Paediatric middle aortic syndrome with endovascular treatment in East Malaysia

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SUMMARY

Middle-aortic syndrome is a surgically curable cause of childhood hypertension. Open surgery is traditionally offered but with the advance of medical technology, endovascular approached is available in many country. Failure to control BP in open surgery is as low as 4.1% compares to 13% in endovascular approaches. However, mortality is 4% in open surgery almost 2 times higher than 2.3% in endovascular approach. This article presents a case of 10 years old child treated successfully without complication with endovascular balloon dilatation, as a first case of such disease in East Malaysia.

INTRODUCTION

Childhood hypertension with middle-aortic syndrome is a curable hypertension with either open surgery or endovascular surgery. Endovascular surgery resembles one of the recent advancement in Vascular Surgery specialty, which is not available in most part of Malaysia. Now vascular surgery service had started in East Malaysia and the first case of the endovascular treatment for middle aortic syndrome was done in East Malaysia with the immediate success without any major complication.

CASE REPORT

A 10-year-old boy was referred to the vascular surgery unit of our center for consideration of operative management on middle-aortic syndrome with the diagnosis of Takayasu's disease. He initially presented with headache and vomiting. Subsequently, he presentedto casualty with multiple episodes of seizure which necessitated intubation for airway protection. His SBP was 156-161mmHg and DBP of 110-114mmHg, which required IV infusion of labetalol for blood pressure control. During the stay in the tertiary paediatrics hospital, multiple imaging studies were done including Magnetic Resonance Angiogram of brain, carotid arteries and renal arteries. The imaging studies reported as suggestive of Takayasu's arteritis involving abdominal aorta and left renal artery with multifocal cerebral infarct. There was long segment irregularity with mild narrowing of upper abdominal aorta. There was a short segment (1cm in length) of >70% narrowing involving abdominal aorta just below renal arteries. The origin of left renal artery is stenosedbut right renal artery is normal. Clinically there is audible abdominal aorta bruit. He was diagnosed by paediatric nephrologist after discussion with the radiologist as Takayasu Arteritis involving abdominal aorta and left renal artery Type 3. The diagnosis is by American College of Rheumatology 1990 criteria with the presence of the 3 criteria: Age at onset of disease less than 40 years, bruit over abdominal aorta and arteriographic abnormality.He was thus referred to interventional radiologist, which renal angiogram and renal angioplasty were done.

Upon seeing him in our outpatient clinic in 1 month after the intervention, his BP was 100-133/54-80 with the 95th centile at 120/80, despite on tablet amlodipine 5mg od and tablet atenolol 50mg BD. He was on tablet prednisolone 45mg OD as well. Physical examination revealed no absent, weak or delayed in the limb pulses. Tablet aspirin 75mg OD was started and a conservative management was offered initially. He was under vascular surgery three monthly follow up. Over the course of one and a half year, he was prescribed with 3 anti-hypertensive medications including hydrochlorothiazide, atenolol, and amlodipine.

As the trend of needing more anti-hypertensive medications for the BP control, our team decided for a percutaneous aortoplasty for him. Vascular access was obtained via the right common femoral artery and aortogram performed. There was significant stenosis of the abdominal aorta just distal to the right renal artery extending to distal to the left renal artery. Aortoplasty was performed with Boston Scientific Mustang™ angioplasty balloon of diameter 6mm and subsequently 8mm. The procedure was successful from completion aortogram and there was no immediate complication noted. He was discharged on the next day.

Three months post-procedure review in the clinic showed that the BP control is better with reduction of 3 anti-hypertensive medications to one medication.

DISCUSSION

Middle-aortic syndrome (MAS) is a term first used by Sen et. al.(1963) to describe the stenosis of aorta in between the arch and the bifurcation of aorta. It is used for obstructive lesions of the mid-aorta without specifying the aetiology.[1] It is an important condition as it represents one of the curable causes of childhood hypertension. There is fibromuscular hyperplasia that involves the abdominal aorta including the renal and visceral branches.

