Mid-aortic syndrome: a rare cause of heart failure in infants

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Abstract

This case reports describe a rare disease, mid-aortic syndrome (MAS), that can cause severe heart failure and hypertension in infancy. The typical images, key points of diagnosis, and therapy methods of the disease have also been presented. We report two critical thoracoabdominal aortic coarctation cases in infants aged 2 and 11 months with severe heart failure. The patients were initially misdiagnosed as dilated myocardiopathy, with the correct diagnosis confirmed through imaging. Both patients underwent balloon angioplasty; one patient also had bare-metal stents implanted. The patient treated with balloon angioplasty alone died after the procedure, whereas the other patient recovered well. In conclusion, careful physical examinations, especially upper and lower extremity blood pressure differences and palpation of upper and lower limb pulses, are critical in unexplained infant heart failure cases. Stent implantation may be a safer and more effective treatment than simple balloon angioplasty in infants with MAS.

Keywords Heart failure infants; Mid-aortic syndrome

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Introduction

Mid-aortic syndrome (MAS) is a rare clinical condition characterized by segmental narrowing of the abdominal or distal descending thoracic aorta.¹ MAS is usually considered a morphological pattern resulting from more than one disease process, such as genetic abnormalities, inflammation, or viral infection. Most patients with MAS present with severe arterial hypertension, symptoms of intraventricular haemorrhage, or cardiac failure.²,³ It is typically diagnosed in late childhood or young adulthood. Cases of MAS in infancy are sporadically reported. To the best of our knowledge, only a handful of cases of infants with MAS have been reported. Herein, we report two infants with severe heart failure due to MAS who underwent percutaneous interventional treatment.

Case reports

Case 1

The 11-month-old infant was admitted to our hospital due to progressively poor feeding and shortness of breath. On admission, physical examination revealed lower limb blood pressure of 78/53 mmHg (upper extremity blood pressure was not obtained), respiratory rate of 52 per minute, and heart rate of 163 beats per minute. The baby had an S3 gallop prior to S1 and S2 heart sounds, and rales were audible in the lung bases. Abdominal auscultation was not performed. The chest X-ray showed cardiomegaly (Figure 1). Clinical data and imaging findings are detailed in Table 1. Echocardiography showed the origin of the coronary arteries was as expected, and no other structural anomalies of the coronary...
arteries were found. The left ventricle was enlarged, left ventricular function was decreased, and the echogenicity of the endocardium was increased. ECG and Holter monitoring did not identify tachycardia arrhythmias, premature contraction, ventricular pre-excitation, or bundle branch block. Initially, the child was diagnosed with primary dilated cardiomyopathy and given cardiotonic drugs and diuretics, though symptoms did not significantly improve. An incidental finding of a severely stenotic aortic segment from T10 to L2 was identified when the patient underwent an MRI to assess the myocardium and cardiac function (Figure 1). After a multidisciplinary team (MDT) discussion, the patient underwent balloon angioplasty. Angiography confirmed the result of the MRI and showed the segment of interest as nearly atretic. Arteries distal to the coarctation were narrow. 2 × 20/2.5 × 80/3 × 30 mm diameter balloons (Sterling Monorail™ and NC QUANTUM APEX™, Boston Scientific) and a 4 × 40 mm diameter balloon (SABRE®, Cordis) were sequentially used to dilate the stenotic lesion repeatedly (Figure 2). However, the response to angioplasty was poor. The patient’s clinical situation gradually worsened over time, despite medical treatment. One week after angioplasty, CT examination revealed the formation of an aortic dissection at the lesion. Because of the uncertain clinical prognosis, the family refused any further interventions. The child died 2 weeks after the procedure due to worsening cardiac dysfunction and acute renal failure.

