Giant cell arteritis (GCA) is a systemic immune-mediated granulomatous vasculitis of large- and medium-sized arteries mainly affecting elderly people. Death from GCA alone is rare and usually results of ruptured aorta. In this paper is reported a case of a 83-year-old woman who unexpectedly died during treatment of GCA. Necropsy revealed inflammatory involvement of the coronary arteries with left descendent anterior artery thrombosis, myocardial infarct and rupture of the anterior wall of the left ventricle, as well as hemopericardium and cardiac tamponade. Myocardial infarction leading to sudden death is an exceptional complication of GCA.

Case Report

Patient, 83 yrs, female, was admitted at Hospital das Clínicas/UFMG due to accentuated lower-limb pain, especially at the thigh extension, in the previous seven months. She had presented ocular pain and progressive loss of sight in both eyes, within an interval of a few days. She also complained of headaches, adinamy, weight loss and later, pain at mandible movement and appearance of scalp nodules.

Physical examination showed subcutaneous mobile and painless scalp nodules of up to 2 cm of diameter. BP was 140 X 90 mmHg and HR was 72 bpm. The patient was alert and oriented. Visual acuity assessment showed the absence of luminous perception in the right eye, where there was total cataract, and luminous perception in the left eye. Left eye fundoscopy showed accentuated optic atrophy. The patient presented difficulty to open the mouth, with pain at the masseter region.

Complementary exams showed hemoglobin of 11.4 g/dL, RBC of 4,500,000/mm³, MCV of 79.4 fl, MCH of 26.1 pg, ESR of 110 mm (1 hour), leukocytes: 21,900/mm³, platelets: 873,000/mm³, glycemia: 112 mg/dL, SGOT: 63 U/L, SGPT: 29 U/L, protein electrophoresis: albumin – 3.44 g%, α1- 0.27g%, α2- 1.74g%, β1- 0.98g%, β2-0.80g%, γ-1.87g%; positive C-reactive protein: 12mg/dl.

The patient was treated with 250 mg intravenous methylprednisolone every 6 hours, analgesics and potassium chloride. Her evolution was free of significant modifications with up to 80% of lumen obstruction. A previous thrombosis was observed in one of the vertebral arteries, obstructing 90% of the lumen. Very recent ischemic necrosis of the rare optic nerve fascicle. Multiple focal necrosis in organization in the cerebellar cortex and more discrete in the left frontal lobe and base of the bridge. Old selective focal and microscopic neuronal necrosis. Acute myocardial transmural...
infarction, measuring approximately 3.0 x 3.0 cm with rupture of the anterior wall of the left ventricle; consequent hemopericardium and heart tamponade (800 ml of blood). Recent and obstructive thrombosis of the anterior descending artery, a branch of the left coronary artery (Figures 2-A, B).

Moderate/intense edema of the encephalon (weight: 1,200g); uncal, posterior orbital region and cerebellar tonsil herniation. Encephalon hypotrophy and mild to moderate evidence of neurohistological alterations caused by aging.

Mild to moderate generalized atherosclerosis (thoracic and abdominal aorta, common carotids, encephalon base vessels and coronaries), with fibrous and calcified plaques.

Bronchioloalveolar multifocal carcinoma (peripheral nodules of approximately 1.5 to 2.9 cm of diameter), in a cicatricial fibrosis area. Generalized chronic adult emphysema.

Discussion

The American College of Rheumatology has defined five clinical criteria for the diagnosis of GCA: 1) age ≥ 50 yrs; 2) presence of a new headache; 3) tenderness or decreased pulse of the temporal artery; 4) hemosedimentation velocity (HSV) > 50 mm/hr and 5) positive temporal artery biopsy. In addition to all these positive data, the patient presented a history of adynamy, poor appetite and weight loss, fever, subcutaneous scalp nodules, mandibular claudication and bilateral loss of sight, demonstrating the broad clinical spectrum of GCA. The mandibular claudication is due to the maxillary artery arteritis, with ischemia of the mastication muscles, being a symptom with low sensitivity, but a specificity of 97.5%.