For most of the MAS cases, the aetiology is idiopathic, where the pathogenesis of the disease mainly remained unknown. There had been theory explaining MAS, which proposed that

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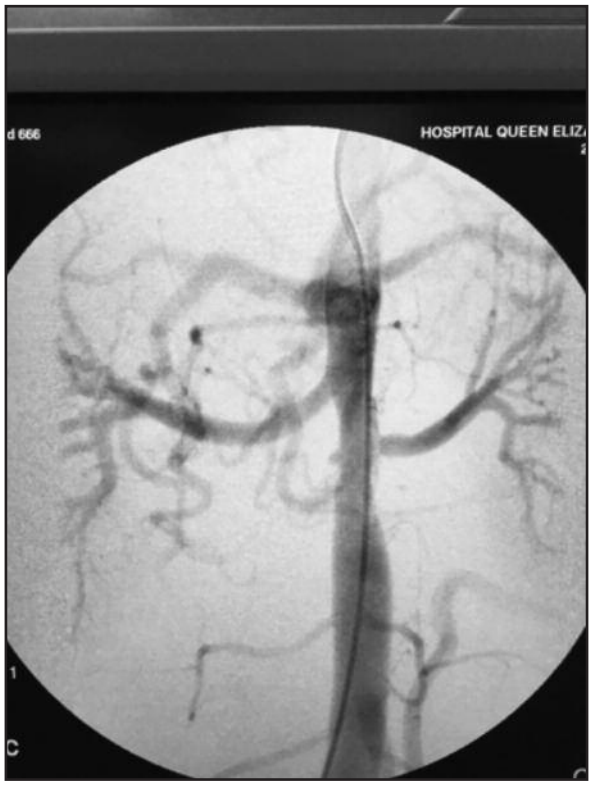

Fig. 1: Pre-dilatation angiogram: the stenosis and the collateral branches are seen.

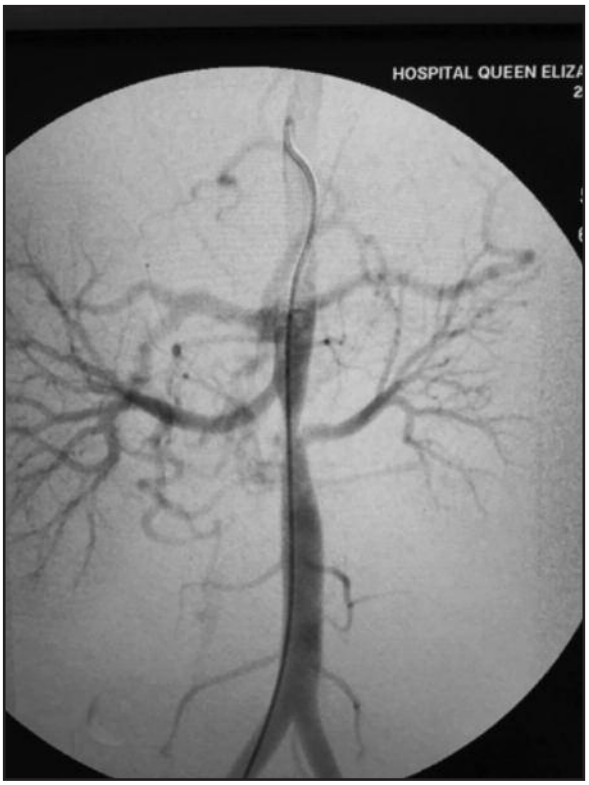

Fig. 2: Angiogram after the dilation showed a good size of aorta.

there is embryological defect followed by failure of normal fusion of the 2 dorsal aortas.[2] MAS may have a genetic cause, such as neurofibromatosis(von Recklinghausen disease) type I,Alagille's syndrome, or Williams' syndrome. It can also be due to acquired inflammatory diseasessuch as Takayasu's arteritis or intrauterine infection (especially Rubella).[3]

