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Case Report

Secondary Hypertension due to Abdominal Coarctation in Children: How Do We Identify and Manage It?

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ABSTRACT

Background: Coarctation of the abdominal aorta (AoA) is a very uncommon condition, accounting for 0.5-2 percent of all aortic stenosis cases. We present an 11-year-old child who has been diagnosed with abdominal aorta coarctation

Case Illustration: An 11-years old boy complained about dizziness 1.5 years ago with hypertension. He underwent hypertension treatment for 1.5 years with a pediatrician, but there has been no improvement in his blood pressure. His current blood pressure was 150/95 mmHg in upper extremities and 120/80 mmHg in lower extremities with three antihypertensives drugs. We found bruit in the abdomen and decreased pulse in the lower extremities. An echocardiogram shows no congenital defect. The first was on suspicion of renal stenosis and underwent ultrasonography of the abdomen but was not conclusive. Computed Tomography Angiographic (CTA) showed severe stenosis in the abdominal aorta on level thoracal 11-12. Aortogram shows significant stenosis in the abdominal aorta with a pressure gradient was 47 mmHg. He underwent percutaneous transluminal angioplasty with Bare Metal stent (BMS) self-stent implantation. At two months of follow-up, his blood pressure target was achieved with a minimal dose of oral antihypertensive drugs.

Discussion: Patients with young age and persistent hypertension should be more careful in diagnosis. A bruit in the abdomen with a weak pulse in the lower extremity raises suspicion of Coarctation of the Abdominal Aorta. An angioplasty procedure, either with or without a stent or surgery, might be used as the primary therapy.

Conclusion: Causative treatment of CoA such as surgery or stent implantation is recommended to improve outcome.

1. Introduction

Hypertension at a young age is not common. The most common etiologies in children, in whom 70 to 85 percent of cases of hypertension have a secondary cause. The primary cause is renal parenchymal disease, coarctation of the aorta, and aldosteronism. For those diagnosed with a "coarctation," the phrase refers to an instantaneous constriction of one or more of the major arteries in one's body directly next to the ductus arteriosus. This results in an increased pressure gradient. The constriction of the aorta results in differential blood pressure in the arm and leg and is found as hypertension in physical examination. CoA may be found in several places.[1,2]

An abdominal aortic coarctation occurs when the abdominal or distal descending thoracic aorta is constricted by congenital or acquired causes. "Middle aortic syndrome" or "mid-aortic dysplastic syndrome" refers to coarctation of the descending thoracic or abdominal aorta. Major arterial branches and visceral arteries may be involved in this condition (renal, superior mesenteric, or hepatic arteries). Abdominal aortic coarctation is a rare condition, affecting just 0.5–2 percent of those suffering from CoA3. Coarctation may occur as a primary (native) occurrence or as a recurring event due to a prior repair.[4]

Coarctation left untreated results in morbidity and early death via hypertension, congestive cardiac failure, stroke, aortic rupture, and progressive cardiac and renal dysfunction. Without correction till age 14, the survival percentage after twenty years is 79%. The life expectancy of mortality is between 33 and 35 years, and 90% of individuals with untreated coarctation die by the sixth decade. It is estimated that three cases of the condition occur for every 10,000 live births. About 5% to 8% of people with aortic coarctation have some congenital cardiac diseases, such as mitral valve prolapse or an accompanying bicuspid aortic valve. In the general population, men outnumber women.[5,6] Coarctation of the aorta is often underdiagnosed

