Asymmetric Brachial Blood Pressure as an Unusual Presentation of Pseudocoarctation of the Aorta with Giant Aortic Arch Aneurysm

Chih-Wei Lin, Chun-Chi Chen, Tien-En Chen, Ming-Jer Hsieh and I-Chang Hsieh

Coarctation of the aorta, a cause of secondary hypertension, usually presents with asymmetric blood pressure of the upper and lower extremities. However, pseudocoarctation of the aorta does not have pressure gradient cross the stenotic area, so blood pressure is the same over four extremities. We describe a case of pseudocoarctation of the aorta, which presented unusually as a giant aortic arch aneurysm with asymmetric brachial blood pressure in a 52-year-old male. He was hypertensive and had a history of relatively lower blood pressure in his left arm. A diagnosis of pseudocoarctation of the aorta with giant aortic arch aneurysm was made according to the results of chest X-rays, computed tomography, cardiac echocardiography and catheterization. Under cardiopulmonary bypass, the giant aortic arch aneurysm were resected and replaced with an 18-mm woven graft. Based on a MEDLINE search up to 2007, we believe that this is the first reported case of pseudocoarctation of the aorta presenting with asymmetric brachial blood pressure. Physicians should keep in mind that measurement of blood pressure in the four extremities is necessary in all cases of suspected aortic arch disease with hypertension.

Key Words: Aortic aneurysm • Hypertension • Pseudocoarctation of the aorta

INTRODUCTION

Coarctation of the aorta is a congenital abnormality with an incidence of 40 to 50 per 100,000 births. The symptoms are observed during the first year of life, and early surgical intervention is necessary due to the risk of aortic dissection and rupture. Usually, presenting symptoms are associated with proximal systemic hypertension in both arms; however, rarely, adult patients present with aortic wall pathology such as aneurysm formation, aortic dissection or rupture [1]. A true aortic aneurysm is currently defined as a localized dilatation of the aorta, 50 percent over the normal diameter, which includes all three layers of the vessel: intima, media, and adventitia [2]. Thoracic aortic arch aneurysms are less common than aneurysms of the abdominal aorta. Pseudocoarctation of the aorta is a rare condition described as an elongation of the aortic arch with kinking at the level of the ligamentum arteriosum. In this article, we present a patient who had a huge aortic aneurysm between the left common carotid artery (CCA) and left subclavian artery as an etiology of asymmetric brachial blood pressure.

CASE REPORT

A 52-year-old male patient presented with asymptomatically asymmetric blood pressure in both arms. The blood pressure was 167/87 mmHg in the right arm and 124/72 mmHg in the left arm. He denied syncope, angina, hoarseness, claudication, peripheral cyanosis, dysphagia, hemoptysis, stridor or wheezing before admis-
sion. His body height and weight were 178 cm and 72 Kg, respectively. He did not have history of Down syndrome, chest trauma or rheumatic fever. Physical examination was negative for continuous murmur or supraclavicular pulsatile mass. The heart rate was regular at 65/min. The blood pressures were 148/82 mmHg and 146/80 mmHg in the right and left leg, respectively. EKG showed normal sinus rhythm. Hemoglobin was 11.0 g/dL. The early morning cortisol level was 10.4 ug/dL, potassium was 4.0 meq/L and creatinine was 1.4 mg/dL. The neck Doppler showed no stenosis in both carotid arteries. Chest X-ray revealed an opaque shadow with continuity with the aortic arch in the left upper lung zone, which was shown to be a giant thoracic aneurysm about 10 cm in diameter by computed tomography (CT). CT angiography showed the giant aortic arch aneurysm between the left CCA and left subclavian artery. However, no evidence of dissection or intramural hematoma was found. This lesion was tortuously connected to the descending aorta in the isthmic region with transitionally smaller caliber proximally and distally. The descending aorta was tortuous without aneurysm or thrombus. The left subclavian artery originated from the normal caliber of thoracic descending aorta, just distal to the aneurysmal aorta. Transthoracic echocardiography showed a 10.5-cm-in-diameter aortic arch aneurysm with a peak trans-stenotic pressure gradient of 16.2 mmHg via supra-sternal notch view. The patient did not have bicuspid aortic valve, patent ductus arteriosus, ventricular septal defect, atrial septal defect, aortic stenosis or regurgitation. Cardiac catheterization was performed percutaneously from the right femoral vein and radial artery approach due to some difficulty in traversing the aortic arch. Left heart catheterization showed 11.7-cm-in-diameter giant saccular aneurysm of the aortic arch between the left CCA and left subclavian artery (Figure 1a). Selective left subclavian angiography revealed no focal stenosis (Figure 1b). The coronary arteries and aortic root were normal without a systolic pressure gradient cross the stenotic area. The blood pressures of ascending and descending aorta were 153/93 mmHg and 149/87 mmHg, respectively. The abdominal aorta was normal without renal artery stenosis by aortic angiography. There was no step-up gradient of oxygen saturation in the right heart catheterization. The cardiac output was 5730 ml/minute, and stroke volume was 99.3 ml, calculated during left ventriculography. The aneurysm was approached via a left posterior thoracotomy, resected, and then replaced with an 18-mm woven graft. The postoperative course was uneventful. The pathological examination of the resected aneurysmal aortic wall with H&E stain revealed dilated aortic wall with artherosclerosis. After operation, the blood pressure was 123/73 mmHg in the right arm, 122/89 mmHg in the left arm, 138/76 mmHg in the right leg, 140/80 mmHg in the left leg. The blood pressure returned to normal during post-operative hos-

