Midaortic Syndrome and Hypertension in Childhood Revealed by Enuresis

Abstract

Middle aortic syndrome (MAS) results from a diffuse narrowing of the distal thoracic or abdominal aorta commonly involving both the visceral and renal arteries. Congenital, acquired, inflammatory, and infectious etiologies have been described. Symptoms occur within the first three decades of life. Revelation modes are dominated by hypertension, lower extremity claudication, and mesenteric ischemia. We herein report a pediatric case of MAS with an original revelation mode that has never been described before in medical literature.

Keywords
Midaortic Syndrome; Childhood; Enuresis.

Introduction

Middle aortic syndrome (MAS) is a clinicopathologic term referring to significant segmental tubular narrowing of the suprarenal, inter-renal or infrarenal abdominal aorta frequently associated with concomitant stenoses in the renal and visceral arteries. Ultrasound with Doppler and flow velocity investigations can directly detect the stenosis itself and demonstrate increased flow velocity and mono or biphasic waveforms of arteries distal to the stenosis (as opposed to the normal triphasic waveforms). CT or MR angiographies can confirm the diagnosis. MAS can lead to a series of severe complications, including cerebral hemorrhage, cardiac infarctions, renal insufficiency and mesenteric ischemia. [1].

It’s important to consider the possibility of MAS in specific patient and the setting of classical clinical symptoms. However, MAS can be revealed by an atypical symptom like enuresis as described in this case report.
Case report
A 13-year-old girl was referred to pediatric service for evaluation of severe hypertension. Her hypertension (180/90 mmHg) was incidentally found during a medical consultation for enuresis. Very little attention was paid to the hypertension from the patient due to her showing no particular symptoms. Thus she was admitted to our hospital for more investigations to found the cause of her severe systemic hypertension. The physical examination demonstrated that the patient was in good condition with a body weight of 53 kg. Both the systolic and diastolic blood pressures were elevated. There was no significant pressure gradient between her upper and lower extremities. There was a weak femoral pulse, and no heart murmurs or abdominal bruits. The clinical and laboratory evaluations for primary aldosteronism, pheochromocytoma, Cushing’s syndrome and hypothyroidism were all negative with a normal glomerular filtration rate (82 ml/min/1.73 m²). The patient also denied taking any oral contraceptives or any other particular drugs. There was no past history of a prolonged fever of unknown origin suggesting a diagnosis of Takayasu’s arteritis. Her family history revealed no evidence of secondary hypertension.

Figure 1.
A: subcostal echocardiographic view showing an aliased flow of the abdominal aorta.
B: acceleration of flow in the abdominal aorta with systo-diastolic chronology

Figure 2. CTscan showing the abdominal aorta severely narrowed with normal renal arteries
The ultrasound confirmed that both kidneys were equal in size and contour. The abdominal computed tomography scan revealed no evidence of an adrenal tumor. However, a transthoracic echocardiography with Doppler sonography revealed an increased peak flow velocity of 4.7 m/s in the abdominal aorta, suggesting severe stenosis (Figure 1), which was confirmed by Thoracic and abdominal CT angioscanner demonstrating a generalized hypoplasia of the abdominal aorta below the origin of the renal arteries. Left and right renal arteries were normal (Figure 2).

There was no obvious involvement of the celiac artery or inferior mesenteric artery (IMA).

Her blood pressure was controlled by taking three different antihypertensive agents (valsartan, nebivolol and hydrochlorothiazide). Administration of amlodipine has caused a skin reaction (rash) to our patient in relation to the side effect of this drug. The surgery was postponed until the end of the child’s growth.

Discussion

MAS was first described as a subisthmic coarctation by Dr. Schlessinger in 1835, but it was not until 1963 when Sen et al. described what we now call the middle aortic syndrome [2]. The exact etiology of MAS is still unknown. The young age of these patients suggests that it may be a congenital condition, and the commonly associated findings of neurofibromatosis, Williams syndrome or other congenital anomalies suggest a possible genetic component. Arnott and Louw et al. [3] hypothesized that MAS may represent the failure of the normal fusion of two dorsal aortas. Further, other authors related it to intrauterine injury or infection (e.g. rubella) [4]. The presence of multiple renal arteries, extremely rare involvement of the IMA, and lesions usually located between the celiac axis and IMA suggest that MAS may be related to the embryological development of the kidneys. However, some authors, based on anatomoclinic elements similar to Takayasu’s disease, believe that an origin acquired cannot be excluded [8]. In our patient, the absence of the supra-aortic trunks and the normal inflammatory assessment, are in favor of a MAS of congenital origin. The Takayasu disease was not retained.

The different symptoms depend on the variable degree and location of the blood vessel narrowing. The largest series of 17 MAS patients reported by O’Neill et al. [2], included all patients presenting with severe malignant hypertension, 5 with heart failure, 3 with renal failure, and 2 with severe leg claudications. The majority of the patients in this study did not have either renal failure or leg claudications, probably due to the slow and gradual occlusion of the related blood vessels allowing the development of an effective collateral circulation. Intestinal angina has not been noted despite the celiac and/or superior mesenteric artery (SMA) involvement. As to the extent of the visceral vessel involvement in MAS, Panayiotopoulos et al. [6] demonstrated the characteristics of at least a 90% presence of multiple renal arteries and renal artery stenosis (RAS), 20-40% associated with a celiac axis and SMA stenosis.

No study has described the relationship between enuresis and MAS, it can be hypothesized that this enuresis may be secondary to a circadian anomaly of ADH secretion with increased urine production at night in these patients (Primary or secondary abnormality) [7].

If possible medical management for MAS is preferred in the pediatric population until the child ceased growing so as to prevent a second surgery later in life. The treatment of choice for MAS is now either a one-stage reconstructive prosthetic or autologous venous surgical arterial bypass graft. [1] Percutaneous transluminal renal angioplasty PTRA may play a role in ‘buying time’ for young patients with RAS waiting for surgery. [9] (Figure 3)

The saphenous vein and internal iliac artery are both preferable grafts of choice. O’Neill et al. [2] re-
commended a reinforcement of the saphenous vein using a Dacron net tube mandrill which can prevent aneurysmal degeneration and graft loss. Due to the possibility of recurrent disease in the remaining renal unit, every effort must be made to preserve as much renal tissue as possible. Nephrectomy should be postponed in RAS patients as long as their renal artery is believed to be reconstructable.

The timing of the surgery for patients with surgically remediable hypertension is important. Chalmers et al. [10] recommended that the absolute indications for surgery include a poor BP control with high doses of antihypertensive agents, evidence of end-organ damage (left ventricular hypertrophy and hypertensive retinopathy) and evidence of the deterioration in the renal function and loss of renal mass. The relative indications for surgery include poor compliance with the medication regime and the patient has reached an age where only a single-stage definite operation should be required, due to a young age requires a change in the graft as the patient grows.

Major complications are graft thrombosis, aneurysmal development and stenosis of the anastomosis, which can lead to persistent hypertension, consecutive renal failure, and early postoperative mortality [11].

Conclusion

MAS remain a little-known entity. Its severity is mainly represented by the complications of renovascular hypertension secondary to the aortic stenosis and the attacks of the renal arteries often associated [12]. A systematic measurement of blood pressure in pediatric consultation even in the presence of sometimes atypical signs would allow early diagnosis and adequate management before the complications stage.

Acknowledgments

The authors thank anyone who contributed towards the study by making substantial contributions to conception, design, acquisition of data, or analysis and interpretation of data and who was involved in drafting the manuscript or revising it critically for important intellectual content.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

References


