Diagnosis and indications for revascularization in Takayasu’s Arteritis: Report of two cases and literature review

Introduction

Takayasu’s arteritis (TA) is a chronic inflammatory disease of the vessels. The aorta and its main branches such as the supra-aortic trunks, renal and digestive arteries are predominantly affected. This inflammation leads to thickening of the vascular wall due to fibrosis that is the most characteristic early sign of Takayasu’s disease [1,2]. This thickening leads to stenosis and formation of thrombus which are responsible for various complications such as ischemia [3]. The diagnosis of this disease is based on a set of radio-clinic arguments including the clinical condition, the topography of involved artery, its appearance (stenosis or ectasia) and the association with cutaneous and visceral lesions [4].

Case Report

Case 1

Mrs. DB was 62 years old. She consulted for bilateral impotence of the upper limbs associated with paresthesia of the upper limbs. On examination, she showed a sign of left carotid arterial pressure > 140/90 mmHg, stenosis or occlusion of the middle portion of the left carotid artery or distal third of the brachiocephalic trunk in the angiography and many others. Therefore, a rating based on clinical, radiological and histological criteria appears to be more useful in practice [4]. On the other hand, coronary and arterial pulmonary lesions can be observed during this disease. Indeed, the presence of 2 major criteria or 1 major criterion + 2 minor or 4 minor criteria suggests a high probability of Takayasu’s disease [5]. Above all, the treatment of TA is medical and consists of giving corticosteroid and even immunosuppressive treatment sometimes. Revascularization is considered in case of non-response to medical treatment or in case of complications such as visceral ischemia, aneurysm or thrombosis. Regarding the revascularization, open surgery and endovascular treatment are currently discussed [6]. We report two cases of Takayasu arteritis with arterial complications that were managed in our department. Then we will discuss the diagnosis and indications of revascularization while we will be reviewing the current data of the literature.

Abstract

Takayasu’s arteritis (TA) is an inflammatory disease of large vessels that predominantly affects the aorta and its main branches such as supra-aortic trunks, renal and digestive arteries. The diagnosis is based on criteria proposed by the American College of Rheumatology and modified by Sharma. These vascular lesions present a problem of surgical indications because of their pathogenic particularity. In this work, we report our experience on the diagnosis and management of two cases of TA. The case 1 was a 62-year-old female patient diagnosed with stenosis of the common carotid artery and the right subclavian artery. A bypass between the carotid artery and the subclavian artery was indicated but not performed. The second patient was a 23-year-old female patient diagnosed with renovascular hypertension. Investigations showed a significant stenosis of the left renal artery. She underwent angioplasty-stenting of the left renal artery and the result was good. Her echocardiography showed left ventricular and atrial hypertrophy and both. The two patients had no indirect signs of myocardial ischemia and arterial pulmonary injuries.
and a feeling of coldness of the hands. She was diabetic type 1 and was being treated with insulin. Also, she was treated for hypertension. The clinical examination did not found trophic changes or upper limb deficit. At the right arm, the pulses were abolished on humeral, radial and ulnar arteries. Clinically, she had no cardiac and pulmonary signs. In addition, a right carotid bruit was auscultated. The arterial ultrasound showed many lesions including a severe occlusion of the right subclavian artery ostium (Figure 1), a stenosis (greater than 60%) of the middle segment of the right common carotid artery, an occlusive stenosis of the right internal carotid artery and a stenosis (50% of the lumen) of the left common carotid artery. The angio CT of the supra-aortic trunks also showed more lesions that consisted of a severe stenosis of the proximal right subclavian artery, an occlusion of the vertebral arteries on the V0 and V1 segments and a stenosis of the right internal carotid artery (Figures 1,2). There was not calcification. The echocardiography showed a conserved ejection fraction of left ventricle and a lack of myocardial ischemia. CRP and platelet counts were normal. Taking into account the clinical arguments and complementary investigations, the diagnosis of Takayasu’s disease was made. Immunosuppressive therapy was started in addition to naftidrofuryl and acetylsalicylic acid. Six months later, the symptomatology was persisting. Then an arteriography was prescribed. It showed a 5 cm-long occlusion at the origin of the right subclavian artery.

Also, it revealed an occlusion of the right renal artery. It was impossible to catheterize the right subclavian artery and as such it was impossible to perform endovascular procedure for revascularization. Subclavian artery bypass was indicated but was not performed because of refusal of the patient. She discharged from hospital but later, she was missing during the follow up.

Case 2

RB was a 23-year-old female patient who was suffering from hypertension for 3 years. Despite her triple anti-hypertensive therapy (bisoprolol, amlodipine and indapamide), her blood pressure remained high. On examination, RB presented good general condition. In the upper left limb there was an abolition of radial and humeral pulse without signs of ischemia. The left carotid pulse was absent. Clinically, she had no pulmonary and cardiac signs. The Doppler ultrasound of the supra-aortic trunks revealed diffuse wall thickening in the brachiocephalic artery and common carotid with significant reduction of their lumen responsible of damping of internal carotid flow. The flow of the right vertebral artery is damped on its segment 2. The angio CT showed a long and severe stenosis of the axillary artery (6 cm) and of the left axillary artery on 13 mm in a context of diffuse parietal arterial thickening (Figure 3). In addition, there was bilateral stenosis of the renal arteries just after theirs origins. The length was about 13 mm on the right and 10 mm on the left (Figure 4). The echocardiogram noted a left atrial

Figure 1: Angio CT of supra-aortic trunks showing pre-occlusive stenosis of proximal right subclavian artery.