![Figure 1](image-url)  
*Figure 1. a: Aortography showed a 11.7-cm-in-diameter giant aortic aneurysm between the left common carotid artery and left subclavian artery. The pigtail tip was located in the aortic root through the brachiocephalic artery (the arrow). The arrow-head is the origin of the left common carotid artery. The star was located near the ostium of the giant aneurysm. The left subclavian artery and descending aorta were not fully opacified due to most dye having accumulated in the lumen of the giant aneurysm. b: Selective left subclavian angiography revealed no focal stenosis.*
pitalization. The patient was discharged on the 14th post-operation day and has been free of symptoms for 5 months until now.

**DISCUSSION**

Untreated coarctation of the aorta results in high morbidity and mortality from systemic arterial hypertension and associated problems, including myocardial infarction, heart failure, intracranial hemorrhage and aortic rupture. Campbell et al. [3] reported that 50% of untreated patients die before the age of 30, and 90% die before the age of 58. However, pseudocoarctation of the aorta has been described as a “benign” entity warranting no specific therapy. It has been proposed that the embryological cause of pseudocoarctation is a feature of compression of the third through the tenth segments of the dorsal aortic roots and the fourth arch segment [4].

The diagnostic criteria include abnormal chest film, a pressure gradient of less than 25 mmHg between upper and lower extremities, and no evidence of increased collateral circulation (e.g., rib notching) [5]. Aortic angiography provides a definite diagnosis. CT angiography and magnetic resonance imaging (MRI) play a role in the tentative diagnosis. In this case, there was an unusual presentation of pseudocoarctation of the aorta with a giant arch aneurysm between the left CCA and left subclavian artery. Thoracic aortic aneurysms most often result from cystic medial degeneration, which occurs normally with aging and is increased with hypertension. When it occurs in young patients, it is most often due to Marfan syndrome or Ehlers-Danlos syndrome. However, our patient did not have cystic medial degeneration, Marfan syndrome or associated congenital anomalies. Asymmetric brachial blood pressure in thoracic aneurysm is rarely seen. The pathogenesis of lower left brachial pressure in our case is presumably related to the “diverged-flow mechanism” in the giant aneurysm, which was proximal to the left subclavian artery. The evidence was provided during aortography; most dye went into the lumen of the aneurysm, with little opacification of the left subclavian artery and descending aorta. The volume of aneurysm was about 4500 ml by the equation for the size of a spherical ball, but we could not measure the exact fraction of diverged flow into the giant aneurysm during each cardiac beat. The phenomenon means most contrast medium diverged into the aneurysm and resulted in decreased flow to the descending aorta and left subclavian artery. Therefore, lower blood pressure was measured in the post-stenotic vessels. The other less likely possibility is that the lower blood pressure of the left subclavian artery resulted from partial compression by a huge aortic aneurysm, but this mechanism were not supported by angiographic and hemodynamic studies. In a series of 35 adult aortic coarctations, Bouchart et al. [7] encountered only one patient with a descending aortic aneurysm, but none had asymmetric brachial blood pressure. In our case, there was a saccular aortic arch aneurysm with a diameter of 11.4 cm, which resembled a pseudocoarctation of the aorta and was confirmed by catheterization and operation. However, there was no cervical aortic arch noted, which presented spontaneously with pseudocoarctation of the aorta in other series [8].

