



Abdominal Aortic Hypoplasia, a Rare Cause of Hypertension

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Authors' contributions

This work was carried out in collaboration between all authors. Author LP designed the case report and wrote the first draft of the manuscript. Author UT designed the figures and contributed to the correction of the draft. Authors BA, DA and SK managed the literature searches. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/BJMMR/2015/17402

Editor(s):

(1) Francesco Angelico, Professor, Department of Public Health and Infectious Diseases, Sapienza University Medical School, Rome, Italy.

Reviewers:

(1) Anonymous, Japan.

(2) Anonymous, Turkey.

Complete Peer review History: <http://www.sciencedomain.org/review-history.php?iid=949&id=12&aid=8637>

Case Study

Received 12th March 2015
Accepted 20th March 2015
Published 31st March 2015

ABSTRACT

Hypoplasia of the abdominal aorta is an exceedingly rare vascular abnormality. Congenital, acquired, infectious and inflammatory etiologies have been described. Hypertension is the most common presenting symptom. Besides hypertension, lower extremity claudication and mesenteric ischemia can be seen. Symptoms typically occur within the first three decades of life. Hypoplasia of the abdominal aorta is considered a life threatening condition as a result of complications associated with severe hypertension.

In this article we present a hypertensive patient with hypertrophic left heart, dilation of the ascending aorta and the aortic root, hypoplastic stenosis of the abdominal aorta, celiac trunc, and the places of origin of superior mesenteric and renal arteries and arch of Riolan (collaterals between the mesenteric arteries). Abdominal aortic hypoplasia is presented as a rare cause of hypertension.

Keywords: Aortic hypoplasia; abdominal aorta; hypertension; arch of riolan; computed tomographic angiography; doppler ultrasonography.

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1. INTRODUCTION

Abdominal aortic hypoplasia is a rare condition which includes tubular hypotrophy of a large abdominal aortic segment, rarely thoracic or both [1,2]. Coarctation of the aorta is defined as significant luminal narrowing of the aorta. This process may affect a short isolated segment of the aorta. Segmental coarctation is most commonly located at the connection site of ductus arteriosus to the thoracic aorta, accounting for 98% of focal lesions.

Abdominal aortic coarctation accounts for less than 2% of the cases, which is extremely rare [3]. The term hypoplasia is used when longer segments of the aorta are stenotic. Aortic hypoplasia has been reported in all thoracic and abdominal segments of the aorta, being more rare than segmental coarctation. Hypoplasia of the aorta is frequently associated with narrowing or hypoplasia of main abdominal aortic branches. The most common clinical manifestation of aortic hypoplasia is severe uncontrolled hypertension in adolescent and young adults [3].

This case report is presented through a single patient with a rare form of hypertension related to hypoplastic narrowing of the abdominal aorta and its main branches.

2. CASE REPORT

A 26-year old man with hypertension and abdominal pain was admitted to the hospital. His history was unremarkable and he had no history of stroke, eye problems, opioid addiction or smoking. The blood pressure was 200/110 mmHg in the right arm. Erythrocyte sedimentation rate, C-reactive protein, antineutrophil cytoplasmic antibodies and gammaglobulins were normal. Echocardiography revealed hypertrophied left atrium and left ventricle, and dilatation of the aortic root and ascending aorta.

The patient was referred to the radiology department for renal artery Doppler ultrasonographic examination. The sizes of the kidneys were normal. Doppler ultrasonographic examination revealed pulsus parvus and tardus flow pattern (low velocity and resistance with high acceleration) in both of the renal arteries. High flow velocities were encountered in the narrowed segments of the celiac trunc and superior mesenteric artery (SMA). The flow

volume increased in the inferior mesenteric artery (IMA) (Fig. 1).

On computed tomography angiography (CTA) hypoplasia of the abdominal aorta with severe narrowing of both renal arteries, accessory renal arteries, superior mesenteric artery and celiac trunc origins were determined. The aortic arch branches and cervical arteries were normal. The wall thickness of the abdominal aorta and its branches were normal. The transvers diameter of ascending aorta above the level of aortic valves was 42 mm. Arcus aorta, descending aorta and abdominal aorta measured 31, 19.8 and 17.2 mm respectively. The diameter of the abdominal aorta between the celiac trunc and IMA was 11 mm. Collateral vessel called arch of Riolan was encountered between SMA and IMA (Fig. 2).

The patient had an operation after the diagnosis. A 14 mm-diameter dacron graft was used to perform an aorto-aortic bypass, extending from descending thoracic aorta to the infrarenal abdominal aorta and two saphen venous grafts were used between the abdominal aorta and left and right renal arteries (Fig 3). The patient's postoperative course was uneventful. He was discharged on antihypertensive medications and called one month later for hypertension follow-up.

