Family Medicine & Primary Care Review 2012, 14, 4: 628–630

PL ISSN 1734-3402

PRACE KAZUISTYCZNE • CASE REPORTS

Bilateral giant cell arteritis

Obustronne zapalenie tętnicy skroniowej

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A – Study Design, **B** – Data Collection, **C** – Statistical Analysis, **D** – Data Interpretation, **E** – Manuscript Preparation, **F** – Literature Search, **G** – Funds Collection

Summary Giant cell arteritis is an autoimmune necrotizing arterial vasculitis affecting mostly people over the age of 50, more frequently women. Symptoms vary depending on which artery is affected. Patients may present with fatigue, weight loss, headache localized in the temporal region, polymyalgia rheumatica and visual disturbances. In the following article we present the case of a woman with bilateral giant cell arteritis affecting temporal arteries. The necessity of making a prompt diagnosis using the criteria of the American College of Rheumatology and of starting therapy with glucocorticoids in order to prevent serious complications was emphasized.
Key words: giant cell arteritis, early recognition and treatment, ophthalmic features.

Streszczenie Olbrzymiokomórkowe zapalenie tętnic jest autoimmunologicznym martwiczym zapaleniem naczyń tętniczych występującym głównie u osób po 50. roku życia, częściej u kobiet. Objawami choroby, zróżnicowanymi zależnie od zajętego naczynia, są najczęściej: osłabienie, utrata masy ciała, ból głowy okolicy skroniowej, polimialgia reumatyczna i epizody zaniewidzenia. Opisany został przypadek kobiety z obustronnym olbrzymiokomórkowym zapaleniem tętnic przebiegającym z zajęciem tętnic skroniowych. Zwrócono uwagę na konieczność szybkiego postawienia rozpoznania na podstawie kryteriów American College of Rheumatology i wdrożenia leczenia glikokortykosteroidami, by uniknąć ciężkich powikłań choroby.
Słowa kluczowe: olbrzymiokomórkowe zapalenie tętnic, wczesna diagnoza i leczenie, powikłania okulistyczne.

Background

Giant cell arteritis (GCA), also called Horton's disease or temporal arteritis, is the necrotizing inflammation of large and medium-sized arteries leading to disturbances in organs' blood supply. Most often it involves the aorta and its branches [1].

In the typical disease course, an inflammation affects extracranial arteries of the head, primarily the temporal artery [1, 2]. GCA is the most common primary vasculitis. It affects 200 people per million per year [2, 3], usually individuals above the age of 50, women twice as often as men.

GCA most frequently develops gradually. Initially patients complain of systemic symptoms such as fatigue, anorexia, weight loss and/or subfebrile body temperature. Subsequently, in case of lesions affecting the temporal artery, patients complain of severe headaches, usually unilaterally localized in the temporal region, hyperesthesia and, in some patients, one may observe erythema and oedema of a tempus with a thickened temporal artery. In case of visceral branches of external carotid artery being affected, intermittent claudication of a jaw and dysphagia can be observed. Inflammation of subclavian arteries leads to weakness of limbs and of the aorta in its thoracic part to dyspnea and chest pain [4].

Half of the patients with GCA [5] present with symptoms of polymyalgia rheumatica, namely pain of a shoulder and the pelvic girdle muscles and neck muscles. Muscles are tender but muscle strength is preserved. Morning stiffness affecting proximal muscles and lasting for longer than 30 minutes may occur. Additionally, difficulties with getting out of bed or getting dressed are observed.

Severe visual disturbances occur in 36% of untreated patients in a period of 3 days to 10 months from the first symptoms of GCA. [6] In more than a half of patients, Horton's disease affects one eye and if it is bilateral usually 12–17 days pass before the occurrence of the first symptoms in the second

eye. Sometimes ophthalmic symptoms are found isolated, without systemic symptoms [6, 7].

Case report

A 74-year-old woman was admitted to the Department of Rheumatology and Internal Medicine presenting with bilateral vision loss and suspected of GCA. The patient complained of headaches with oedema and erythema of the skin in temporal regions aggravating for the past two months. Antibiotic therapy received as an out-patient didn't bring much improvement. One month later the patient suddenly, within three days, lost vision in her left eye and headaches worsened considerably. On the ground of the angiography of an eye-ground performed at that time the ophthalmologist initially diagnosed GCA and sent the patient to the Rheumatology Department in order to confirm the diagnosis and initiate treatment.

On admission, the patient complained of severe temporal region headache, mostly on the right side, radiating to a jaw and making eating difficult, neck pain and shoulders muscle pain, loss of vision in her left eye and vision deterioration in her right eye. Physical examination revealed disturbances of skin sensation affecting head and fingers with a severe burning sensation. In laboratory tests, elevated levels of inflammation parameters (ESR – 56 mm/hr; CRP – 17.8 mg/l) were found. During ultrasonographic examination of the temporal arteries, superficial temporal arteries and their branches with a presence of hypoechogenic halo and blood flow restriction were visualized (narrow lumen, low-resistance flow, low flow velocities) and in some fragments blood flow was not visualized at all (Figs 1, 2).

