OCULAR ISCHEMIC SYNDROME AS AN INITIAL PRESENTATION OF TAKAYASU ARTERITIS

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Abstract. Takayasu arteritis is rare systemic inflammatory disease that involves large vessels in the process. Clinical presentation of Takayasu arteritis varies and many patients may not have characteristic symptoms for a quite long time. In this case report we described a female patient that presented to the ophthalmologist with complaints characteristic for episcleritis but after profound diagnostics appeared to have Takayasu arteritis.

Keywords: Takayasu arteritis, ocular ischemic syndrome, rheumatology.

Окулярний ішемічний синдром як початковий прояв артеріїту Такаясу

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Резюме. Артеріїт Такаясу — рідкісне системне запальне захворювання, при якому в процес залучаються великі судини. Клінічна картина артеріїту Такаясу різна, і багато пацієнтів можуть не мати характерних симптомів протягом досить тривалого часу. У клінічному спостереженні ми описали пацієнтку, яка звернулася до офтальмолога зі скаргами, характерними для епісклериту, але після поглибленої діагностики було встановлено діагноз «артеріїт Такаясу».

Ключові слова: артеріїт Такаясу, окулярний ішемічний синдром, ревматологія.

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Introduction

Takayasu arteritis (TA) is rare systemic inflammatory disease that involves large vessels (predominantly aorta and its branches) leading to a restricted blood flow (due to stenosis and occlusions) or aneurismal degeneration [1]. It was first reported in 1908 by professor ophthalmologist Mikito Takayasu at the 12th Annual Meeting of the Japan Ophthalmology Society as a case of a 21 year old women with an eye disease (arteriovenous anastomosis around the papilla) [2]. The etiology is still poorly understood, but it is considered by now that cell-mediated immunity (in particular CD4+ and CD8+ T cells) plays an active role in the development of the disease, supporting the granulomas formation in genetically predisposed individual [1, 3]. Clinical presentation of TA varies from one patient to the other. Constitutional symptoms of the disease may include headaches, malaise, arthralgias, fever, and weight loss [4]. About 10% of all patients are reported to be asymptomatic [5], making the diagnosis to become a waif during routine examination. Visual disturbance/loss are considered to be the most common ocular symptoms of TA [6, 7].

In this report we tried to highlight a quite rare disorder, TAthat may be easily missed at first presentation of a patient.

Casereport

A 48 year old Caucasian female N. with no significant history of disease presented to the ophtalmologist with complaints of mild pain in both eyeballs and tear over-secretion. A diagnosis of episcleritis was made and topical steroidswere prescribed that had a temporary positive effect. In one month the patient N. came again because both theintensity of eye pain and lacrimation increased as well as visual impairment appeared. After ophthalmoscopic examination ocular ischemic syndrome was suspected. The patient was admitted to the Internal medicine department for full medical and neurological evaluation. On admission clinical examination of organs and systems revealed low-grade fever (37.7℃), increased arterial blood pressure up to 170/90 mm Hg without difference on both arms, claudication and chilling of handsand feet, intermittent arthralgia (mainly of large and medium joints). Neurological assessment showed no organic disorders or focal neurological deficits. The rest was unremarkable.

Diagnostics

Common blood count showed increased erythrocyte sedimentation rate (ESR) up to 45 mm/h. To exclude possible chronic infection or malignancy, tests for viral hepatitis B, C, HIV, Lues, liver function tests, abdominal ultrasound, transthoracic echocardiography and ultrasonography of the mammary glands, gastroscopy, chest X-ray, head MRI were performed and were negative. To exclude systemic disorders tests for ANA, ANCA, C-reactive protein, rheumatoid factor were done however were negative as well.

Figure 1

CT-angiography, coronal view: thickening of the walls of right and left common carotid arteries with narrowing of the lumen

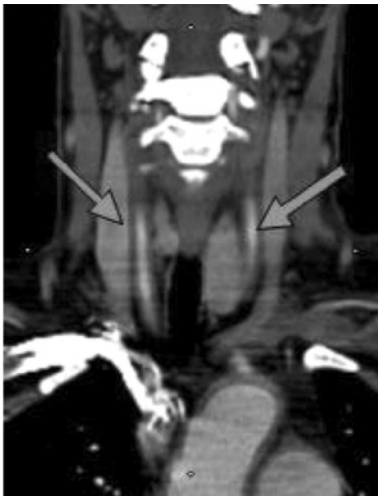


Figure 2

CT-angiography, coronal view: thickening of the walls of right and left internal carotid arteries with narrowing of the lumen

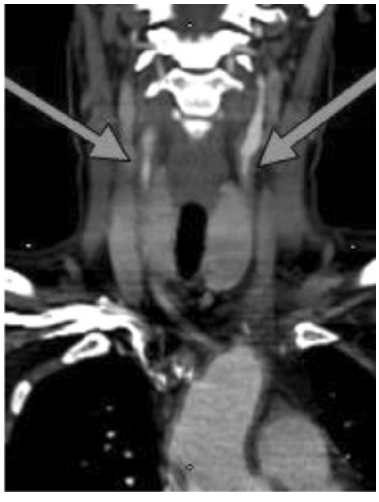
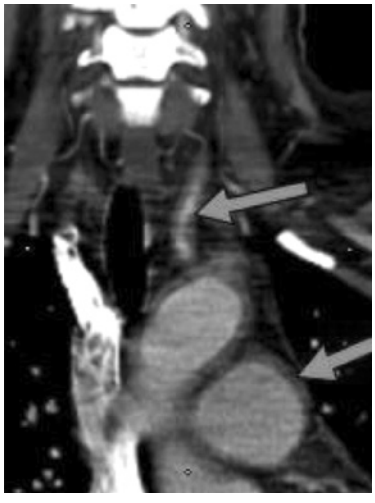


