36 Visceral Artery Involvement in Takayasu Patients: Treatment Options

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Takayasu arteritis (TA) is a chronic non-specific inflammatory arteritis which affects large and medium caliber arteries, predominantly the aorta and its main visceral branches. Its cause remains unknown. The disease mostly occurs in young patients during the second or third decade and has been now identified in both genders and many ethnic and racial groups worldwide. It is a lifetime affection with recurrence and progression of symptoms related to occlusive of aneurysmal complications. When vessel inflammation leads to wall thickening, fibrosis, stenosis, and thrombus formation, symptoms reflect end-organ ischemia. More acute inflammation can destroy the arterial media and lead to aneurysm formation. The coexistence of occlusive and aneurysmal lesions is highly evocative of the diagnosis of Takayasu arteritis.

Whether isolated or more frequently associated to aortic occlusive or aneurysmal lesions, lesions of visceral arteries may be responsible for specific symptoms. Although commonly encountered in thoracoabdominal aortic occlusive disease, renovascular hypertension may also be directly related to severe renal artery stenosis. Less frequently, due to the usual development of collateral circulation, occlusive lesions of the main intestinal arteries may lead to the occurrence of chronic or acute mesenteric ischemia.

This chapter aims to describe the specific involvement of visceral arteries during Takayasu arteritis and the current treatment options.

36.1 Involvement of Visceral Arteries

The vast majority of occlusive aortic lesions, especially when they affect the interrenal or abdominal aorta, coexists with lesions of visceral, namely, renal or intestinal, arteries. These lesions may be occlusive and aneurysmal or may associate a post-stenotic dilatation distal to a tight stenosis.

Involvement of renal arteries during Takayasu arteritis might be diagnosed in 40–60% of patients submitted to systematic angiography. Lesions are more frequently occlusive and bilateral. Occlusive lesions of the renal arteries are usually located at the level of their ostia or close to it. Most of the time, they do not extend distal to the first 2 cm of the main renal trunk. In that situation, they represent a direct extension of aortic lesions, whereas more distal and extending lesions may occur independently of aortic occlusive disease. In rare cases, they may affect the hilar division branches of the artery. During their progression, these lesions may lead to renal atrophy following complete occlusion of the

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renal artery. In more favorable cases, the occlusion remains located to a short segment of the artery, and blood supply to the kidney is provided by a rich collateral network issued from adrenal, lumbar, and ureteral arteries. This providential development of collaterals preserves the viability of the kidney and allows to consider an adapted revascularization in the absence of renal atrophy. A part from post-stenotic dilatations, there are rare cases of true fusiform or sacciform aneurysms of the renal arteries.

Hypertension is the main revealing symptom of renal artery involvement in the Takayasu disease. Most of the times, it is uncontrolled hypertension despite intensive medical therapy. Alternatively, hypertension may be caused by a severe aortic coarctation and then predominates at the level of the upper limbs, while femoral pulses are absent unless an extensive collateral network has already developed. Hypertension may be difficult to diagnose in case of bilateral subclavian or axillary artery stenosis. Sometimes, lesions of renal arteries are depicted by the imaging workup in the absence of renovascular hypertension.

Intestinal arteries are frequently involved in Takayasu disease. The superior mesenteric artery (SMA) is the most frequently diseased. The lesions are similar to that observed at the level of the renal arteries. They may include short ostial and paraostial stenoses or more frequently long lesions of the arterial trunk. Therefore, occlusive lesions of the SMA are usually longer than atherosclerotic lesions. In case of a single involvement of this artery, blood supply is coming from the celiac artery via peripancreatic anastomoses. In case of combined celiac artery involvement, an ascending Riolan artery provides an efficient supply to the intestine and supramesocolic organs. This artery may also develop and provide the unique blood supply to the lower limbs in case of severe stenoses or complete occlusion of the infrarenal aorta. Subsequent overflow may be responsible for the development of aneurysms of this collateral vessels.

Acute and chronic intestinal ischemia are rare during Takayasu disease. In most cases, the extensive development of a rich collateral network precludes the occurrence of intestinal symptoms, even in case of multiple occlusions of intestinal arteries.

36.2 Preoperative Workup

CT scan appears to be the most relevant preoperative imaging technique. At the acute phase of the disease, CT scan may depict the severity of the arteritis which typically appears as a wall thickening preexisting to any vessel stenosis of dilatation. Preoperative corticosteroid therapy may reduce this thickening. At the chronic stage of the disease, CT scan may depict circumferential aortic wall calcifications with potential extension toward the visceral arteries. CT angiogram provides an accurate workup of renal and intestinal vessels and of their main branches. Distal arterial runoff supplying the kidneys and the intestine is also addressed by CT angiogram. Measurement of the long axis of the kidney and evaluation of the corticomedullary index are easily done.

TEP scan is probably the most accurate examination to predict the intensity of periarteritis, both at the level of the aorta and at the level of the visceral arteries. TEP scan is essential to estimate the inflammatory status of the disease before attempting to a direct surgical revascularization [1].