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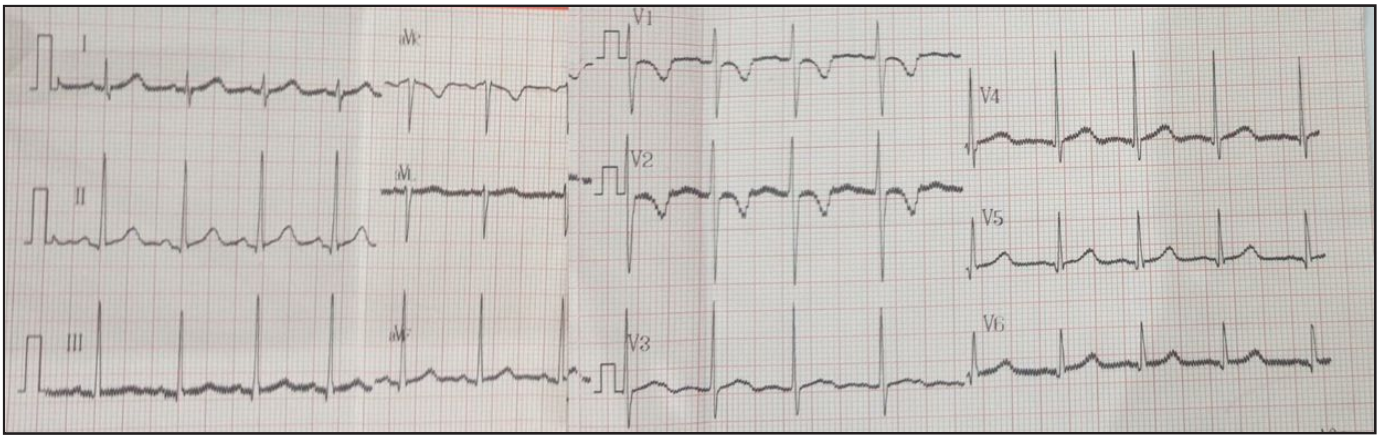


Figure 1 . Electrocardiogram recording showed normal sinus rhythm with a heart rate of 100 bpm.

because of is usually asymptomatic. The major symptoms are general symptoms with dizziness and high blood pressure similar to other underlying diseases such as renal artery stenosis and aldosteronism.

We report a case of Coarctation of the abdominal aorta treated with percutaneous transluminal angioplasty and stent placement

2. Case Illustration

We discuss a case of an 11-year-old boy who was sent to Saiful Anwar General Hospital with symptoms of occasional dizziness in the last 1.5 years and elevated blood pressure. The resting blood pressure in his left arm was 150/90 mmHg, 145/90 mmHg in his right arm, and 120/80 mmHg in both legs. He is already using three antihypertensive medications. At rest, the patient was asymptomatic but noted occasional dizziness during effort. He had no recent febrile episodes, no congenital abnormalities, no neurofibromatosis or cafe-au-lait spots, no claudication, and no mesenteric insufficiency. Clinical examination revealed that the patient was growing normally (125 cm tall and 23 kg in weight), with clear lung examination and normal respiration sounds.

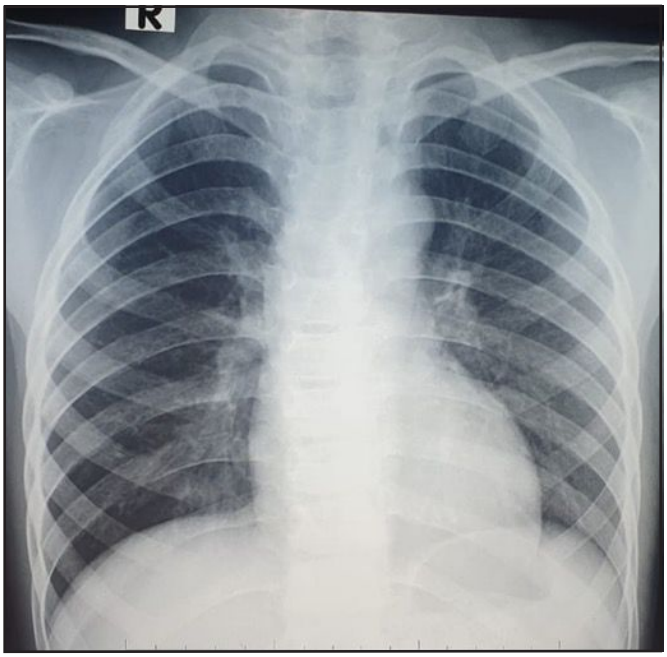


Figure 2. Chest X-ray showed cor and pulmo within normal limit.

