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Takayasu arteriopathy — rare, but worth to remember disease

Choroba Takayasu — rzadka, ale godna zapamiętania

Commentary to the article:

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Takayasu's arteritis (TA) is very rare in general and also Polish patient population (2.6 patients per million population per year in US) [1] therefore this case report is an important opportunity to discuss both the disease and its therapeutic options. Its nature is autoimmune although some genetic predisposition was also reported (HLA haplotype B52 and DR4 are more common in this patient population). Moreover some infectious factors may coincide with the onset of the disease. Heat shock protein HSP-65 found in aortic tissue taken from the patients suffering from TA is similar to Mycobacterium tuberculosis 65-kDa HSP [2]. This HSP can be a target for the gamma-delta T-cells secreting in massive amounts a membrane-disrupting protein — perforin. The same authors [2] suggest that not only cellular but also humoral immunologic response is activated in this disease. The arterial wall that is involved in inflammatory process is affected through vasa vasorum and all layers of its structure are being destroyed.

Clinically disease starts with symptoms consistent with typical virus infection including fever, weight loss, myalgia and arthralgia. In this phase it causes inflammatory infiltrates in all three layers of the arteries. In the chronic phase intimal proliferation and fibrotic changes in the vessel wall lead to stenosis or aneurysm formation [3]. It is easier to establish the diagnose in the first phase if the patient presents with the pain at palpation of the affected artery (most often one of the carotids). Later ischaemic symptoms and age of the patient (by the rule below 40 — the main criterion in Ishikawa clinical diagnosis but not so strongly supported in newer classifications) are the main clinical data for beginning right diagnostic evaluation.

The disease can be devastating in the group of young patients that consists predominantly of women (2–8 times more frequently suffering from TA) [1] and in the group of children the mortality can be as high as 35% at 5 years [4].

In the case report presented by Kalawski et al. [5] 37 years old female patient presented typical vascular changes observed in TA involving aortic arch branches, right coronary artery (proximal narrowing) and aortic valve insufficiency. This distribution of malformations is typical for Asian patients who present vascular changes localised mainly in aortic arch, its branches and ascending aorta [6], but in other groups renal arteries and abdominal aorta can be also the main localisation of the disease [7] even in 38% of patients. The operation was prompted by TIA that was the main cause for patient admission and evaluation that has lead to semielective surgery that was mainly aimed at solving the problem of RCA proximal occlusion and severe aortic valve insufficiency (patient was referred to cardiac surgery department but not to vascular surgery department). Coronary arteries are reported to be affected by inflammatory narrowing or occlusive changes in 10 to 30% of TA patients [8] and optimal results are achieved

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using surgical treatment predominantly with saphenous vein grafts (36% of restenoses after surgical treatment *vs* 78% of restenoses after angioplasties). This finding seems to be related to report from American group [9] that stated that more than two thirds of patients present difficulties in performing routine daily activities and about one-fourth of all patients are unable to work. Very good decision was made to revascularise the supraaortic vessels at the same time because the danger of the brain perfusion disturbances during operation and perioperatively could have been high with a poor blood supply diagnosed preoperatively. Unfortunately the case report does not bring the clear information whether the perfusion of the brain was maintained through the graft during cardiopulmonary bypass time. If the brain and upper extremities revascularisation would not be possible during the same operation the aortic valve replacement could have been reconsidered having in mind that heart failure can progress slowly in the group of patients with TA and severe aortic insufficiency when inflammatory parameters and systemic hypertension are under control [10]. Two more issues are worth mentioning here. First: regarding not easy problem of the perioperative antinflammatory treatment of the patients and second: the durability of the anastomosis performed in aortic tissue that can be affected by inflammatory changes at the time of symptom-prompted operation or later on. Matsuura et al. [11] presenting the results of 21 consecutive arch and hemiarch operations in patients with TA supports the idea to hamper the inflammatory process with preoperative steroid administration lasting 8.1 years (mean). The aim of the treatment in his strategy is bringing the ESR and CRP level to normal limits before the operation. From other hand Miyata et al. [12] in his publication reporting the results of surgical interventions in 103 TA patients (treated over 40 years) does not find any correlation between formation of the aneurysm in the anastomosis and anti-inflammatory treatment regime. In his analysis higher frequency of anastomotic aneurysm formation was observed in patients in whom the anastomosis was performed in the aneurysmatic but not stenotic tissue. The overall anastomotic aneurysm formation in TA patients was 12% at 20 years.

In the end we would like to congratulate dr Kalawski and his coworkers for the successful treatment of the very complex case.

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