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LETTER TO THE EDITORS

Cerebrovascular lesion in idiopathic midaortic syndrome in children

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I read with great interest the report by Sethna and colleagues on the idiopathic midaortic syndrome (MAS) in children [1] as they nicely reviewed the presentation, diagnosis and management of this rare syndrome with their own cases. I here draw attention on some points based on my own experience.

I encountered the 2-month-old boy with systemic fibromuscular dysplasia (FMD) who was reported as the youngest case in the literature [2]. He presented with congenital renovascular hypertension due to stenosis of the right renal artery and later developed renal infarction on the contralateral side resulting in renal failure. The boy subsequently died of intracranial haemorrhage at the age of 14 months. During the course, hemiconvulsion caused by a Moyamoya-disease-like cerebral vascular lesion was noted. Stenotic lesions of both the abdominal aorta and its branches, which agreed with the diagnosis of MAS, were also revealed by angiography. Post-mortem examination confirmed that the coronary, splenic and mesenteric arteries were also affected, and their histological findings were compatible with FMD. In this case, multivessels in both intracranial and extracranial arteries were involved.

MAS is an uncommon condition characterised by narrowing of the abdominal aorta and stenosis of its major branches, including renal arteries, causing renovascular hypertension [3], and the diagnosis is based on angiographic findings. Accordingly, it encompasses aetiologically diverse conditions, including FMD. In fact, there are many similarities of reported cases both in pathological findings [4] and in the incidence of the affected vessels between MAS and FMD: lesion in the renal artery was found in 91% [1] and in up to 75% of patients [5], respectively. It was, therefore, surprising that there was no case with cerebrovascular lesions out of 102 patients with MAS [1], whereas 25–30% of patients with FMD were reported to have them [5].

Taken together, it is quite reasonable to speculate that the occurrence of cerebrovascular lesions in MAS must be higher because it was not looked for in most reported subjects, as suggested by Sethna et al. [1]. Therefore, we should have a high index of suspicion regarding cerebrovascular diseases, which can be fatal, as in my patient's case, and seek them by angiographic imaging, including magnetic resonance angiography when making a diagnosis of MAS.

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