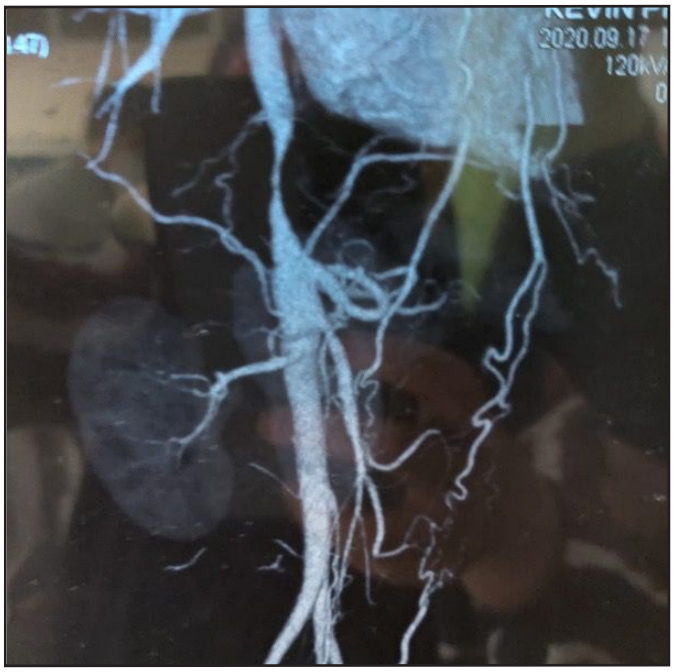
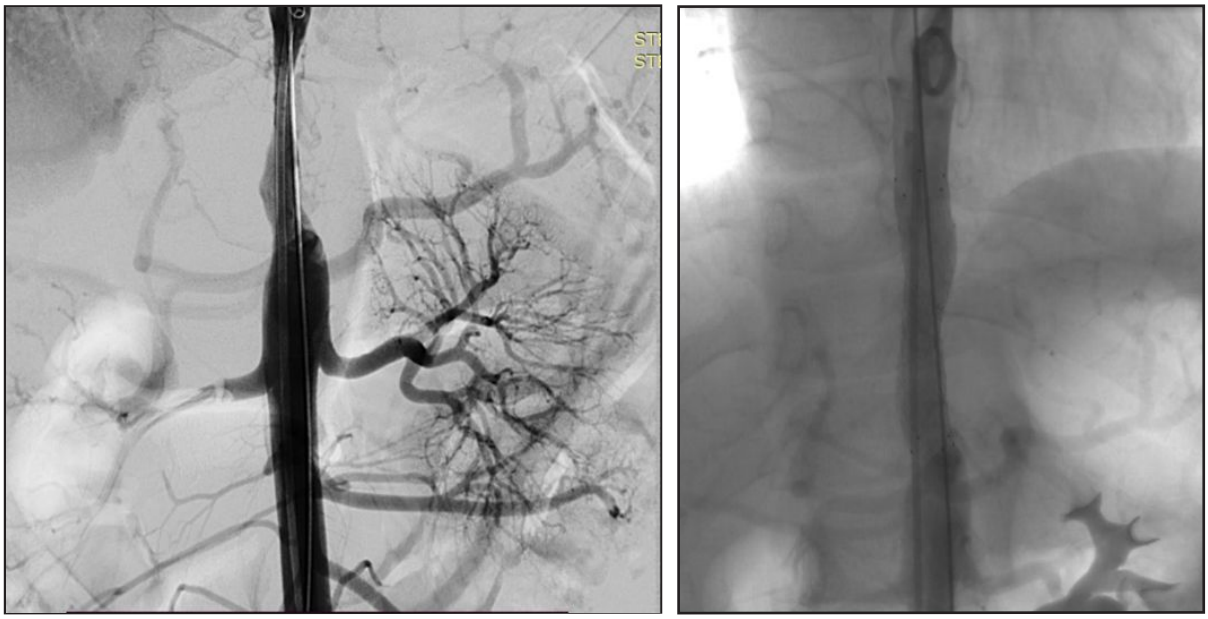


Figure 3 . CT Scan of abdominal aorta show stenosis at level T11-12 with the narrowest segment diameter was 3 mm

A cardiovascular examination revealed a heart rate of 100 beats per minute, no cardiac dilatation, and no murmur. Both femoral arteries had a weak pulse. We also found a bruit in the abdomen examination. As for the rest of the clinical assessment, it was considered normal.

A standard 12 leads electrocardiogram (ECG) was normal at rest, in sinus rhythm, and with no signs of left ventricular hypertrophy or strain. Transthoracic echocardiography showed normal systolic function with no congenital defect found. Abdominal ultrasonography was inconclusive with suspicion of stenosis artery renal. An aortic angio-CT scan was performed on the patient to provide a more accurate representation of aorta anatomy and its branches. It verified coarctation in the abdominal suprarenal aorta with a minimum lumen diameter of 3 mm. The epigastric artery was dilated with collateral circulation.

