



## Middle Aortic Syndrome (MAS) –A Rare Cause Of Hypertension In A Child with Rheumatic Heart Disease

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### Abstract

*The estimated prevalence of hypertension (HTN) in paediatric age group is about 2.5%. These are mostly due to secondary causes, of which renovascular causes constitute (60-70%). Middle aortic syndrome (MAS) is a rare cause of HTN.*

*Here we present a case of 10 year old girl child, a known case of rheumatic heart disease (RHD) under treatment for carditis. She was on steroids and penicillin. She was admitted in the emergency paediatric department with complains of continued fever for 7 days. On examination the blood pressure was 180/110mm in the right arm supine position and it was thought to be steroid induced HTN. Detailed clinical examination revealed absent B/L femoral pulsation. On further investigation by MR Aortogram, a diagnosis of MAS was confirmed. MAS is a rare disease with only 200 reported cases and this is the first case report citing co-presentation of RHD and MAS.*

**Keywords:** Renovascular cause, carditis, MR Aortogram, co-presentation of RHD and MAS

### Introduction

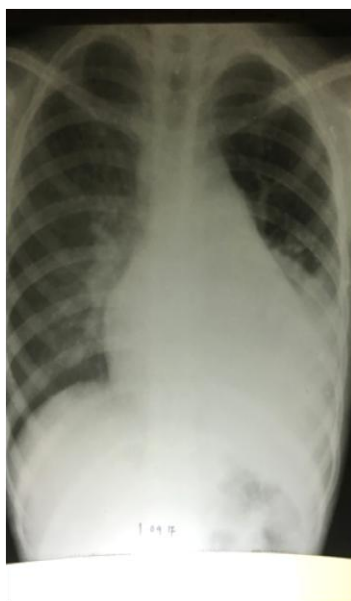
The term MAS was first used by Sen et al to describe narrowing of the sub-isthmal aorta, in distinction to the Takayasu's type aortic arch and Leriche type aortic bifurcation obliterative disease<sup>[1]</sup>. It typically leads to stenosis of the lower thoracic and upper abdominal aorta with or without involvement of renal arteries. MAS is unusual cause of arterial HTN in upper extremity secondary either to congenital anomaly in development of aorta or due to one of the several acquired conditions like NF, retroperitoneal fibrosis, fibromuscular dysplasia, MPS and Takayasu's arteritis. This may lead to narrowing of abdominal aorta and other vessels<sup>[2]</sup>

### Case report

A 10 year old girl was admitted in emergency paediatric department of MKCG with complains of fever for 7 days. She is a known case of RHD and was admitted 1 month back for carditis, for which she was taking steroid and penicillin. Her clinical examination revealed normal radial, very feeble femoral and other limb pulses. The BP was 180/110mmHg in both upper limbs in supine position and lower limb BP was 40/10 mmHg. There was a grade 3 pansystolic murmur best heard at apex and radiating all over the chest. The breath sounds were grossly reduced on the left side. The diagnosis of pneumonia (left) with/without collection was made.

Chest radiography revealed a non homogenous opacity on the left side with evidence of cardiomegaly. No evidence of mediastinal shift was there (Fig 1).Pleural tap was a dry. She was put on IV antibiotics (ceftriaxone and ampiclox). Six days later, she became afebrile and breath sounds on the left side improved. The ECHO cardiography showed RHD with MR. Regarding hypertension which was initially thought to be steroid induced but MR Aortogram showed short segment smooth narrowing of distal descending thoracic aorta just above the diaphragm(60-70% narrowing), suggestive of distal descending thoracic aortic stenosis due to supradiaphragmatic MAS (Fig 2).Aortic arch and its branches were normal. And there was no stenosis in renal arteries. The patient on treatment with anti hypertensives and antibiotics showed improvement. Patient was counselled to go to higher center for cardiothoracic surgical intervention. In the follow up of the child the hypertension was not controlled (140/96mmHg). She was on regular penicillin prophylaxis for RHD and taking anti hypertensives (nifedipine and enalapril). She failed to go to higher center because of financial constrain. However she was managing to go to school.

**Fig 1:** chest X-ray showing cardiomegaly non homogenous patch left side



**Fig 2:** MRI showing distal descending and thoracic aortic stenosis



### Discussion

MAS is an uncommon condition characterized by segmental narrowing of the abdominal or distal descending thoracic aorta. It constitutes about 0.5-2% of all the cases of aortic coarctation [3]. It can be congenital or acquired. Acquired causes of mid-aortic syndrome include neurofibromatosis, fibromuscular dysplasia, retroperitoneal fibrosis, Williams syndrome, mucopolysaccharidosis, giant cell arteritis (Takayasu disease, temporal arteritis), and acquired insults in utero or in early life that result in developmental disorders of the growing aorta<sup>[4]</sup>. Congenital mid-aortic syndrome is caused by a developmental anomaly in the fusion and maturation of the paired embryonic dorsal aortas and typically manifests itself in young patients.<sup>[5,6,7]</sup> MAS can present as hypertension or lower limb claudication or abdominal angina. The most common anatomic form in congenital or idiopathic middle aortic syndrome is inter renal (19-52%), followed by supra-renal (11-40%), infra-renal (19-25%) and diffuse (12%). Stenosis of the renal arteries is common (60-90%), with less common involvement of the coeliac and superior mesenteric arteries (20-40%), and infrequent involvement of the inferior mesenteric arteries.<sup>[8,9,10]</sup> The exact cause is unknown in most cases though it is probably due to an insult during

intrauterine life. Whilst intimal and subintimal fibrosis and fragmentation of the elastic media are seen, arteritis and atherosclerosis are characteristically absent in this condition. MAS is characterized radiologically by severe narrowing of abdominal aorta and its branches. Most of these patients usually die due to progressive severe hypertension. The imaging differential diagnosis are Takayasu arteritis and neurofibromatosis <sup>[11]</sup>. An initial effort to control hypertension with oral antihypertensives may be successful in a few cases with mild to moderate aortic and/or renal stenosis. The severity of hypertension is the primary indication for intervention and the factor determining procedural timing. Endovascular therapy may provide a sound minimally invasive treatment in MAS caused by discrete aortic stenoses that do not encompass the mesenteric and renal arteries.

Open surgery is the primary treatment of tubular aortic narrowing associated with renovascular hypertension and visceral artery stenosis <sup>[4]</sup>. For patients with active vasculitides surgery is not recommended in active phase of disease. For children best results are achieved if definitive treatment can be delayed till they have achieved full growth.

### Conclusion

A child presenting with feeble pulse and HTN should be first suspected for co-arctation of aorta. But considering initial 10 years of life being normal, secondary cause like Takayasu arteritis and MAS should be kept in mind.

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