3. DISCUSSION

Hypoplasia of the abdominal aorta is extremely rare and few cases have been reported in the literature [1-7]. Less is known about the etiology of aortic hypoplasia. Developmental defects, infection response and inflammation have been proposed as possible mechanisms. It may also be a manifestation of hereditary syndromes such as neurofibromatosis, Williams-Bauren syndrome and hypomelanosis of Ito [4-7]. Most of the authors favor the developmental defect because of frequent association of aortic hypoplasia with other congenital anomalies. Familial form of aortic hypoplasia has been described in a report [4]. Blood vessels start to develop after the 18th day of embryonic folding [8]. As a result of errors in many stages of the development, congenital vascular anomalies such as aortic coarctation and hypoplasia occur. Most of the stenosis are probably related to conditions occurring around the 25th day of fetal development. Meanwhile, two embryonic dorsal aortas translocate towards each other, fuse and the intermediate wall disappears, forming a single vessel. Most aortic stenosis are thought to originate by the

overfusion of the embryonic dorsal aortas or their inability to fuse [8].

Localized segmental narrowings are termed coarctations, but when the narrowing extends over a longer distance the term hypoplasia is preferred (3). Certain viruses, notably rubella, may interfere with cell growth and may have direct cytopathic effects on developing tissue. It is believed that smooth muscle cell organisation and proliferation is inhibited by viruses, thus preventing the normal growth of the aorta. The possible correlation with arteritis such as Takayasu's disease, has given rise to inflammatory etiology. In most cases merely, very little evidence of inflammatory processes exist (5). Neurofibromatosis, fibromuscular displasia, syphilis, tuberculosis and radiation therapy at an early age, Williams-Bauren syndrome (elastin gene mutation) have been all associated with hypoplasia of the abdominal aorta, but no causal relations have been identified [6]. In our patient, all these conditions were excluded. Hypoplasia of the aorta most commonly is seen with hypertension and its sequelae, including fatigue, headaches and epistaxis. Claudication and visceral vessel ischemia can be seen.

Hypertension is most likely the result of global renal hypoperfusion and activation of renin-angiotensin system. Hypertension if left untreated, may result with life-threatening complications, such as stroke and hearth or renal failure [1-3]. In our patient both renal arteries were narrow and severe hypertension was detected. Hypoplasia of the aorta is commonly related to stenosis or hypoplasia of renal arteries (33-81%) and less commonly of celiac trunc (9%), SMA (4%), and IMA (1.3%) [1]. Mesenteric ischemia is uncommon, presumably because of collaterals from an intact IMA. The collateral vessel between the SMA and IMA called 'arch of Riolan' or 'meandering mesenteric artery'. Riolan's arch is believed to be characterized by enlargement of pre-existing collateral vessel in the ground of a major stenosis of the SMA or IMA. Depending on the location of stenosis, the blood flow may be antegrade or retrograde. In proximal SMA occlusion, the Riolan's arch provides collateral flow from the IMA to the SMA territory. In proximal IMA occlusion, it provides collateral flow from the SMA to the IMA territory [9]. Our patient had severe stenosis of the SMA, however, IMA was intact. Arch of Riolan was encountered between SMA and IMA by CTA.

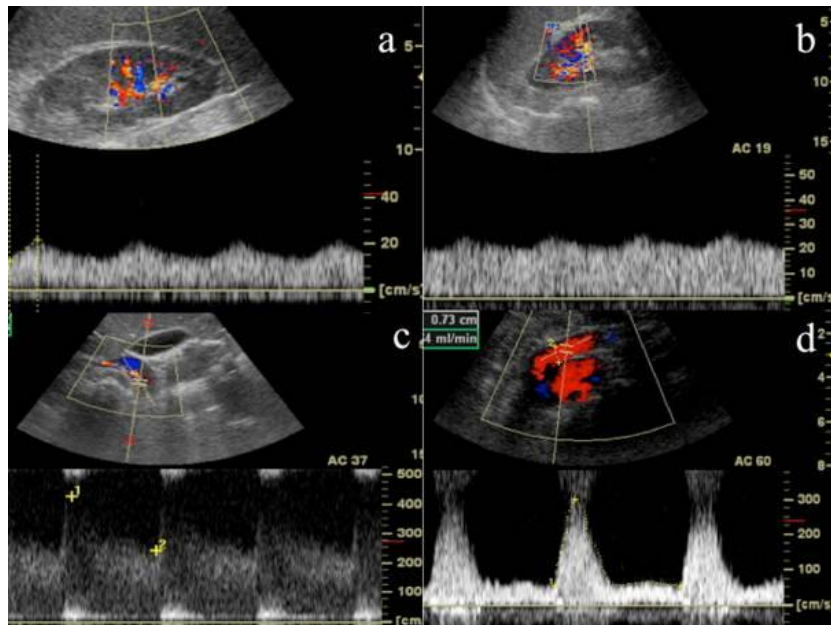


Fig. 1. a-d. (a,b) Doppler ultrasonography shows pulsus parvus and tardus flow pattern in the interlobar renal arteries, (c) high flow velocity in the narrowed segment of the celiac trunc (c), and (d) high flow volume in the inferior mesenteric artery