An aortogram revealed a significant stenosis >70% compared to distal the abdominal aorta. The length of aortic narrowing was 50-60 mm, before the celiac branch. The pressure gradient pre stenotic and post stenotic was 47 mmHg. He underwent percutaneous transluminal angioplasty and performed BMS self-stent implantation 14x60x120 cm.



(A) (B)

Figure 4a. The abdominal aortogram showed >70% stenosis of the abdominal aorta proximal to the celiac branch. 4b Abdominal aorta after stenting

After the procedure, there was no specific treatment; we tapered down the dose of antihypertensive drugs with monitoring blood pressure periodically. After six months of follow-up, the blood pressure returned to normal without using antihypertensive medication. Additionally, he is gaining weight gradually

3. Discussion

Secondary hypertension is a type of hypertension with an underlying disease. Secondary hypertension prevalence and its etiology vary according to age. In children and adolescents, the primary cause of secondary hypertension is renal parenchymal disease and coarctation of the aorta. Coarctation of the aorta has two forms: pre-ductal and post-ductal. Coarctation of the descending thoracic and abdominal aorta, also termed atypical coarctation of the aorta (atypical CoA) or middle aortic syndrome (MAS), is a rare condition with an estimated relative incidence of 1:62.500 unselected mortality with a broad spectrum of morphologic findings. Depending on the origin and degree of stenosis, MAS may manifest in infancy but is commonly diagnosed in teenagers or adults. Coarctation of aorta boys is more prevalent, but the prevalence varies in the case of abdominal coarctation when the lesion occurs in a similar proportion in both genders. Morphologically, coarctation may appear as luminal narrowing of the aorta spanning several centimeters in length or as a sharp, web-like constriction.[6,7,8]

Aortic coarctation results in varying degrees of luminal compromise. The truncus arteriosus terminates in the aortic sac, where the paired aortic arch arteries branch off and terminate in one of the paired dorsal aortas. The abdominal aorta develops during the fourth week of gestation by the union of the twin embryonic dorsal aortas. Congenital coarctation has been attributed to either incomplete fusion or fusion of the paired (primitive) embryonic dorsal aortas. A patent ductus arteriosus may be present in 50% of individuals with preductal coarctation, although its prevalence declines significantly with postductal aortic coarctation. Stenosis of the proximal renal artery occurs in up to 80% of cases, while stenosis of the celiac and superior mesenteric (SMA) arteries occurs in at least 25% of cases.[9]

The intensity and degree of this constriction are the primary causes of symptoms such as hypertension, intermittent claudication, and abdominal discomfort. MAS is often associated with significant arterial hypertension in children, which may result in major consequences such as congestive heart failure, left ventricular hypertrophy, and cerebrovascular accidents. The symptoms vary depending on the degree and location of vessel stenosis. The majority of patients exhibit signs of severe renovascular hypertension, such as absent femoral pulses, abdominal bruit, and claudication of the lower limbs. Patients with long-standing refractory hypertension may present with hypertensive encephalopathy and retinopathy symptoms.[10] In our case, this patient has known hypertension since 1.5 years ago with dizziness. He was controlled by the pediatrician and got three antihypertensive drugs. Blood pressure in his arm and leg varied greatly, with weaker femoral pulses and abdominal bruit. We didn't find abnormalities in heart and lung examination. These clinical findings support a suspicion about stenosis in the abdominal aorta.

There have been many documented subtypes of abdominal aortic coarctation, depending on the location of the lesion: inter-renal, suprarenal, infrarenal, and diffuse. In one of the biggest recorded series, stenosis occurred most often in the inter-renal portion of the aorta (52%), followed by the infrarenal segment (25%). In 13%, the whole aorta was diffusely implicated, whereas 11% impacted the suprarenal area. About 81% percent of patients had stenosis of the renal arteries, whereas 22% had the splanchnic occlusive disease. According to the ACHD 2018 Guidelines, initial and follow-up aortic imaging with CMR or CTA is indicated in people with aortic coarctation (Class I, Level of Evidence B).[9,11,12] An abdominal CT scan and aortography on this patient show that he has suprarenal stenosis.

