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Case report

Secondary Hypertension due to Isolated Interrupted Aortic Arch in a 60-Year-Old Person – One-Year Follow up

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A B S T R A C T

Interrupted aortic arch (IAA) is a congenital defect characterized by loss of luminal continuity between the ascending and descending aorta[1]. It is a rare malformation with an estimated incidence of perinatally diagnosed cases of 3 per million live births[3]. The condition is considered extremely rare in adults. However, its true prevalence in this population is unknown. We have found 30 case reports of IAA in adults in literature, 5 of whom were older than 50 years. Four of them had type A IAA. Arterial hypertension is a typical co-morbidity. In this report we describe a 60-year-old male patient who had a type A asymptomatic IAA. Although we initially suspected the aortic coarctation, further invasive procedures revealed complete interruption of the aortic arch just distal to the origin of the left subclavian artery. The patient underwent surgical repair, followed by full recovery and near-normalization of blood pressure.

Key words: *interrupted aortic arch, aortic atresia, coarctation of the aorta, congenital heart disease in adults, secondary hypertension, hypertension*

Introduction

Interrupted aortic arch (IAA) is a congenital malformation characterized by loss of luminal continuity between the ascending and descending portions of aorta[1]. IAA was first described by Steidele in 1778. In 1959 Celoria and Patton proposed a classification that is based on the localization of the aortic arch discontinuity – type A, distal to the left subclavian artery; type B, between the left carotid and left subclavian arteries; type C, between the innominate and left carotid arteries[2]. IAA is usually associated with other intracardiac congenital malformations such as ventricular septal defect, patent ductus arteriosus, bicuspid aortic valve, left ventricular outflow tract obstruction or aortopulmonary window. The prognosis of IAA is poor in those who do not undergo surgical repair[3]. Majority (90%) of the affected infants die at a median age of 4 days[4]. Incidence of IAA was assessed in a report published more than 40 years ago and was estimated to be as low as 3 cases *per* million live births[3]. This report was based on the cases diagnosed in the perinatal period. Some patients may develop collateral aortic circulation during gestation and go undiagnosed during early periods of life[5]. It is unknown what proportion of IAA patients survives until adulthood. We have found 30 case reports of IAA in adults in the literature. Five of these individuals were older than 50 years and 4 of them had type A IAA. Arterial hypertension is a typical co-morbidity present in individuals with IAA. Other typical sign is limb swelling with blood pressure

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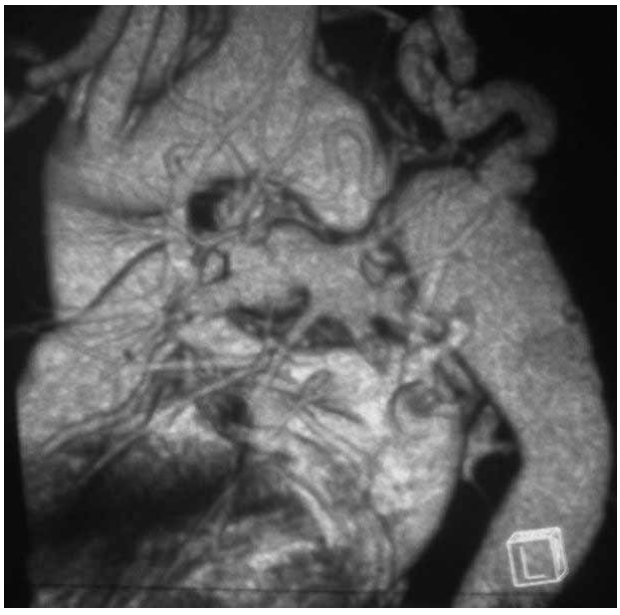

Fig. 1. Multi-slice computed tomography (MSCT) shows a total interruption of the aorta. Collateral vessel inflowing distal of the interruption can be seen.

difference (gradient) between extremities[3]. Recently, we diagnosed a patient with adult form of IAA, who initially was presented with severe systolic and diastolic hypertension.

Case Report

A 60-year-old male patient was admitted to our clinic in March 2008 because of poorly controlled blood pressure (BP). The highest measured BP value in the period immediately preceding hospitalization was 220/140 mmHg. His hypertension was first diagnosed in 2002 and was treated with lizinopril 20 mg/day and hydrochlorothiazid 25 mg/day. At hospital admission the patient did not report any symptoms. His BP was similar on both arms with the highest value 190/100 mmHg (on Day 9 post-admission). His physical examination was remarkable for weak peripheral pulses in both legs. BP was similar in both legs with the mean value of 91/66 mmHg on the right side and 89/63 mmHg on left side. The patient was treated with carvedilol 25 mg/day, lisinopril 40 mg/ day, hydrochlorothiazid 25 mg/day, amlodipin 10 mg and moxonidine 0.2 mg. Chest X-ray indicated aortic configuration of the heart and elongation of the supra-valvular aorta. Trans-thoracic echocardiography showed dilated aortic root (4.3 cm), dilated left atrium (diameter 4.4 cm), and mild diastolic dysfunction of left ventricle. Trans--esophageal echocardiography showed interrupted aortic continuity distal to left subclavian artery (membrane of unknown origin completely obstructing the flow was suspected). The descending aorta displayed no abnormalities. A strong inflow of a collateral vessel that irrigated descending aorta was also seen on these images. A multi--slice computed tomography (MSCT) indicated a total interruption of the aorta due to fibrous membrane 4 cm distally of the origin of left subclavian artery. Blood supply distal to the membrane was maintained through abundant collateral vessels (Figure 1). A contrast-enhanced intraarterial magnetic resonance (MR) aortography scan yielded similar findings. We performed aortography with two catheters, one placed distally in the femoral artery and another one placed proximally in the brachial artery (Figure 2). With this approach a clear aortic discontinuity was confirmed, accompanied by the presence of a collateral vessel in left subclavian area connecting to the distal portion of aorta. These findings were consistent with type A IAA. The patient underwent cardiac surgical procedure that included resection of the fibrous

