Mid-aortic syndrome in childhood: case series

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Introduction

Mid-aortic syndrome (MAS) is defined as the stenosis of the abdominal or descending thoracic aorta and can be accompanied by narrowing of the visceral arteries (up to 66% of MAS involve renal arteries). MAS is an infrequent pathology, as it constitutes only 0.5-2% of all the coarctation of the aorta. The etiology of this disease is unknown and most of the cases are idiopathic, but some of them can be genetic or secondary to vasculitis such as Takayasu’s arteritis. In childhood, it is typically presented as severe arterial hypertension.

Management of these patients is still unclear, as the treatment can be pharmacological or, in severe cases, surgical or endovascular.

Methods

Retrospective review of four cases of MAS in a university hospital.

Results

Four patients diagnosed with MAS were included in the review, with a mean age of 11.3 years (6-14 years old), 3 of them were women. All the children were referred to our centre after the discovery of asymptomatic arterial hypertension, while MAS was diagnosed by magnetic resonance angiography in 3 of them and computed tomography angiography in the other patient.

Regarding the location of the lesions, one patient had stenosis of the suprarenal aorta, 2 of them of the paravisceral aorta and one of the descending thoracic aorta. Two of the children also had involvement of the visceral arteries: one of them, the celiac and both renal arteries, and the other patient the celiac, superior mesenteric and both renal arteries.

Endovascular treatment with percutaneous angioplasty and stenting was performed in 2 patients with good angiographic result and short-term arterial hypertension control. Although, one of them required two more angioplasties at 13 and 27 months due to refractory arterial hypertension.

The youngest patient was managed conservative with medical treatment with normalization of blood pressure. The remaining patient is waiting for endovascular treatment.

Conclusions

MAS is an infrequent disease with complex management due to the lack of scientific evidence. Invasive procedures are recommended in patients with severe arterial hypertension with end-organ damage or failure of the medical treatment. In those cases, endovascular procedures may be a safe option with acceptable results.