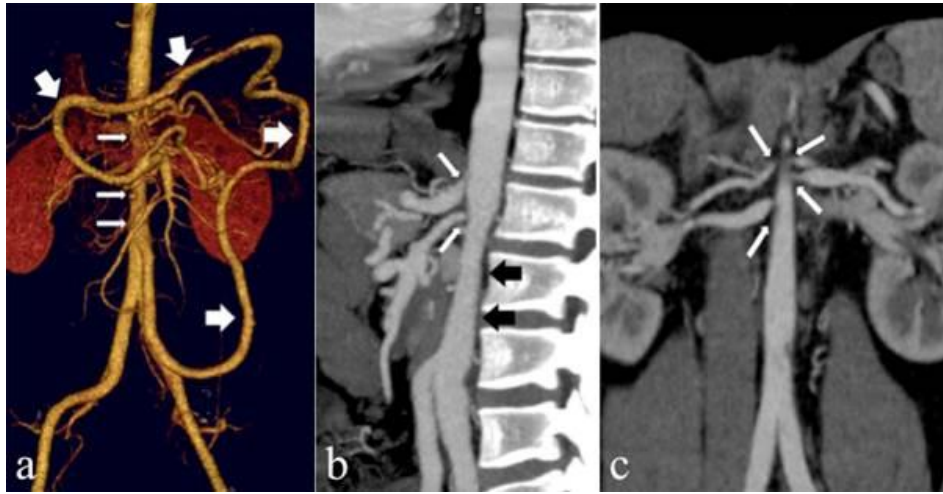


Fig. 2. a-c. (a) 3D volume rendering CT image shows the hypoplastic abdominal aorta (thin arrows) and Riolan arc (thick arrows), (b) sagittal maximum intensity projection (MIP) CT image shows the narrowings of abdominal aorta (black arrows), celiac trunk and superior mesenteric artery origins (White arrows), (c) coronal MIP CT image shows narrowings of native and accessory renal arteries (arrows)

Young patients with hypertension should be evaluated for aortic coarctation or hypoplasia. The diagnosis of aortic hypoplasia can be made by using DSA, CTA, magnetic resonance angiography (MRA) [1-4]. DSA shows the location and extent of the aortic hypoplasia, and reveal associated visceral or renal vessel abnormalities and collateral vessels. Direct measurement of pressure gradient can be performed to access the hemodynamic significance of stenotic lesions and direction of flow within collateral vessels can be obtained. CTA and MRA have been used with increasing frequency to delineate vascular anatomy and diseases. CTA and MRA can provide additional information regarding the presence of any associated inflammatory changes that DSA cannot evaluate. True hypoplasia doesn't show any inflammatory changes [5,6]. In our patient there was no aortic wall thickening on CTA and we can easily exclude any inflammatory process.

Aortic hypoplasia often has a poor long-term prognosis. Most of the patients die due to cardiac failure or cerebrovascular accident at early adulthood. It is treated with surgical revascularization. The length of hypoplastic segment and related renal or visceral artery involvement should be the guide through the operation. End-to-end anastomosis followed by stenotic segment resection or interposition graft placement are the options. For patients with longer segments of hypoplasia, bypass grafting

around the narrowed segment is more appropriate. The bypass graft should originate proximally from uninvolved aorta, usually the descending thoracic aorta. The distal anastomosis is generally made to the infrarenal abdominal aorta. Patients with visceral or renal artery stenosis, may also require by-pass. Every patient with renal artery stenosis of hemodynamic relevance in the presence of hypertension should be treated. Medical control of the hypertension is often difficult, and thus patients usually require renal artery revascularization. Arterial pressure determinations has been found especially useful in assessing the significance of the stenosis and as a guide to surgical therapy. Surgical management should optimize the use of autogenous methods of renal artery reconstruction including saphenous vein aortorenal bypass, splenorenal arterial anastomosis, hepatorenal saphenous vein bypass, and renal autotransplantation [10].

Our patient had an operation after the diagnosis. An aorto-aortic bypass graft was used between the descending thoracic aorta above and infrarenal abdominal aorta below. Two saphenous venous grafts were used between abdominal aorta and left and right renal arteries. The patient's postoperative course was uneventful. He was discharged on antihypertensive medications and called one month later for hypertension follow-up.



Fig. 3. Three dimensional volume rendering CT image shows aortoaortic dacron graft (arrows)

4. CONCLUSION

In conclusion, hypoplasia of the aorta is a rare vascular lesion frequently seen in the first and third decades of life. The usual presenting symptoms are hypertension, claudication and visceral vessel ischemia. Surgical bypass graft is the optimal treatment option. Hypoplasia that involves both the thoracic and abdominal aorta is extremely rare. Young patients with hypertension should be evaluated for aortic coarctation or hypoplasia. We report a case of abdominal aortic hypoplasia with the radiologic findings.

CONSENT

The patient has given his informed consent for this case report to be published. It is applicable. All authors declare that 'written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

It is not applicable for this study.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:

The peer review history for this paper can be accessed here:
<http://www.sciencedomain.org/review-history.php?iid=949&id=12&aid=8637>