Figure 3: Angio CT showing a long and severe stenosis of left subclavian and axillary artery. There was a diffuse wall thickening.

Figure 2: Stenosis of the interne carotid after the sinus of the right carotid. There was no calcification.

Figure 4: Bilateral stenosis of the renal arteries. It was nearly occlusive at left.
and ventricular hypertrophy, a conserved ejection of the left ventricle and a lack of indirect signs of myocardial ischemia. The biology revealed an increased erythrocyte sedimentation rate (.42 mm Hour 1), the CRP was also increased to 96 mg/L with leucocytosis at 16,700 / mm3. The Waaler Rose test was negative. The final diagnosis was vascular lesions in a context of Takayasu’s disease. Endovascular revascularization of the renal arteries was decided. The perioperative arteriography confirmed a stenosis of 90% of the left renal artery and 35% of the right. Angioplasty-stenting was performed on the left. The postoperative course was uneventful and the patient was discharged from the hospital on day 3. The second postoperative control on day 45 was good; in particular the blood pressure was normalized (120/70 mm Hg). However a follow up of the other vascular lesions is still ongoing.

Discussion

Takayasu’s arteritis (TA) was described for the first time in 1908 by Mikito Takayasu, a Japanese ophthalmologist [7]. Like in our two cases, it is a pathology that mainly affects women in about 80 to 100% of the cases [8]. It occurs in the young patients during theirs 20’s or 30’s [1]. Indeed in the series of Kerr et al. [9], only 2 of 10 patients were aged over 40 years old at the period when diagnosis was made. In fact, the delay between the first symptoms and the diagnosis is variable and the disease can be discovered late like in our case 1 (62 years old) because the signs are not very specific during the pre-occlusive phase of this disease. In addition, the absence of cephalic, ophthalmic and articular signs made us eliminate the Horton’s disease. On the other hand, a biopsy of the temporal artery would be more formal. The diversity and disparity of clinical signs led the American College of Rheumatology to suggest diagnostic criteria in 1990 [3] which were modified by Sharma in 1996 [7]. This intends to facilitate the diagnosis. Thus, the case 1 presented 2 major criteria and the case 2 had 1 major criterion and 3 minor criteria. Taking into account the Sharma score, both our patients were diagnosed with TA with a sensitivity of 92.5% and a specificity of 95% [5]. No biopsy or biomolecular tests were prescribed because the pathogenesis of TA remains elusive and there is no biological specific test [10,11]. New imaging tools such as computerized tomography or magnetic resonance angiography, fludeoxyglucose positron emission tomography – computed tomography and recently contrast–enhanced ultrasonography are frequently used in the diagnosis and to assess vascular inflammation. Accumulating evidence shows that biological agents such as anti-tumor necrosis factor agents, tocilizumab and rituximab could be used effectively in refractory cases [12]. According to literature data, the corticosteroid therapy is the first-line treatment and in the event of failure the addition of methotrexate may stabilize the disease [13]. The medical treatment was different in our 2 patients. In the case 1, it consisted of an arterial vasodilator, a platelet aggregation inhibitor and an immunosuppressive drug in the place of corticosteroid due to diabetes whereas the case 2 benefited from antihypertensive and corticosteroid therapy. This is mainly related to the difference in the clinical features of the patients. In the literature, no therapeutic protocol is unanimously recognized [8] although corticosteroid is recommended during the acute resurgences. The efficacy of anticoagulants and antiplatelet agents has not been demonstrated because hypercoagulability and thrombosis are not common complications of TA [4].

The Takayasu arteritis surgical treatment is indicated in case of failure of medical treatment. Revascularization can be done by endovascular or by bypass. It was necessary in our patients who were already in the occlusive stage of the disease. For the case 1, the failure to catheterize the right subclavian artery indicated subclavian artery bypass. Moreover, the indication for revascularization of the right common carotid artery was relevant due to the significant stenosis of this artery. Open surgery is an effective method according to Kim et al. [6] as they used 15 surgical bypasses on 25 patients (60%) with lesions of the supra-aortic trunks. This would avoid major complications such as stroke. The case 2 underwent endovascular treatment: angioplasty – stenting of the renal arteries with a good result. This indication was justified by the Chaudhry’s study which suggests renal revascularization in all cases of renal artery stenosis ≥ 70% with or without reno–vascular hypertension [11]. It is a less invasive and less morbid method compared with surgical bypass and which shortens the hospitalization delay [11]. To refresh, the case 2 stayed only 3 days in the hospital. Angioplasty only is associated with a high rate of re-stenosis [6], hence the interest of stenting. The long-term prognosis depends on major complications such as aortic insufficiency, retinopathy, arterial aneurysms and increasing erythrocyte sedimentation rate [11]. According to current data regarding TA revascularization, conventional surgery is indicated in large vessel lesions [6,14], whereas endovascular techniques remain more effective in stenosis of the renal artery [11]. This attitude is explained by the pathogenesis of the vascular lesions of this disease. Indeed, the latter is characterized by a pan-arteritis associating adventitial thickening, focal infiltration of the tunica media and intimal hyperplasia. The association of these micro-lesions results in stenosis or occlusion [15]. Thus, in the case of angioplasty–only or associated with stenting there is a continuous intimal cell proliferation that will create a secondary stenosis [6]. Other therapeutic modalities are being explored for the treatment of TA such as TNF inhibitors and active angioplasty balloons but they have not proved their worth yet [15].

Conclusion

The surgical management of vascular lesions of Takayasu’s disease remains accessible in the developing countries because conventional open surgery is much more appropriate than endovascular surgery. The main challenge is the early diagnosis of these vascular lesions.

References