The pathophysiology of CoA is still largely unclear. Even though inherited and acquired illnesses are connected to a few cases, most are unrelated. Neurofibromatosis (von Recklinghausen disease) type I, Alagille's syndrome, or Williams' syndrome may be genetically linked to MAS. The onset of Takayasu's arteritis or an infection in the intrauterine might also be a contributing factor (particularly rubella)

but remain unclear. If stenosis of the aorta related to inflammation such as Takayasu disease, its often be systemic stenosis. The stenosis will occur not only in one place but also in other arteries. [10,13,4] In this case, there was no sign or symptom of inflammation such as fever, joint pain, and elevated inflammation marker. We also didn't find any stenosis in other arteries.

A delayed repair beyond early childhood does not prevent the persistence or late recurrence of systemic hypertension; it is best to correct coarctation in infancy or childhood to avoid the formation of chronic systemic hypertension. Untreated abdominal aortic coarctation is associated with increased mortality and morbidity. Percutaneous transluminal angioplasty, surgical placement of a stent, or both are possible treatments. In the 1980s, balloon angioplasty was used to execute the first transcatheter aortic coarctation procedures, and intravascular stent therapy became more widely accepted in the 1990s. Children and adults with native aortic coarctation are being treated with the transcatheter method in numerous hospitals.[14-15]

Infant surgery has a mortality rate of fewer than one percent and is considered the treatment of choice. A greater re-intervention rate due to restenosis and an increased risk of aortic rupture and aneurysm development than surgical treatment, balloon angioplasty is not indicated in newborns and children. There are several variables to consider, including the patient's age, where the stenosis is located, how long the segment is, and how much of the visceral vessels are affected. Because of the higher risk of problems after stent insertion in tiny children who have not yet grown up, surgery prefers stent repair. Surgical repair has a greater mortality risk, although stent insertion still has a high reintervention rate.[10,14,16] In this patient, we decided to do endovascular treatment with stent implantation after discussing with inter multidiscipline team. A balloon angioplasty procedure was performed with stent implantation. Based on the guideline in native CoA, there was an indication for stent implantation in a patient with a pressure gradient >20 mm, with systemic hypertension (class IIa).[11] Hence in this patient, the pressure gradient was 47 mmHg with significant stenosis and systemic hypertension. After stent implantation, the pressure gradient becomes 10 mmHg. There was no difference in the incidence of coarctation and pseudoaneurysm development between patients who had stent implantation using a bare-metal stent and those who used a covered one after 31 months of follow-up in a randomized study.[16]

A further intervention or antihypertensive medication is sometimes required following repair in circumstances where persistent hypertension is widespread. As children and teenagers develop, the fibrosis caused by instrumentation or the visible lack of development predicted from grafts or stents may cause restenosis and decreased flow.[17] This patient population's life expectancy necessitates that long-term improvement and monitoring be a key but generally unknown part of therapy. In this patient, the was clinically improved six months after repair. His blood pressure was within normal limits without any antihypertensive drugs. But, the possibility of restenosis and reintervention in this patient still cannot be ruled out. Periodic imaging is necessary to monitor complications.[18,19]

4. Conclusion

Coarctation of the abdominal aorta is uncommon, and the cause is as yet unclear. Children or adolescents with systemic hypertension and different blood pressure in extremities should increase our awareness about the possibility of coarctation—the earlier diagnosis and treatment in the patient with coarctation related to a better outcome. Causative treatment of CoA such as surgery or stent implantation is recommended to improve outcome. After repair, monitoring of complications should be done periodically. [20,21]

5. Declarations

5.1. Ethics Approval and Consent to participate
This study was approved by local Institutional Review Board, and all participants have provided written informed consent prior to involvement in the study.

5.2. Consent for publication
Not applicable.

5.3. Availability of data and materials
Data used in our study were presented in the main text.

5.4. Competing interests
Not applicable.

5.5. Funding source
Not applicable.

5.6. Authors contributions
Idea/concept: OPS. Design: OPS, NK. Control/supervision: NK, AF, HM. Literature search: OPS, NK. Data extraction: OPS, NK. Statistical analysis: OPS, NK. Results interpretation: OPS, NK. Critical review/discussion: NK, AF, HM. Writing the article: OPS. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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