

Chronic Occlusion of Hypoplastic Aorta in a Patient with Right Renal Artery and Right Kidney Agenesis

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Abstract

A 53-year-old woman presented in vascular emergency department with black change at the top of the right toe and blue staining fingers of the both feet. Patient was examined clinically, with CDS (Color Duplex Scan) and MSCT angiographic examination. It has been confirmed the absence of peripheral pulses of both legs, decreased ABI (right 0.23; left 0.35), thrombosis of the juxta- and infra-renal small sized aorta (diameter 16 mm just above LRA, 11 mm just below LRA and only 9 mm just above AB) and the absence of the right renal artery and right kidney. Patient was operated through transperitoneal approach and it has been performed thrombectomy and infrarenal aorta replacement with 12 mm Dacron tubular graft.

Keywords: Abdominal aorta; Hypo-plasia; Small aorta syndrome

Introduction

First description of small sized aorta was performed in 1848 by Richard Quain, professor of anatomy and surgery at the University of London [1]. There are several diagnoses that indicate existence of unusually small diameter of the abdominal aorta, but they do not have same meaning. Small aorta syndrome (SAS), introduced by Greenhalgh in 1979, was characterized by presence of aortic diameter smaller than 13.2 mm just below renal arteries, or an aortic diameter smaller than 10.3 mm just above the aortic bifurcation [2]. Middle-aortic syndrome (or mid-aortic syndrome - MAS), which has been described by Sen in 1963, implies the existence of segmental narrowing of the abdominal or distal descending thoracic aorta with accompanied ostial stenosis of its branches [3]. Hypoplastic aorto-iliac syndrome (HAIS) implies high bifurcation of the abdominal aorta, straight course of the iliac arteries without the normal characteristic bowing, acute angle of the aortic bifurcation (AB), aortic diameter of 14 mm or less, and iliac artery diameter of 7 mm or less. Other, less common terms for similar aortic disease are small artery syndrome and small blood vessel syndrome [4,5]. All of these conditions have been linked to a strong tendency toward arterial occlusive disease and consequent thrombosis which is the main reason for necessity of surgical treatment especially in the case of symptomatic patients [6]. Small sized aorta diseases are usually manifested with chronic lower limb ischemia, early developed hypertension and its complication, ischemic stroke, myocardial infarction, heart failure. Developing model of congenital abdominal aortic narrowing has been proposed by William Maycock in 1937 with particular emphasis on failure of the paired dorsal aorta fusion during the fourth week of gestation [7].

Case Report

A 53-year-old woman presented in vascular emergency department with black change at the top of the right toe and blue staining fingers of the both feet (Figure 1). Previously, more than a year she had claudication especially in leg muscle that later dropped below the knee. Trophic changes and blue fingers are present last two to three weeks.

Initially, patient was examined clinically when stated absence of all arterial pulses in both legs. The next step was the ultrasound examination

which confirmed significantly lower levels of ankle-brachial index (ABI; right 0.23; left 0.35) and the absence of flow through the final segment of the abdominal aorta. Also, it was not possible to identify the right renal artery (RRA) or right kidney (RK) with ultrasound examination; therefore we indicated multislice computed tomography (MSCT) angiography. Results of MSCT angiography showed presence a juxtarenal thrombosis of small sized aorta that loses a significant percentage of its diameter just below the left renal artery in comparison with suprarenal aorta (16 mm just above LRA, 11 mm just below LRA and only 9 mm just above AB). MSCT also showed the absence of the RRA and the RK (Figure 2). According to the findings of the examination and the local status in lower extremities it was indicated open surgical treatment.