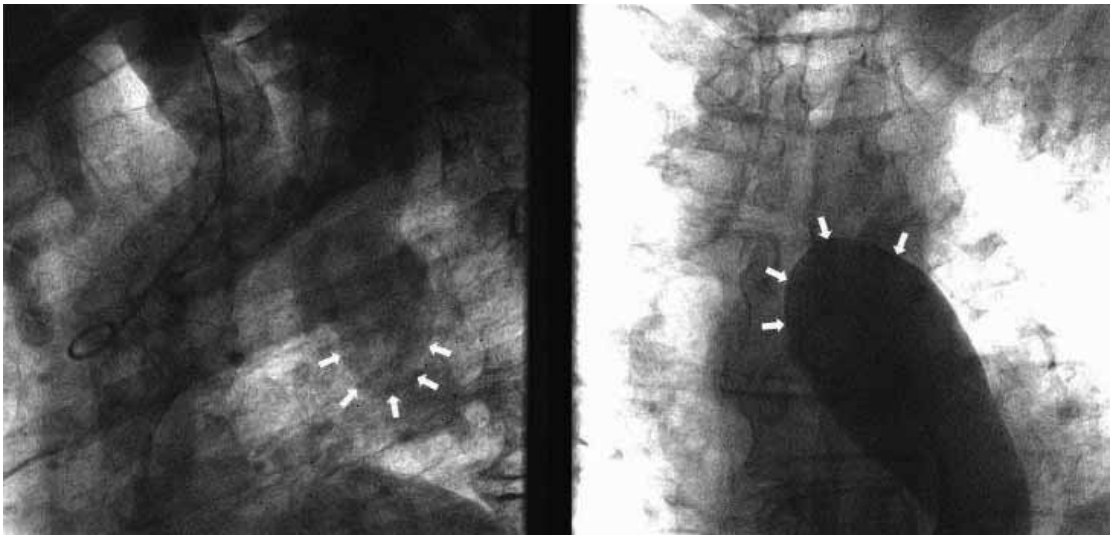

Fig. 2. Aortography with two catheters, one placed proximally in the brachial artery and one placed distally in the femoral artery. A clear aortic discontinuity is seen.

membrane and luminal reconstruction using vascular graft (Intervascular graft 18 mm/15 cm, cat.num. IGW0018--15, Datascope Corp., Montvale, NJ) Intervascular graft length 18 mm. A single-stage extra-anatomic bypass was preformed by a 18 mm Dacron graft between the ascending and descending portions of aorta. Postsurgical recovery was complete and without complications. At the time of release from the hospital his BP was maintained at normal range with substantially reduced number and doses of antihypertensive medications (liznopril 10 mg/day, bisoprolol 2.5 mg/day). At 1-year-follow up his BP was in the normal range with only modest dose of lisinopril (5 mg/day).

Discussion

This patient was diagnosed with isolated type A IAA at the age of 60, being the oldest patient reported in the literature, indicating that some of the individuals with this condition may develop collateral circulation to the extent that no symptoms or signs develop for many years or decades. This puts in question earlier reports on the incidence of IAA as reported by Messner et al.[3] However, we can only speculate about the true frequency of this congenital defect and additional investigations are needed to resolve this question. From a clinical point of view it is important to emphasize that this and some other reports indicate that IAA may be presented with isolated arterial hypertension in those who survive until adulthood. We described in this report almost complete amelioration of, an otherwise poorly controlled, arterial hypertension by a surgical reconstruction that included restoration of the aortic lumen. Throughout the period of postsurgical recovery and at six- and twelve-month-follow-up assessments, BP of the patient was significantly lower as compared to the period before the surgery, despite greatly reduced number and doses of antihypertensive medications. Therefore, IAA may be considered as a cause of secondary hypertension, although, a very rare one.

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SEKUNDARNA HIPERTENZIJA KAO POSLJEDICA PREKINUTOG LUKA AORTE KOD 60-GODIŠNJEG PACIJENTA – JEDNOGODIŠNJE PRAĆENJE

SAŽETAK

Prekinuti luk aorte (PLA) je prirođena mana kod koje je kontinuitet lumena između uzlazne i silazne aorte prekinut. To je rijetka malformacija s procijenjenom, perinatalno dijagnosticiranom, incidencijom 3 na milijun živorođenih. Smatra se da je preknuti luk aorte iznimno rijedak kod odraslih. Međutim, stvarna prevalencija prekinutog luka aorte u našoj populaciji je nepoznata. U literaturi smo pronašli 30 slučajeva prekinutog luka aorte kod odraslih, od kojih je 5 bilo starije od 50 godina. Četvoro od njih imalo je prekinuti luk aorte tip A. Arterijska hipertenzija je, kod takvih bolesnika, tipični komorbiditet. U ovom prikazu opisujemo 60-godišnjeg muškarca s asimptomatskim prekinutim lukom aorte. Iako je u početku postavljena sumnja na koarktaciju aorte, daljnji invazivni postupci su otkrili potpuni prekid luka aorte distalno od polazišta lijeve podključne arterije. Pacijent je zatim bio podvrgnut reparativnom kirurškom zahvatu, nakon kojeg je slijedio puni oporavak i gotovo potpuna normalizacija krvnog tlaka.