Digitalized angiography is now rarely indicated since the accuracy of CT angiogram is excellent. The technique may remain useful in case of distal extension of occlusive lesions of the renal or intestinal arteries, less accessible to CT angiogram. Selective catheterization of renal or intestinal arteries may help to diagnose precisely the development of collaterals and the distal extension of stenotic lesions.

36.3 Treatment Indications

The goal of the treatment of Takayasu arteritis is to relieve inflammation in the arteries and to prevent potential complications related to end-organ ischemia. Even with early detection and treatment, Takayasu arteritis may be challenging to manage, although recent advancements in medi-

cal and surgical treatments, including endovascular procedures, have improved the prognosis of patients with Takayasu arteritis [1]. Biological inflammation at the time of revascularization increases the likelihood of postoperative complications in patients with Takayasu arteritis. Therefore, first-line treatment is immunosuppression, primarily with corticosteroids. With glucocorticoid treatment, remissions may occur in 40–60% of all patients. About 40% of all steroid-resistant patients respond to the addition of cytotoxic agents. Approximately only 20% of all patients are resistant to any kind of medical treatment [1]. Therefore, aggressive medical and surgical treatment is required for patients suffering from major complications and a progressive disease course.

Severe hypertension should always be medically addressed before surgery. Several medical treatments may sometimes be associated to obtain a significant decrease in blood pressure level. Renal percutaneous arterial angioplasty may temporarily help to decrease blood pressure in order to schedule a short-delayed more complete surgical repair.

In most cases, lesions of visceral arteries during Takayasu arteritis are surgically treated in combination to aortic repair, either for occlusive or for aneurysmal abdominal or thoracoabdominal aortic lesions [2]. The surgical approach is thus mainly designed according to aortic patterns of lesions, given that (right) renal and distal intestinal arteries are best approached by midline laparotomy than by isolated thoracophrenolumbotomy. Techniques of open revascularization of visceral arteries are numerous. They should take into account the type and extension of the visceral arterial involvement and the estimated technique of aortic revascularization.

Direct reimplantation of visceral arteries into the native aorta is rarely indicated. Most frequently, visceral artery lesions are not limited to the ostia, and they extend distally, thus precluding the opportunity of such a revascularization. Thickening of the aortic wall confers also a high risk of anastomotic intimal hyperplasia.

Transaortic visceral artery endarterectomy is technically hazardous, if not impossible, due to the severity of aortic wall involvement and to the frequent distal extension of visceral artery lesions.

Bypasses are the most appropriate technique of revascularization for visceral artery involvement during Takayasu disease. They offer a treatment for severe and long occlusive lesions of both renal and intestinal arteries. In this specific young population, we advocate the use of autografts rather than prosthetic (Dacron or PTFE) grafts. Given the frequent late deterioration and dilation of venous autografts, we recommend the use of arterial autografts and especially the harvesting of the superficial artery (or sometimes the internal iliac artery). Superficial femoral artery autograft is often a congruent conduit which provides excellent long-term anatomical and hemodynamic outcomes.

The proximal anastomosis is rarely made on a healthy segment of the native abdominal aorta or iliac arteries, due to potential future involvement of these arteries by an evolutive disease. Distal thoracic aorta, if healthy, may be at the origin of the bypass which thus takes an anterograde conformation. In rare cases, mainly in patients already approached at the level of the descending or abdominal aorta, bypasses can be proximally implanted into the ascending aorta with good long-term results [3]. Less frequently, due to potential secondary evolution of the intestinal arteries, bypasses to the renal arteries may originate from the splenic, the hepatic, or the superior mesenteric arteries [4]. In these last alternatives, it is mandatory to assess precisely the absence of aortic lesions proximal to the level of the renal arteries, in order to avoid a secondary deterioration of the inflow intestinal vessel. Therefore, redo surgery is probably the most appropriate condition for such extra-anatomic bypasses to the renal arteries.

The most reliable origin of a bypass to the visceral arteries in Takayasu disease is certainly an aortic prosthesis. The proximal anastomosis of an arterial or venous autograft should be done after excision of a small circle of the prosthesis, in order to avoid future hyperplasia and development of an anastomotic stenosis. A prosthetic excision far larger from the native diameter of the

autograft exposes to secondary disruption of the anastomosis, especially when venous grafts are used, and may lead to acute or chronic formation of an anastomotic false aneurysm.

Intestinal arteries are quite easy to treat with an anterograde bypass. The right renal artery may be revascularized from the infradiaphragmatic segment of a ventral aorta (bypass originating from the ascending aorta), with the graft thus positioned behind the hepatic pedicle. The right renal artery is less easy to approach during a dorsal aortic revascularization (bypass originating from the descending thoracic aorta). In this case, it is better to bypass from the lower abdominal segment of the prosthesis with the graft positioned behind the duodenum of the inferior vena cava as it would be done during an isolated right aortorenal bypass. Bypasses to the left renal artery are sometimes difficult to position due to the relatively short distance separating the dorsal aortic prosthesis from the hilum of the left kidney. Direct left renal artery reimplantation into the prosthesis is an alternative to short bypasses or to long bypasses running laterally to the aortic prosthesis. If necessary, left renal bypass may originate from the superior mesenteric artery or from a bypass to this artery.

