to the above-mentioned cases of Amato et al.[5] and Bollinger et al.[11] However, Bollinger's patient had neither a history of bronchial asthma nor allergic sinusitis, which are important symptoms for the diagnosis of AGA. Therefore, our case is the second one that showed both clinical and laboratory findings supporting a diagnosis of AGA and an association with the temporal artery with granuloma formation and necrosis similar to the findings in Amato's patient. These findings therefore confirmed the diagnosis of AGA with temporal artery involvement. The pathological changes in AGA rarely permeate into a comparatively middle-sized vessel such as the temporal artery, and accordingly we

should always keep the existence of some exceptional cases for the disease classification in mind in order to diagnose such cases accurately.

Acknowledgment This work was supported in part by the Research Committee for Intractable Vasculitic Syndrome of the Ministry of Health, Welfare, and Labor, Japan.

References

1. Jennette JC, Falk RJ, Andrassy K, Bacon PA, Churg J, Gross WL, et al. Nomenclature of systemic vasculitides. Proposal of an international consensus conference. Arthritis Rheum 1994;37:187–92.
2. Churg J, Strauss L. Allergic granulomatosis, allergic angiitis and periarteritis nodosa. Am J Pathol 1951;27:277–301.
3. Masi AT, Hunder GG, Lie JJ, Michel BA, Bloch DA, Arend WP, et al. The American College of Rheumatology 1990 criteria for the classification of Churg–Strauss syndrome (allergic granulomatosis and angiitis). Arthritis Rheum 1990;33:1094–100.
4. Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, Calabrese LH, et al. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. Arthritis Rheum 1990;33:1122–8.
5. Amato MBP, Barbas CS, Delmonte VC, Carvalho CR. Concurrent Churg–Strauss syndrome and temporal arteritis in a young patient with pulmonary nodules. Am Rev Respir Dis 1989;139:1539–42.
6. Vidal E, Lioson F, Rogues AM, Cransac M, Berdha J, Liozon E. Concurrent temporal arteritis and Churg–Strauss syndrome. J Rheumatol 1992;19:1312–4.
7. Guardiola JM, Rodriguez-Pla A, Lopez-Contreras J, Llobot JM, Domingo P, Franco M, et al. Simultaneous presence of C-ANCA and P-ANCA in a patient with concurrent Churg–Strauss syndrome and giant cell temporal arteritis. Scand J Rheumatol 2000;29:68–9.
8. Conn DL, Dickson ER, Carpenter HA. The association of Churg–Strauss vasculitis with temporal artery involvement, primary biliary cirrhosis and polychondritis in a single patient. J Rheumatol 1982;9:744–8.
9. Serrano-Castro PJ, Hernandez Ramos FJ, Gomiz Camello A, Minguez Castellanos A, Garcia Gomez T. Non-giant cell temporal arteritis as a manifestation of Churg–Strauss syndrome. Presentation of a case and review of literature. Neurologica 1996;10:82–4.
10. Lie JT, Gordon LP, Titus JL. Juvenile temporal arteritis, biopsy study of four cases. JAMA 1975;234:496–9.
11. Bollinger A, Leu HJ, Brunner U. Juvenile arteritis of extracranial arteries with hypereosinophilia. Klin Wochenschr 1986;64:526–9.
12. Tanaka U, Arimura Y, Nagasawa T. A case of juvenile temporal arteritis with tissue eosinophilia (in Japanese with English abstract). Ryumachi-ka (Rheumatology) 1995;14:77–81.
13. Lie JT, Nagpal S. Churg–Strauss syndrome with nongiant cell eosinophilic temporal arteritis. J Rheumatol 1994;21:366–7.
14. Adoue D, Dongay G, Rego J, Couret B, Ariet-Suau E, Fedou R. Association maladie, De Horton, periarterite noueuse? Rev Med Toulouse 1983;1:167–9.
15. Endo T, Katsuta Y, Kimura Y, Kikuchi A, Aramaki T, Takano T, et al. A variant form of Churg–Strauss syndrome: initial temporal non-giant cell arteritis followed by asthma – is this a distinct clinicopathologic entity? Hum Pathol 2000;31:1169–71.