Taking under consideration the full clinical picture and laboratory test results, the patient was diagnosed with GCA and treatment with methylprednisolone (total dose of 1.5 g) was initiated. Subsequently treatment with oral prednisone in a dose of 1 mg per kilogram was initiated and clinical improvement regarding headache and hyperesthesia was achieved. Considering progressive deterioration of vision in the right eye, after ophthalmological consultation, it was decided to introduce treatment with intravenous cyclophosphamide in a total dose of 1600 mg. No improvement of vision was observed. After several days complete bilateral loss of vision was diagnosed.

Control laboratory tests revealed decrease of ESR value (20 mm/hr) and normalization of CRP level (1.37 mg/l).

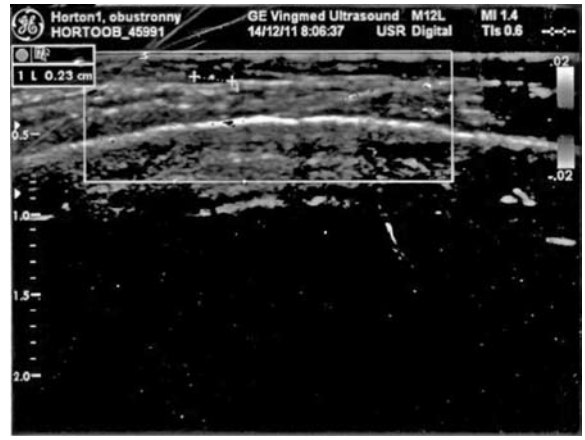

Figure 1.

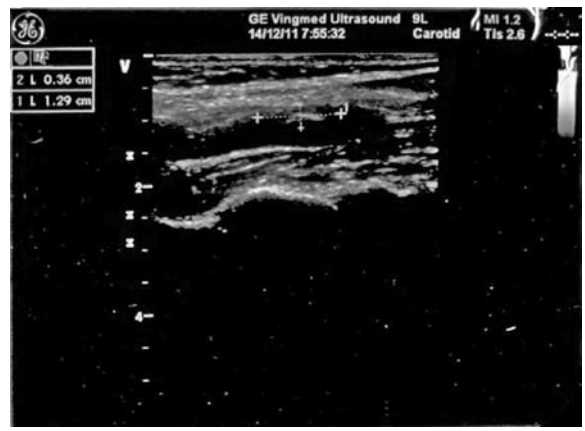

Figure 2.

Discussion

According to the American College of Rheumatology, meeting 3 of the 5 criteria mentioned below indicates GCA diagnosis:

- Disease onset over the age of 50.
- New onset of or new localization of headache.
- Tenderness or decreased pulsation of temporal artery not related to cervical arteries atherosclerosis.
- ESR ≥ 50 mm/hr.
- Abnormal result of temporal artery biopsy [8].

The described case met 4 of these criteria and presented with characteristic symptoms of temporal arteritis.

Chronic activation of the innate and specific immune response is taken into consideration in GCA pathogenesis [9].

In the described case the performed ultrasonographic imaging of temporal arteries proved itself to be helpful because it visualized the dark halo or hypoechogenic space surrounding wall of arterial vessel. It is a classic image of vessels in GCA occurring in 69% of patients [10, 11]. Patients presenting with headaches and systemic symptoms such as malaise, fatigue, loss of weight and hyperesthesia of skin, must be suspected of having temporal arteritis. Onset of GCA symptoms may be sudden and early symptoms may resemble influenza. Un-

treated GCA may lead to loss of vision and the risk of blindness increases with each day. That is why it is so important that physicians of different specializations, especially general practitioners whom patients see most often, make prompt diagnosis and start treatment or send a patient to an ophthalmologist or rheumatologist. It is important not to ignore episodes of vision loss, headaches or skin hyperesthesia that a patient may complain of because they may suggest GCA with posterior ciliary artery, central retinal artery or posterior cerebral artery being affected. If not diagnosed at a proper time it leads to blindness.

Improvement of cooperation between general practitioners and ambulatory specialists may increase the quality of patient health care and eliminate serious complications resulting from delays not dependent on general practitioners [12].

The base of GCA therapy are glucocorticoids (GCS). The suggested initial dose is 60–80 mg of prednisone per day in GCA and 15–20 mg in polymyalgia rheumatica. CRP level should normalize in several weeks or months [10, 12]. Most of the patients require maintenance of treatment with prednisone in a dose of 15–20 mg per day for a couple of months and sometimes even 2 years. Unfortunately not every patient responds to treatment with GCS. Until now standard procedures in the case of GCA refractory to standard treatment have not been worked out. In 2011, a work was published in which researchers evaluated results of treatment with cyclophosphamide (CYC) in patients with GCA refractory to GCS or with severe constriction of arteries in a course of the disease. It revealed that in 90% of patients after 10 cycles of intravenous CYC in a dose of 750–900 mg/m^2 every three weeks, clinical remission and decrease in the disease activity evaluated with PET was achieved [11].

Prompt diagnosis and therapy with glucocorticoids are extremely important because they improve prognosis in GCA and therefore they allow the avoidance of serious complications such as irreversible loss of vision, aneurysms and arterial walls dissection.

In the presented patient, despite intensive treatment with GCS and application of CYC, efforts to spare her vision were not successful. Supposedly, sooner initiation of treatment would have allowed the avoidance of vision loss in her right eye and hence improved the patient's quality of life.

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Praca wpłynęła do Redakcji: 2.10.2012
Po recenzji: 8.10.2012
Zaakceptowano do druku: 10.10.2012