Takayasu's disease is a disease with chronic inflammatory arteriopathy of unknown aetiology leading to stenotic and ectatic changes, including occlusion of main branches of aorta.[4] The anatomic distribution of disease is most commonly classified using the 1994 Tokyo International Conference Classification of Takayasu's Arteritis, and in this patient is type III as evidenced by long segment upper abdominal aorta involvement, renal artery involvement and the stenosis at aorta distal to renal arteries bifurcation. Usually if left untreated, Takayasu's disease would lead to vascular complications and patient might even die of the disease. Untreated Takayasu's disease commonly ended up with cardiovascular problems, such as aneurysm formation, concentric arterial wall fibrosis, and thrombotic complications. The mortality of Takayasu's disease children in the recent cohort study in UK published in 2015 by Despina Eleftheriou was reported as high as 27%.[5] The fibrosis of aorta in Takayasu's disease can happen in anywhere in the aorta. When the fibrosis is in the middle of aorta, it is known as MAS.

The typical presentation of children with MAS is severe arterial hypertension, which can be complicated with coronary artery disease, congestive heart failure, left ventricular hypertrophy and cerebrovascular accidents. The systematic review done by Rumman et al. (2015), showed that 86.5% of the MAS children presented with hypertension. The other symptoms may vary according to the degree and location of vessel stenosis. Clinical findings other than hypertension followed by frequency includes systolic murmur, abdominal bruit, diminished femoral pulse, absent femoral pulse, neurologic deficit, positive Mantoux, facial palsy, and seizures. The abdominal aortic stenosis is reported in 97% of cases, where as only 3% of cases have distal thoracic aortic stenosis. In the abdominal aorta stenosis, the most common reported anatomical site is suprarenal, followed by suprarenal to infrarenal stenosis, and then infrarenal involvement. The extra-aortic vessel involvement is not uncommon, as 66% of renal artery stenosis cases are noted at initial presentation. Other involvement may include retinal, cerebrovascular, carotid and subclavian.[3]

The management of MAS is aimed at controlling blood pressure, preventing long term complications related to hypertension, and preserving end-organ function. To achieve the above-mentioned target, there can be pharmacological, endovascular and surgical treatments. Oral anti-hypertensives can be used with a satisfactory result in some cases, which if treated conservatively, 14% became normotensive and 36% had an improvement in BP with drug therapy. In others, their hypertensionsare refractory to medication and needed operative management. [3]

Endovascular management of MAS can be done via percutaneous transluminal aortoplasty with or without

stenting. The intervention may include balloon dilatation alone or involve stenting. Percutaneous interventions were reported as acutely successful in decreasing the degree of stenosis, and low re-stenosis rate after intervention especially the cases with intervened renal arteries.[1] The patients who were treated with endovascular approach can achieve good results with only 13.3% of them BP uncontrolled. The mortality due to endovascular intervention is low about 2.3%. However, the endovascular intervention is not without problem as the complications rate was reported about 13%. The reported complications include vascular tears and development of aneurysms at angioplasty site. There is also a high incidence of restenosis and reintervention.[3]

On the other hand, surgical interventions are usually successful in relieving obstruction and resulted in a longer freedom from re-intervention and fewer complications.1 The surgical intervention may be consists of aortic patch plasty, aorto-aortic bypass, thoraco-abdominal bypass and reconstruction patch graft. The rate of uncontrolled BP for the group after surgical intervention is about 4.1%, which is better than endovascular intervention. Nevertheless, the mortality rate for surgical intervention is 4% which is higher than endovascular approach. Thus the risk and benefit must be weighed before the decision for intervention.[3]

In our patient, so far he has only one endovascular intervention and it is the first time the middle-aortic syndrome managed by this approach. The intervention is considered successful as the anti-hypertensive medication able to reduce from three medications to one medication. There is no standard follow up protocol for the endovascular approach. However, he would need to have a long term follow up by vascular surgery and our paediatric colleague to monitor the BP control and radiological surveillance, as to decide if any further intervention is needed.

CONCLUSION

The treatment of MAS necessitates a multi-discipline team management, which includes timely diagnosis of the disease, optimal medical treatment and early vascular surgery treatment. With the advances of technology in endovascular field, a less invasive approach with a higher success rate can be reached with endovascular intervention.

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