The ocular involvement in GCA is reported in 14 to 70% of the cases. The most common cause of loss of sight in GCA is the arteritic anterior ischemic optic neuropathy, due to occlusion of one or more posterior ciliary arteries and consequent optic nerve infarction. The sequential involvement of both nerves is common. Amaurosis fugax is a frequent symptom, which precedes the irreversible loss of sight. This also can arise due to the obstruction of the central artery of the retina and the cilioretinal artery. Diplopia can occur, occasionally, due to ischemia of the extraocular muscles.

The ocular alterations are considered permanent; sometimes it is possible to obtain the stabilization of the process and more rarely, slight improvement with treatment. Nevertheless, the anterior ischemic optic neuropathy and loss of sight have been verified during the use of high-dose steroids, in spite of the disappearance of the systemic manifestations.

In the case reported here, the diagnosis was corroborated by the high levels of HSV (> 50 mm/hour) and positive C-reactive protein, which have a sensitivity > 97%. There was also thrombocytosis, leukocytosis, microcytic and normochromic anemia, increase of transaminases and plasma protein electrophoresis compatible with the diagnosis of GCA and observed in cases subject to complications.

The biopsy of the temporal artery is the gold standard for the diagnosis. The anatomopathological evaluation through the necropsy allowed the confirmation of the diagnosis, but especially, the distribution of the GCA in the several arterial territories. The classically described lesions are observed, represented by fibrointimal hyperplasia, mononuclear cell inflammatory infiltrate between the intima and the media, fragmentation of the internal elastic limitant and the presence of giant cells (Figures 1 and 2B, C, D); degeneration of the muscular layer and lymphohistiocytary infiltrate in the adventitia. These typical morphological aspects can present under other patterns, including without an active process, resulting in false-negative biopsies in 5-13% of the cases. The intima thickening, if present, can be related to atherosclerosis, common in the elderly. The differential diagnosis includes Takayasu arteritis, characterized as granulomatous vasculitis accompanied by accentuated cicatricial fibrosis throughout the vessel, which affects the aorta and its branches and can affect the coronaries; it is predominant, however, in women younger than 50 yrs, constituting clinical syndromes that are dissimilar from GCA.

The atherosclerosis, which frequently coexists with GCA, has a distinct topography that favors the anterior component
of the polygon of Willis and the carotid system. On the other hand, GCA affects more the posterior circulation, has a focal and segmental distribution in the vertebral arteries, rarely close to the polygon of Willis and, in general, do not exhibit lesions after perforating the dura mater, due to the loss of the elastic fibers of the median layer and the external elastic lamina, important factors in the physiopathogeny of the disease^{13}.

In the reported case, the process extended beyond the vertebral arteries, reaching the basilar artery (Figure 1-B), with the observation of old and recent cerebellar necrosis corresponding to the cerebellar arterial vessels. However, the patient did not present clinical and neurological deficits related to such alterations.

The clinical course of GCA is usually auto-limited and non-fatal, can last for months or years and can be followed by complete remission. The prolonged use of corticosteroids and immunosuppressive drugs is indicated with the aim of preventing the reactivation of the inflammatory process. Residual sequelae include amaurosis, mental and neurological disorders^{9}. GCA in general, does not influence survival and the causes of death do not differ from the general population in the same age range. GCA-related deaths occur more frequently due to aorta rupture by aneurysm, stroke, myocardial infarction and arterial thrombosis of the lower limbs^{6,14}.

The coronary involvement is rare and acute myocardial infarction (AMI) as a cause of death directly related to GCA is exceptional^{6,15,16}, although there is evidence of its more significant presence in the coronary arteries and its participation in the genesis of non-fatal infarctions^{17}. The anatomopathological confirmation of myocardial infarction in GCA is rarer, and its real prevalence is yet to be determined.

It is noteworthy that the patient presented AMI during the course of high-dose corticosteroid therapy. Complications are described even during the course of treatment^{6,10,15}.

The present case emphasizes the importance of the necropsy in the accurate diagnosis elucidation, particularly when there is sudden death. Although the diagnosis of GCA had been established by clinical criteria, the immediate cause of death and its relation with GCA would remain undetermined if the necropsy had not been performed. It is important to mention the presence of bronchioloalveolar carcinoma as an exclusive finding at the post-mortem examination.
References