Patients with thoracic aneurysms are often asymptomatic at the time of diagnosis [6]. However, depending upon the location of aortic aneurysm, chest, back, flank, or abdominal pain can be a presenting symptom. Symptoms are usually caused by compression or distortion of adjacent structures or vessels. Ascending and aortic arch aneurysms can erode into the mediastinum. Such patients will present the following symptoms: hoarseness due to compression of left recurrent laryngeal nerve; hemidiaphragmatic paralysis and elevation due to compression of the phrenic nerve; wheezing, cough, hemoptysis, dyspnea, or recurrent pneumonitis if there is compression or erosion of the tracheobronchial tree; dysphagia due to esophageal compression; or the superior vena cava syndrome from the compression of aortic root aneurysm. In this case, however, asymmetric brachial blood pressure probably resulted from significant diverged-flow volume into the giant aneurysm, which had large lumen and lower compliance than the aortic wall. The other causes of decreased left subclavian blood pressure include trauma, cervical rib compression, agenesis of brachial artery, autoimmune arteritis, compartment syndrome, subclavian steal syndrome, thoracic outlet syndrome, atherosclerotic subclavian stenosis or total occlusion, external compression from lung or mediastinal mass, dissection during catheterization, fibromuscular entrapment, thromboangiitis obliterans and cardiac or non-cardiac thromboemboli. Isolated decrease in left
brachial pulse, as an unusual presentation of pseudocoarctation of the aorta, was presented in this case. Measurement of blood pressure in the four extremities is necessary in all cases of suspected aortic arch disease. The major differences between the post-ductal stenotic aneurysm and coarctation of the aorta were asymmetric brachial blood pressure and no pressure gradient between the upper and lower extremities. However, surgical intervention is usually effective in preventing lethal complication such as aortic dissection or rupture.

REFERENCES

不對稱之上肢血壓為假性主動脈狹窄合併巨大主動脈弓瘤之罕見臨床表現

林智偉  陳俊吉  陳恬恩  謝明哲  謝宜璋
桃園縣  長庚紀念醫院 林口院區 第二心臟內科

主動脈狹窄為次發性高血壓原因之一，通常以不對稱之上下肢血壓表現；然而假性主動脈狹窄則不會於狹窄段有任何壓力差，也就是上下肢血壓相同。我們報告一位 52 歲男性病患以不對稱之上肢血壓為假性主動脈狹窄合併巨大主動脈弓瘤之罕見臨床表現。病患有高血壓之病史，而且左手臂動脈壓遠低於右側。根據胸部 X-光，斷層掃描，心臟超音波及心導管檢查的結果，診斷為假性主動脈狹窄合併巨大主動脈弓瘤。經體外循環手術，巨大主動脈弓瘤被切除並以 18-mm 之主動脈人工血管取代。依據 MEDLINE 直到 2007 年的搜尋，我們相信這是第一個假性主動脈狹窄以不對稱之上肢血壓為臨床表現的病例。對於懷疑有高血壓合併主動脈疾病之患者，臨床醫師應有高度的警覺來测量四肢的血壓。

關鍵詞：主動脈瘤、高血壓、假性主動脈狹窄。