Figure 3

CT-angiography, coronal view: thickening of the walls of left internal carotid artery with narrowing of the lumen



Carotid duplex revealed extensive stenosis of common carotid arteries up to bifurcation: right 40-45%, left 30-35%. Aa. vertebrales without stenosis.

For differential diagnosis computed tomography-angiography of head, neck and thorax with intravenous iodine enhancement was performed: thickening of the walls of ascending aorta and aortic arch, brachiocephalic trunk, right and left common andinternal carotid arteries up to 1.5-2.6 mm with narrowing of the lumen was revealed (fig. 1-3). The diagnosis of TA wasmade.

Discussion TA is considered to progress through three stages [4]: 1. early systemic stage (prevasculitic) — constitutional symptoms may occur; 2. vascular inflammatory stage — clinical presentation depends on the vessel involved; 3. burned out stage — remission due to fibrosis formation of an actively involved vessel.

Thorough examination is required paying the most attention to systolic blood pressure difference between arms (and legs), peripheral pulses difference and ocular examination not to miss the diagnosis. In our case the patient N. had no difference of systolic blood pressure between arms though she had arterial hypertension.

Ophthalmologic examination often reveals retinal pathology (haemorrhages, micro aneurysms, arteriovenous shunting, and ischemia), optic atrophy and cotton wool spots [8]. Ocular ischemic syndrome is a condition associated with stenosis (or occlusion) of a carotid artery [9].

First diagnostic criteria were presented by Ishikawa in 1988 [10] that later in 1996 were modified by Sharma et al. [11] (table 1). For the diagnosis of TA our patient fulfilled Sharma's criteria, but not the Ishikawa's one as the age at onset was 48 years.

The other diagnostic criteria for TA that are widely used are the one of American College of Rheumatology (ACR) [12]. Due to the latter the patient is considered to have TA if at least three out of six criteria are present which makes it possible to establish the diagnosis of TA in our patient.

In our case TA should be differentiated with another systemic granulomatous vasculitis–giant cell arteritis (GCA). They have lot of similarities in clinical presentation, arterial lesions that causes the debates whether TA and GCA are two diseases or the same one [13, 14]. Using the ACR criteria for GCA [15] our patient couldn't fulfil the diagnosis of GCA (table 2).

Case history outcome

The treatment using Methylprednisolone (MP) 32 mg/day was initiated together with symptomatic therapy (antihypertensive drugs, antiplatelet agents, anticoagulants). The dose of MP was gradually tapered in one month after the symptoms regression.

Table 1

Comparison of Ishikawa and Sharma criteria for TA

Ishikawa diagnostic criteria [10]	Sharma diagnostic criteria [11]
Obligatory criterion: age at onset <40 years	-
Major criteria: 1. Left mid subclavian artery lesion 2. Right mid subclavian artery lesion	Major criteria: 1. Left mid subclavian artery lesion 2. Right mid subclavian artery lesion 3. Symptom duration at least one month duration
9 Minor criteria: 1. High ESR 2. Carotid artery tenderness 3. Hypertension 4. Aortic regurgitation or annuloaortic ectasia 5. Pulmonary artery lesion 6. Left mid common carotid lesion 7. Distal brachocephalic trunk lesion 8. Descending thoracic aorta lession 9. Abdominal aorta lesion	Minor criteria: 1. High ESR 2. Carotid artery tenderness 3. Hypertension 4. Aortic regurgitation or annuloaortic ectasia 5. Pulmonary artery lesion 6. Left mid common carotid lesion 7. Distal brachocephalic trunk lesion 8. Descending thoracic aorta lession 9. Abdominal aorta lesion 10. Coronary artery lesion
To fulfill the diagnosis of TA	
1 obligatory + a. 2 major or b. 1 major and 2 or more minor or c. 4 or more minor	a. 2 major or b. 1 major and 2 or more minor or c. 4 or more minor

Table 2

The comparison of ACR criteria for TA and GCA

Takayasu arteritis [12]	Giant cell arteritis [15]
1. Age at disease onset <40 years 2. Claudication of extremities 3. Decreased brachial artery pulse 4. Blood pressure difference >10 mm Hg 5. Bruit over subclavian arteries or aorta 6. Arteriogram abnormality	1. Age at disease onset >50 years 2. New headache 3. Temporal artery abnormality 4. Elevated ESR 5. Abnormal artery biopsy
To fulfill the diagnosis of TA	To fulfill the diagnosis of GCA
3 of 6 (necessary 3 of 6)	1 of 5 (necessary 3 of 5)

Informed consent

The informed consent for the publication of this report was obtained from the patient by the corresponding author.

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