Giant cell arteritis as a cause of death

R. Andersson

Department of Infectious Diseases, Sahlgren University Hospital/Östra, Göteborg, Sweden

Please address correspondence and reprint requests to: Rune Andersson M.D, Ph.D, Department of Infectious Diseases, Sahlgren University Hospital/Östra, SE-41685 Göteborg, Sweden. E-mail: rune.andersson@medfak.gu.se

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ABSTRACT

It is not well known that giant cell arteritis can cause fatal complications due to rupture of aortic aneurysms or cerebral and myocardial infarctions. If corticosteroid treatment is started early, most of these complications can be avoided.

Introduction

Giant cell arteritis (GCA) can threaten the life of its victims due to inflammatory lesions in vital vessels such as the coronary arteries, the aorta and the cerebral arteries. However, only a few fatal cases are reported in the literature and most clinicians are not aware that GCA can cause cerebral and myocardial infarctions as well as aortic aneurysms. Fatal GCA cases are probably even more often undetected today due to the decreasing frequency of autopsies and the fact that microscopic examination of the arteries is not routinely performed.

Myocardial infarction

The prevalence of GCA in cardiological practice is unknown and How *et al*. described the problem in 1980 as "a cardiological blind spot" (1). Only a few case reports on myocardial infarction due to GCA are available in the literature (2-5).

Dissecting aneurysm of the aorta

Aortitis in GCA was first reported by Sproul and Hawthorn in 1937 (6). Östberg studied 20591 autopsies in Malmö, Sweden 1957-71 and found that out of 443 aortic aneurysms, 85% were due to atherosclerosis, 8% to syphilis, and 7% to GCA (7).

Evans calculated that the risk for GCA patients of suffering thoracic aortic aneurysms was 17.3 times (95% confidence interval (CI) 7.9 - 33) the risk in the general population (8). The corresponding risk for addominal aortic aneurysms was 2.4 (95% CI 0.8 - 5.5).

Liu reviewed 24 cases of aortic dissection in GCA and noted that 11/24 patients (46%) had no previous GCA diagnosis (9). 80% of the patients died within 2 weeks.

In Gothenburg we found aortic aneurysms in 6/90 patients after a median follow-up period of 11.3 years (10). Rupture of the aneurysm was the cause of death in 2 patients.

Cerebral infarction

Gilmour reported arteritis of the internal carotid artery in 1941. Arteritis of the chiasmatic and pituitary arteries were observed in 1959 by Crompton. In 1968 Kjeldsen and Reske-Nilsen found arteritis in the circle of Willis. Wilkinson and Russel reviewed 8 cases in the literature and presented 4 of their own in 1972 (11). They observed an upper border of the arteritis 5 mm above the dural perforation. From Göteborg we reported seven GCA patients with cerebral stroke (5). In 6/7 patients the arteritis even involved the intracerebral arteries. Only one patient had the picture of arteritis outside the dura reported by Wilkinson and Russel.

Caselli noted transient ischemic attacks (TIA) or cerebral stroke in 12/166 (7%) of consecutive patients with biopsy-proven GCA (12). In 1990 Caselli reported 3 cases of multi-infarct dementia due to GCA.

Discussion

The risk of fatal complications stress the importance of suspecting and diagnosing GCA early. The preventive effect of corticosteroid treatment is indicated by many long-term follow-up studies of GCA patients reporting no increase in the overall mortality rate compared to the general population (8, 10, 13).

In an analysis of patients with biopsy-proven GCA, Nordborg & Bengtsson (14) reported an increased mortality during the first 4 months after diagnosis, but not later on. Schaufelberger noted an increased mortality during the first 2 years in patients with polymyalgia rheumatica with no histological signs of arteritis on biopsy of the temporal artery (15). In the latter two studies the patients were treat-

ed by many different doctors without special knowledge of GCA in contrast to the first long-term follow-up study in Gothenburg in which only doctors with a particular interest in the disease were involved.

This may indicate the promising possibility of preventing many fatal complications by a careful monitoring of the patients. It will also be of great interest to study the suggested preventive effect of low dose acetylsalisylic acid or other anti-coagulants on the incidence of vascular complications. To evaluate this problem a multi-center clinical trial is highly recommended.

References

1. HOW J, STRACHAN RW, BEWSHER PD: Giant cell arteritis of coronary arteries: A cardiological blind spot ? *Am Heart J* 1980; 100: 405-7.
2. MORRISON AN, ABITBOL M: Granulomatous arteritis with myocardial infarction: A case report with autopsy findings. *Ann Intern Med* 1955; 42: 691-700.
3. MARTIN JF, KITTAS C, TRIGER DR: Giant cell arteritis of coronary arteries causing myocardial infarction. *Br Heart J* 1980; 43: 487-9.
4. LIE JT, FAILONI DD, DAVIS JR DC: Temporal arteritis with giant cell aortitis, coronary arteritis and myocardial infarction. *Arch Pathol Lab Med* 1986; 110: 857-60.
5. SÄVE-SÖDERBERGH J, MALMVALL BE, ANDERSSON R, BENGTSSON BÅ: Giant cell arteritis as a cause of death- Report of nine cases. *JAMA* 1986; 255: 493-6.
6. SPROUL EE, HAWTHORNE JJ: Chronic diffuse mesaortitis. Report of two cases of unusual type. *Am J Path* 1937; 13: 311-23.
7. ÖSTBERG G: On arteritis with special reference to polymyalgia arteritica. *Acta Pathol Microbiol Scand* Sect A 1973; Suppl. 237: 1-59.
8. EVANS ME, O'FALLON M, HUNDER GG: Increased incidence of aortic aneurysm and dissection in giant cell (temporal) arteritis. A population-based study. *Ann Intern Med* 1995; 122: 502-7.
9. LIU G, SHUPAK R, CHIU KY: Aortic dissection in giant cell arteritis. *Semin Arthritis Rheum* 1995: 25: 160-71.
10. ANDERSSON R, MALMVALL BE, BENGTSSON BÅ: Long-term survival in giant cell arteritis including temporal arteritis and polymyalgia rheumatica. A follow-up study of 90 patients treated with corticosteroids. *Acta Med Scand* 1986; 220: 361-4.
11. WILKINSON IMS, RUSSEL RWR: Arteritis of the head and neck in giant cell arteritis. A pathological study to show the pattern of arterial involvement. *Arch Neurol* 1972; 27: 378-91.
12. CASELLI RJ, HUNDER GG, WHISNANT JP: Neurologic disease in biopsy-proven giant cell (temporal) arteritis. *Neurology* 1988; 38: 352-9.
13. MATTESON EL, GOLD KN, BLOCH DA, HUNDER GG: Long-term survival of patients with giant cell arteritis in the American College of Rheumatology giant cell arteritis classification criteria cohort. *Am J Med* 1996; 100: 193-6.
14. NORDBORG E, BENGTSSON BÅ: Death rates and causes of death in 284 consecutive patients with giant cell arteritis confirmed by biopsy. *Br Med J* 1989; 299: 549-50.
15. SCHAUFELBERGER C, BENGTSSON BÅ, ANDERSSON R: Epidemiology and mortality in 220 patients with polymyalgia rheumatica. *Br J Rheumatol* 1995; 34: 261-4.