**Case 2**

The 2-month-old infant was admitted for severe cardiac dysfunction with dyspnoea. His heart rate was 184 bpm on admission. The infant received non-invasive positive airway

| Table 1 Clinical data of Patient 1 | Baseline | Clinical event |
|-----------------------------------|----------|----------------|
| Weight, kg                        | 9.2      | 1 One week after angioplasty, CT examination revealed the formation of an aortic dissection at the lesion |
| NYHA function, class              | III      | 2 Death at 2 weeks after the procedure due to worsening cardiac dysfunction and acute renal failure |
| ECG                               | Sinus rhythm left ventricular hypertrophic |
| Laboratory parameters             |          |                |
| NT-pro-BNP, pg/mL                 | >35 × 000|                |
| Cardiac troponin I                | Normal   |                |
| Aetiology                         | Negative |                |
| Echocardiographic parameters      |          |                |
| LVEF %                            | 42       |                |
| LVFS %                            | 21       |                |
| LVEDV, mL                         | 46       |                |
| LVESV, mL                         | 37       |                |
| IVSTh, mm                         | 6        |                |
| PWTh, mm                          | 8        |                |
| Systolic pressure gradient (mmHg) |          |                |
| Before angioplasty                | 75 mmHg  |                |
| After angioplasty                 | 68 mmHg  |                |

IVS, interventricular septum; LVEDV, left ventricular end-diastolic volume; LVEF, left ventricular ejection fraction; LVESV, left ventricular end-systolic volume; LVFS, left ventricular fractional shortening; PW: posterior wall.
pressure ventilation with supplemental oxygen, and SpO₂ was maintained at 96%. Invasive arterial blood pressure in the lower limb was 74/58 mmHg, but the upper limb blood pressure was not checked. Clinical examination revealed facial oedema, gallop rhythm, 2/6 grade systolic murmur in the apex, and rales at the bases. Abdominal auscultation of the infant was not performed as in the previous case. Chest radiography demonstrated enlargement of the cardiac silhouette and pulmonary oedema (Figure 3). Echocardiography showed a dilated left ventricle with moderate mitral regurgitation, left ventricular dysfunction with decreased fractional shortening, and enhanced endocardial echogenicity. Doppler showed laminar flow in the aortic arch and down the descending aorta. Laboratory findings are detailed in Table 2. ECG monitoring showed first-degree AV block with sinus tachycardia and left ventricular hypertrophy (Figure 3). The infant was also initially misdiagnosed with primary dilated cardiomyopathy. Pulmonary oedema improved after fluid restriction and commencement of diuretic and cardiotonic drugs. However, clinical symptoms were more likely to recur. A long stenotic segment in the distal descending thoracic aorta was found incidentally when the patient underwent a cardiac-enhanced MRI (Figure 3). After adequate pre-operative evaluation and MDT discussion, the patient underwent angioplasty at 3 months of age. The angiogram of the abdominal aorta showed near-occlusion and excessive development of collaterals at the aortic segment from T9 to L1 (Figure 4). The systolic pressure gradient across the coarctation was 103 mmHg. Initially, efforts were made to salvage the narrowing aorta by balloon dilation. However, the result of angioplasty was unsatisfactory. Subsequently, we chose to place two metal stents (7 ×14 mm, PRECISE PRO RX, Cordis) overlapping in the stenotic lesion. Although local stenosis on sent available, the gradient pressure was remarkably decreased. The diameter of the aorta at the stiff area was 3.6 mm with a peak systolic pressure gradient of 14 mmHg (Figure 4). At 4-month follow-up, the cardiac size and ECG reverted to near-normal. The echocardiogram showed that left ventricular function was as expected, and the mean pressure gradient of in-stent stenosis was only 9 mmHg (Figure 3).

Discussion

These two cases report the diagnosis and treatment of two infants with recurrent left heart failure due to rare aortic lesions. Review of the cause of secondary cardiac enlargement and dysfunction in infants including coronary artery abnormalities, coarctation of the aorta, tachyarrhythmias, exposure to infection, or toxins should always be considered. Laboratory findings and clinical history can quickly rule out other aetiologies and toxins. ECG monitoring can also help detect tachyarrhythmias. Echocardiography can show the origin of coronary arteries, valve lesions, and coarctation of the proximal thoracic aorta. However, coarctations of the distal thoracic aorta and abdominal aorta, defined as mid aortic syndromes, are often overlooked while performing echocardiography. MAS can cause severe hypertension in the upper limbs and diminished pulses in the lower extremities. In theory, an accurate physical examination can help detect the defect. Nevertheless, in infants, blood pressure monitoring is often omitted for various reasons, and palpation of upper and lower limb pulses is often missed, which may lead to misdiagnoses. In this study, these two infants were misdiagnosed as primary dilated myocardiopathy precisely because of the lack of careful physical examination. Vital signs were omitted, such as comparing of upper and lower limb blood pressures and pulses as well as abdominal auscultation. Therefore,
Figure 3  Case 2: (A) Pre-operative chest radiography shows cardiomegaly. (B) 4 months post-angioplasty, the cardiac shadow gradually reduced. (C) The mean pressure gradient of in-stent stenosis was 9 mmHg, and the maximum pressure gradient was 24 mmHg. (D) Pre-operative electrocardiogram demonstrating sinus rhythm and left ventricular enlargement, with complete recovery starting at 4 months after the procedure.
careful physical examinations, especially upper and lower extremity blood pressure differences and palpation of upper and lower limb pulses, are critical in unexplained infant heart failure cases.

