Prevention of serious ophthalmic and cerebral complications in temporal arteritis?

D. Schmidt[1], P. Vaith[2], A. Hetzel[3]

[1]Department of Ophthalmology, [2]Department of Internal Medicine, [3]Department of Neurology, University of Freiburg, Germany.

Please address correspondence and reprint requests to: Prof.Dr. Dieter Schmidt, Department of Ophtalmology, University of Freiburg, Kilianstrasse, 79106 Freiburg; Germany.

Clin Exp Rheumatol 2000; 18 (Suppl. 20): S61-S63.

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Key words: Temporal arteritis, giant cell arteritis, blindness, intracranial arteritis, stroke.

ABSTRACT

Patients

Five patients (mean age 81.6 years) developed bilateral blindness and 3 additional patients suffered cerebral strokes (mean age 58 years) due to temporal arteritis. Bilateral blindness and strokes occurred despite corticosteroid treatment.

Results

In all patients with temporal arteritis, the diagnosis was made too late. Patients with bilateral blindness were referred to the Eye Hospital when one eye had already become blind. The delay between the first symptoms and blindness in one eye was (average) 7 weeks. The interval between blindness of the first and second eyes was (average) 5 days in 3 patients, and simultaneous blindness in both eyes occurred in 2 patients. The other eye also became blind despite mega-doses of prednisone in 3 patients. Three additional patients already showed neurological signs and symptoms at the beginning of the temporal headache. All 3 patients developed strokes after some weeks or months. The wrong diagnosis was made in the first examination(s) by the physician with patients having prodromal signs or symptoms, but who also showed signs of other vascular diseases (diabetes mellitus, hypertension or occlusion of the internal carotid artery) which masked the inflammatory disease of temporal arteritis.

Conclusions

Early diagnosis is essential to prevent severe complications. In patients with a cerebral stroke the early neurological deficits are warning signs which means that one must observe the patient regularly at short intervals. After the diagnosis has been settled, treatment of the patients for several months with a high dosage of corticosteroids is mandatory.

Introduction

Permanent visual loss is one of the most dreadful manifestations of temporal arteritis. Bilateral blindness was reported in 11 out of 239 patients with this disease (4.6%) (1). Some authors have even described blindness that developed during intravenous treatment with a megadosage of prednisone (2-5).

Several questions arise: Can we be sure that corticosteroid really does help to prevent blindness in every patient with temporal arteritis? It has been suggested that some patients with temporal arteritis are refractory to treatment with systemic corticosteroids (5).

Do we begin the treatment too late? Nevertheless, we do know from the literature and from our own experience that corticosteroids can be effective in preventing blindness.

Intracranial arteries are rarely involved in patients with temporal arteritis. We report on 3 additional patients who developed strokes after some weeks or months. In these 3 patients, diagnosis was made very late.

Patients

Between 1996 and 1999, we examined five patients who developed bilateral blindness, and 3 other patients who had suffered a cerebrovascular accident, in each case attributable to temporal arteritis. The mean age of the 5 patients with bilateral blindness was 81.6 years (range: 76-86 years) **(**Table I**)**.

The average time elapsed between the appearance of the first symptoms and the onset of blindness in one eye was 7 weeks (range: 2-14). In the other two patients both eyes became blind simultaneously. In 3 patients blindness of the second eye occurred despite mega-doses of prednisone.

Three additional patients (mean age 58 years, range: 53-67) had already developed neurological signs and symptoms at the onset of the temporal headache. The prodromal symptoms included polymyalgia rheumatica (PMR), headache (temporal or occipital), jaw claudication, and anemia. In 2 patients who were ini-

Table I. Five patients with bilateral blindness.