The patient was operated with trans-peritoneal approach in terms of general endo-tracheal anesthesia. Intraoperatively we have verified small sized aorta with absence of the right kidney and right renal artery (Figure 3). At first, it was performed thrombectomy of infra-renal aorta with Fogarty catheter (size 6 and 7) through transected segment of the aorta below the separation of inferior mesenteric artery which was passable and very hypoplastic. After that we cut-off a 3 cm long segment of aorta and completed the reconstruction with interposition of tubular Dacron 12 mm graft. After surgery, patient had pulses on both feet (ABI right 1.0 and left 1.05), neatly filed postoperative period and was discharged home on postoperative day 7 with a satisfying local status. Blue discoloration of the fingers withdrew, and skin lesions in the right toe began demarcation. Control MSCT angiography examination was made just before the release, with results that shows the complete patency of infra-renal segment of the aorta without residual stenosis.

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Received May 09, 2015; **Accepted** December 03, 2015; **Published** January 02, 2016

Citation: Miroslav JP, Dimitrije S, Velja M, Aleksandar L, Darko B (2016) Chronic Occlusion of Hypoplastic Aorta in a Patient with Right Renal Artery and Right Kidney Agenesis. J Vasc Med Surg 4: 240. doi:10.4172/2329-6925.1000240

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Figure 1: Clinical findings: A before and B after operation. 1. Black discoloration of the right toe (topical gangrene), 2. Blue discoloration (livido), 3. Gangrenous process in regression after operation, 4. Demarcation line, 5. Left toe without blue discoloration 7 days after operation.

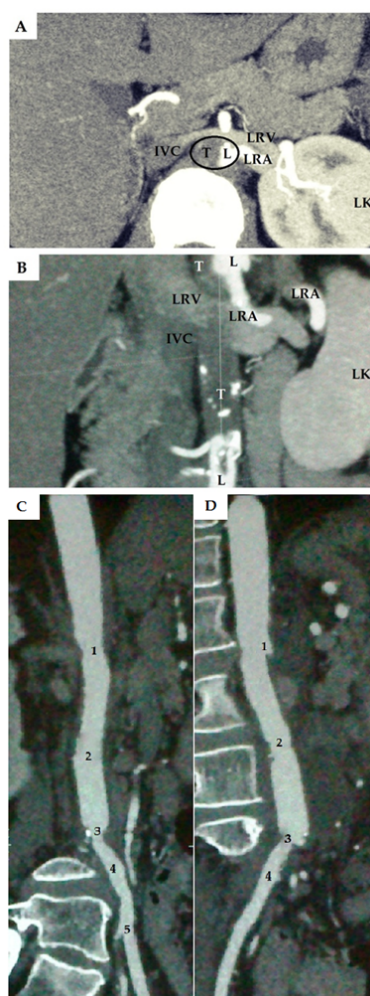


Figure 2: MSCT angiography. A - horizontal transaction in left renal artery separation level, B - sagittal plane transaction, C and D postoperative results. IVC - inferior v. cava, LRA - left renal artery, T - thrombus, L - free lumen, LRV - left renal vein, LK - left kidney; 1. Coeliac trunk separation level. 2. Proximal anastomosis level. 3. Distal anastomosis level. 4. Common (left/right) iliac artery. 5. Left common iliac artery bifurcation.

Discussion

Maycock WA, Arnot RS et al. believes that the basis of small sized aorta development lies in the disturbance of normal genesis of the aortic tube and its occurrence in children [7-12]. It is suggested that the cause

of distal aortic hypoplasia is a congenital one, and is due to excessive fusion of the embryonic dorsal aortae about the 25th day of intra-uterine life. Furthermore, it was observed that patients with reduced diameter infrarenal aorta often have a common lumbar artery tree, which later split into the left and right branch which was found in our case [8]. The external factors that can have a positive impact on the development of small sized aorta are trauma, therapeutic radiation in pregnancy, congenital rubella, hormonal instability, the use of oral contraceptive pills and inflammation. Acquired etiologies were also suggested in fibromuscular dysplasia or Takayasu's arthritis [9]. Small sized aorta has been reported to occur almost exclusively in women and heavy smokers with a relatively early age of onset. Other studies suggest that male-female ratio for SAS is about 1:3. Biochemical analyses usually show lipid disorder; most often familial hypercholesterolemia and familial hypertriglyceridemia are present [3].

