Mortality in giant cell arteritis: analysis of a monocentric cohort of biopsy-proven patients

Sirs.

Giant cell arteritis (GCA) is a systemic vasculitis of the elderly that is characterised by granulomatous involvement of large and medium-sized vessels with a predilection for the extracranial branches of the carotid arteries (1, 2). The wide spectrum of clinical manifestations can extensively vary, from cranial symptoms, such as headache, jaw claudication or visual alterations, to constitutional symptoms, like fever, weight loss or asthenia. GCA may cause death by thromboembolic events, rupture of aortic aneurysm, involvement of coronary or cerebral arteries (3). Many studies have attempted to address whether GCA is associated with an increase in mortality rate, reporting conflicting results (4-13); as a matter of fact, some reports have suggested that GCA does not seem to have an effect on mortality, while others showed that GCA patients are more likely than age- and gender-matched controls to die within the first years after diagnosis.

In the present study we analysed the causes of death in a monocentric cohort of biopsy proven GCA patients. We carried out an analysis by reviewing all the medical documentation and by means of a telephone interview to all patients with a diagnosis of GCA consecutively seen at our centre from 1990 to 2010. Patients who had not undergone a temporal artery biopsy (TAB), who had a negative TAB or with insufficient clinical data were excluded. The collected data also included clinical GCA findings, and steroid and immunosuppressant use. GCA-related causes of death were defined as having reference to a cause compatible to GCA and present during a relapse of disease, while gastrointestinal haemorrhage and severe infections were considered related to its treatment. Out of a cohort of more than 220 GCA patients, 112 biopsy-proven patients (18 males and 94 females, mean ± SD age: 79±3 years; mean ± SD at the onset 72±7 years, mean disease duration 9 years) were studied. All the patients had received high-medium doses of glucocorticoids (GC), followed by a rapid tapering scheme to a low GC oral regimen; moreover, 45% of patients received GC alone (mean GC duration 20±4 months), while the others received GC plus MTX (mean GC duration 11±4 months). At the time of this analysis, 15 of the 112 patients (13%) died, of whom only one had a systemic involvement of disease. At the time of death none of the patients was taking immunosuppressive therapies, 7 of them were receiving daily low doses of GC, and a patient with systemic involvement was receiving high-doses of GC. An analysis of causes of death revealed that all but one subject had died as a result of diseases unrelated to GCA: cancer: 4 (colon cancer 2, lung cancer 1, pancreatic cancer: 1), cardiovascular disease: 7 (existing before GCA onset); intestinal obstruction: 1; senescence: 2; the mean latency period between GCA onset and death was 6±4 years. The only case of death related to GCA was a female patient with a systemic involvement of disease. The fluorodeoxyglucose positron emission tomography with computerised tomography (18)FDG PET-CT performed in this case showed a significant uptake in a circumferential fashion along the aorta and its major branches, including the carotid, subclavian, and common iliac arteries. In spite of prompt treatment with a high-dose of steroids, a high level of disease activity persisted and the patient died of a pulmonary embolic event 3 months after onset. Comparing the subjects deceased with the other patients of the cohort, no differences were observed in terms of sex, age at onset, disease duration, clinical GCA profile, GC and immunosuppressant use.

The literature data show that mortality rates of GCA are comparable to those of the general population (14), however, the debate on the effect of GCA on mortality is still open. Given the age restriction of GCA, it has been suggested that immune-senescence plays a role in the disease pathogenesis and it has also been proposed as a possible mechanism by which an increase in mortality was found in patients with GCA. Many studies showed that the risk conferred by the disease appears to decrease with the passage of time, while it can remain significantly elevated in patients with an insufficiently controlled disease. Since among the patients studied the only patient who died of a cause directly related to GCA was characterised by a high level of disease activity, our results may suggest that the maintenance of disease remission could represent an optimal prevention measure for reducing mortality in GCA.

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References