Renal autotransplantation may be indicated in case of distal occlusive or aneurysmal lesions, extending to the hilar branches, and in case of homolateral multiple and diseased renal arteries [5]. Renal autotransplantation is also useful in case of intense periarterial fibrosis which renders the surgical exposure of the renal artery and its branches particularly difficult. Autotransplantation allows ex situ repair of complex and often distal lesions with the use of an arterial autograft. The heterotopic transposition has our preference, and the kidney is thus replaced in the iliac fossa, the graft being proximally anastomosed to the iliac artery or to a prosthesis. Thanks to renal autotransplantation, our indications of primary nephrectomy are now limited to severe and nonreversible parenchymatous kidney lesions, to small kidneys (long axis less than 7 cm) with non-accessible lesions of the distal arterial branches, or to infarcted kidneys following a failed attempt to revascularization. Primary nephrectomy may also be indicated in combination of contralateral renal revascularization, in case of severe renovascular hypertension complicating an ipsilateral renal infarction.

Given the usual good physical status of young patients with Takayasu disease, we often advocate a single-stage revascularization of both aortic and visceral artery lesions. More complex revascularizations may also include the simultaneous treatment of lesions of the supra-aortic trunks. Logically, supra-aortic trunks are first treated when patients are deemed at high risk for a single-stage procedure.

36.4 Surgical Results

As they are the more frequent symptomatic lesions among visceral artery lesions in Takayasu disease, results of renal artery revascularization are the easiest to analyze. Previous data from our institution, published by Kieffer et al. [4], showed satisfactory early and long-term outcome in 24 patients with Takayasu arteritis who underwent surgery for renal artery stenosis. Renal artery revascularization was unilateral in 46% and bilateral in 54%. During the 61.3-month follow-up, repeated renal artery revascularization was required in only four patients. Hypertension was cured in 63%, improved in 31%, and unchanged in 6%. We also reported [5] a series of 26 renal autotransplantations in 19 Takayasu patients and demonstrated that hypertension normalized or improved in 89% of patients who survived the procedure. On the basis of this experience, we have progressively made renal autotransplantation the procedure of choice for treatment of patients with Takayasu disease with renovascular lesions that would be difficult to dissect because of the inflammatory nature of the disease or extensive occlusive lesions of distal branches. We now also advocate renal autotransplantation in patients with aortic lesions that are not sufficiently severe to warrant reconstruction but that would interfere with aortorenal or iliorenal bypass or direct reimplantation. In this regard, it should be noted that thickening of the aortic wall is almost always associated with involvement of

the hepatic, mesenteric, and splenic arteries, thus excluding their use for bypass inflow. Overall postoperative morbidity, especially represented by pulmonary complications, is linked mainly to the extent of the operative exposure that is necessary to achieve single-stage reconstruction of thoracoabdominal aorta and renal involvement, which we advocate in young patients.

36.5 The Endovascular Alternative

Balloon angioplasty and stenting are now currently proposed to treat involvement of visceral arteries during Takayasu disease [6–9]. Stent indications are residual stenosis of more than 30% of the native diameter, significant dissection following angioplasty, and distal lesions. Due to the fibrotic nature of the arterial lesion, endovascular treatment is in most reports considered technically successful if the arterial lumen after treatment is less than 30% residual stenosis. Similar to surgery, endovascular repair is recommended at a time of quiescent Takayasu arteritis disease to avoid early and late arterial complications. In our opinion, endovascular procedures are best used as a temporary alternative to improve rapidly severe renovascular hypertension or subacute or acute mesenteric ischemia. Nevertheless, several studies have shown enthusiastic results of renal angioplasty in renovascular hypertension caused by Takayasu arteritis. Tyagi et al. [8] reported a 44.2% cure rate and an improvement rate of 48.4% but with a 13.5% restenosis rate. Sharma et al. [9] reported that angioplasty was beneficial in 89% of patients and their restenosis rate was 16%. In these studies, the follow-up period was about 2 years only. In a more recent series [6] with a longer average follow-up period of 5 years, rates of restenosis were 9% after angioplasty and 62.5% in the setting up of a stent. Renal artery patency rates were 100%, 90.9%, and 90.9% in angioplasty alone; the rates of primary permeability by the introduction of a stent were 62.5%, 37.5%, and 37.5% and assisted permeability 87.5%, 75%, and 50% at 1, 3, and 5 years, respectively.

In conclusion, in the treatment of visceral arteries involved by Takayasu arteritis, surgery remains an effective and durable alternative, especially for patients with associated aortic pathologies and in case of failure of first-line endovascular treatment. The establishment of a stent should probably be reserved for the failure of plain balloon angioplasty.

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