The locations and length of aortic narrowing in patients with MAS are highly variable. The stenosis can range from being only a short focal constriction of the aorta to a long diffuse narrowing involving multiple visceral arteries. The most common lesion site is in the suprarenal region. Surgery or endovascular treatment is the mainstay therapy for MAS. The patient's age, the location of the stenosis, the length of the lesion, the extent of visceral vessel involvement, clinical presentation, and response to medical therapy are all factors that should be considered before invasive treatment. In infants, the outcomes from surgical approaches including bypass grafting or aortic patch grafting are poor due to the high rate of reintervention and complications. Any required reinterventions may also be more technically difficult. Therefore, the benefit of delaying surgical revascularization is definite until the child is older. In our cases, the endovascular intervention approach was utilized. In Case 1, the patient only underwent angioplasty with a poor outcome. The aortic stenosis did not improve after the procedure, and aortic dissection was detected on a CT scan 1 week later. Over time, the patient developed worsening heart failure and acute renal insufficiency, which ultimately led to death. The possible contributors to the difficulty in balloon dilatation of the aortic stenosis may be the pathology of the hypoplastic aorta and the length of the aortic lesion. In Case 2, balloon angioplasty's initial result was poor, and aortic dissection was also detected during the procedure. After stents were implanted, the stenosis of the aorta and aortic dissection improved significantly, even though a degree of residual stenosis was present post-procedure. The clinical condition of the patient improved gradually during follow-up. Thus, stent implantation may be a safer and more effective treatment than simple balloon angioplasty in infants with MAS. As infants grow, re-stenosis and impaired flow may develop due to fibrosis related to instrumentation or the apparent lack of growth secondary to stent implantation. The need for surgical

| Table 2 Clinical data of Patient 2 |
|-----------------------------------|
|                                   |
| Weight, kg                       | Baseline | Discharge | Follow-up (4 months) |
| NYHA function class              | IV       | Normal    | Normal               |
| Laboratory parameters            |          |           |                      |
| NT-pro-BNP, pg/mL                | >35 000  | 284.2     |                       |
| Cardiac troponin I, ng/L         | 202.5    | Normal    |                       |
| Aetiology CMV                    |          |           |                      |
| Specific antibodies CMV-IgM      |          |           |                      |
| Echocardiographic parameters     |          |           |                      |
| LVEF, %                          | 42       | 64        | 62                    |
| LVFS, %                          | 20       | 33        | 31                    |
| LVEDV, mL                        | 40       | 16        | 19                    |
| LVESV, mL                        | 23       | 6         | 7                     |
| IVSTh, mm                        | 7        | 7         | 6                     |
| PWTh, mm                         | 7        | 7         | 6                     |
| Systolic pressure gradient, mmHg |          | 103 mmHg  | 9 mmHg (mean)         |
| Before angioplasty               |          | 14 mmHg   | 9 mmHg (mean)         |
| After angioplasty                |          | 9 mmHg    |                       |
| Medication Diuretics + cardiotonic drugs | | Diuretics + ACEI | Diuretics + ACEI |
| IVS, interventricular septum; LVEDV, left ventricular end-diastolic volume; LVEF, left ventricular ejection fraction; LVESV, left ventricular end-systolic volume; LVFS, left ventricular fractional shortening; PW, posterior wall.
treatment in this patient is all but certain, though the timing of open surgical treatment depends on the patient’s clinical condition. Blood pressure and symptoms of heart failure are monitored as indicators during the follow-up.

Heart failure is a sequela in infants with MAS. Accurate blood pressure monitoring and careful physical examination may aid in diagnosis. Primary stent placement may lead to clinical improvement and delay the need for surgical intervention.

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Conflicts of interest

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