A reduced lumen diameter favors the development of atherosclerotic lesions and thrombosis of infra-renal aorta, its bifurcation and common iliac arteries [10]. SAS is manifested as a congenital narrowing of the abdominal aorta below the renal arteries with no changes in visceral branches. Infra-iliac arterial segment is usually hypoplastic and spared from atherosclerotic lesions [11]. In the other case, MAS usually affecting visceral segment of abdominal aorta and outcomes of the visceral branches, but distal infrarenal aortic diameter remain unchanged [12].

Aortic occlusion or thrombosis occurs more frequently in SAS more often than in MAS [9]. Because of these facts described case can be presented more as an unusual form of SAS than MAS, despite the involvement in the level of visceral branches (agenesis of RRA). In accordance to the absence of proper terms for these changes of aorta and its branches we proposed that similar cases of association between SAS and congenital stenosis or agenesis of visceral arteries should be defined as "visceral type of SAS" (vSAS).

Symptoms of hypoplastic aorta usually depend on the involvement of the arterial segment narrowing, presence of visceral origin stenosis and aortic thrombosis. Usually first appears claudication, especially buttock claudication which later descend below the knee. Trophic changes are the end-stage of leg ischemia, particularly present in chronic abdominal aortic occlusion [9]. Renovascular hypertension is the result of narrowing of renal arteries while digestive symptoms are caused by steno-occlusive changes in splanchnic arteries. SAS is not uncommon vascular disorder in Western countries with frequency from 9 to 16% in all patients operated for aorto-iliac occlusive disease (AIOD) [10]. In one study of Japanese authors it has been reported that frequency of SAS in AIOD is around 1.7% (4 cases of SAS in 231 patients with AIOD) [13].

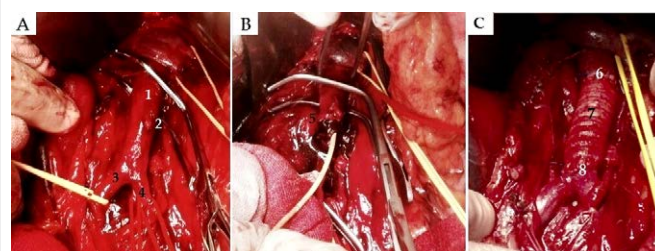


Figure 3: Intraoperative findings. A - postpreparation aortic presentation, B - thrombectomy, C - after Dacron 12mm graft interposition. 1. Infrarenal aorta, 2. Inferior mesenteric artery, 3. Right and 4. Left common iliac artery, 5. Thrombus in aorta, 6. Proximal and 8. Distal line of anastomoses, 7. Dacron 12 mm tubular graft.

The algorithm for determination the correct diagnosis of small sized aorta includes physical examination of the patient, ultrasound examination (Color Duplex Scan or CDS and Ankle-Brachial Index), digital subtraction angiography (DSA), MSCT angiography and magnetic resonance imaging (MRI) angiography [14].

Since patients with SAS had a significantly higher risk of developing AIOD it is strongly important to correct their other risk factors such as smoking and hyperlipdemia to prevent the development of disease consequences early in life. In the presence of symptomatic SAS, reconstructive treatment should be considered. Surgical method of chose for reconstruction in patients with small vessels remains undefined. Most of authorities believe that endarterectomy is not suitable for small sized aorta and iliac vessels. Those who favor bypass techniques advocate the use of end-to-side proximal aortic anastomosis to avoid size discrepancies with the usual prosthetic grafts [15]. In our case we performed thrombectomy and resection of ill infra-renal aorta and reconstruction with 12 mm Dacron graft interposition. Primary indications for this procedure were juxtarenal aortic thrombosis, small sized aorta and relatively healthy aortic bifurcation and iliaco-femoral arterial segment.

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