Patients	Mean delay in therapy	Interval between amaurosis of the 1st and 2nd eye	Signs and symptoms	Eye findings	CRP (mg/dl) (normal 0-0.8) without therapy	ESR (mm/h) without therapy	Fibrinogen (mg/dl) (normal 170-450) without therapy	Treatment
Mean age 81.6 yrs (range 76-86) M:2; F:3	7 weeks (range 2-14)	5 days in 3 patients (mean) Simultaneous blindness in 2 patients	Headache: 3 (temporal pain: 2, occipital headache: 1) PMR: 3 Arterial hypertension: 2 Severe hypochromic anemia: 1 Pain with eye movements: 1 Unilateral occlusion of the internal carotid artery: 1	5 patients with AION 1 with unilateral CRAO (in addition)	16.6 (range 1.6-27.6)	91 (range 54-119)	768 (range 513-1077)	4 patients: 250 mg prednisone every 6 hrs. 1 patient: 500 mg prednisone every 6 hrs. In addition, in 5 pts. heparin and acetyl-salicylic acid

tially treated elsewhere in a hospital, a distinct deterioration with strokes occurred after reduction of the corticosteroids. However, after the referral of the patients to the Department of Neurology, despite maximal treatment additional progression of the neurological defects occurred **(**Table II**)**.

Results

In 3 of the 5 blind patients, the second eye also became blind in spite of megadoses of prednisone.

The 5 patients with bilateral blindness revealed high inflammatory serological changes. These high values, together with the relatively advanced age of the patients, are remarkable and could account for this malignant variant of the disease, i.e. the development of blindness in the 2nd eye in spite of the massive doses of prednisone and in spite of additional treatment with heparin.

All 3 patients with neurological signs and symptoms developed strokes after some weeks or months. The wrong diagnosis was made at the first examination of patients with prodromal signs or symptoms, but who also showed signs of other vascular diseases such as diabetes mellitus, arterial hypertension or occlusion of the internal carotid artery, which masked the inflammatory temporal arteritis.

Most severe neurological breakdowns with spastic pareses and serious neuropsychological involvement (such as mutism or dementia) occurred with the strokes, mainly affecting the territory supplied by the middle cerebral artery.

Strokes occurred despite corticosteroid plus methotrexate and cyclophosphamide.

Conclusions

Immunological and neurovascular examination (ultrasound of the intra- and extracranial vessels) is necessary as soon as the first symptoms are noticed. The early neurological defects are warning signs which indicate the necessity for observing them regularly at short intervals. After the diagnosis has been settled, treatment for several months with corticosteroids is mandatory. In addition, anticoagulation is recommended for high grade stenosis of the carotid artery, particularly of the siphon, together with the administration of acetylsalicylic acid.

It seemed that the intracranial arteries are almost never involved in patients with temporal arteritis (6). However, intracranial arteritis in this disease does occur. In a series of 166 patients with temporal arteritis, Caselli *et al*. found TIA or strokes in 7% of the patients (7).

We think that patients with neurological signs belong to a special group since they differ from other patients with temporal arteritis. They are younger (the mean age of our 3 patients was 58 years, range: 53-67) and the disease runs an unusually serious and malignant course. Many patients with neurological signs, described in the literature, were in the same age group as our patients. The age of the patients was 58 yrs. (8), 66 yrs. (9), 51 yrs. (10), 61 yrs. (11), 52 yrs. (12), 60 yrs. (13), and 65 yrs. (14). The finding of initial neurological signs underlines our belief in the existence of a special group of patients who develop severe neurological defects. In the literature, multi-infarct dementia was described with incomplete improvement of cognitive function in one patient due to corticosteroids (15).

In contrast to the fairly young age of the patients with intracranial artery involvement, we found that patients who developed bilateral blindness were fairly old when blindness occurred (81.6 years, range: 76-86). We think that they also belong to a special group among those

Table II. Three patients with strokes.

Patients	Delay in therapy	First signs and symptoms	Tests	Neurological deficits	Treatment
Mean age 57.7 (range 53-67)	2 to 3 months	Headache: 3 (temporal headache: 2)	CRP: 13.8 mg/dl (range 12-15.5)	Deterioration after stopping or reducing steroids: 2	Corticosteroids Methotrexate Cyclophosphamide Heparin
Sex Male: 2; female: 1		AION: 1	ESR: 89.3 mm (range 70-120)	Initial signs: Paresthesia of one hand:1	
		Weight loss: 2		Episodic pareses and diminished sensibility in both arms: 1	
		History of neurological symptoms: 3	Positive biopsy: 2	Hemipareses: 3 Mutism, dementia: 3	

with temporal arteritis. All had marked serological signs of inflammation. We must, however, admit that early treatment with steroids might have prevented the bilateral loss of sight.

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