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*Letter to the Editor*

# Intramural small vessels in arteriovenous malformations of the heart: a note on prognostic significance

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To the Editor,

We read with interest the article by McCammond et al in the May issue of this Journal.[1] The article describes the well-documented case records of a 12-year-old girl with congenital arteriovenous malformation of the heart. This rare lesion was histopathologically sub-classified as being an intramuscular small vessel arteriovenous malformation by means of endomyocardial biopsy. The authors further report on a benign course with no change in appearance of the mass on serial echocardiograms or any concerning symptoms on the basis of a 30-month follow-up. The description of this interesting case is followed by a review of the literature on vascular lesions of the heart, which reveals a marked heterogeneity in the types of benign vascular lesions ranging from various types of angiomas to congenital vascular malformations. Some were reported to be harmless, whereas others showed unfavourable outcome, including a 15-year-old boy with mixed small vessel and cavernous vascular cardiac malformation who witnessed sudden cardiac death.[2]

In 2010, we also described two patients with cardiac arteriovenous malformation with intramuscular small vessel component, which was confirmed at autopsy in both cases.[3] These are not mentioned in the review section of the article, but can be of particular interest for the interpretation of the long-term outcome of such vascular masses. In our first case, a 22-year-old man, endomyocardial biopsy was performed, which showed a similar pattern of intramuscular microvessels, as is shown in figure 4 of the article by McCammond et al. This patient died 9 months later because of progressive cardiac failure. The second patient was known to have a large mass of the anteroseptal part of the heart since birth, and died of intractable arrhythmias at the age of 14 years. In both cases, the autopsy of the heart showed a large component of small vessels amidst the large malformed vessels of an arteriovenous malformation. Occurrence of a microvascular component in arteriovenous malformations is not so rare. In a series 109 patients with symptomatic vascular malformations of soft tissue and skin, we observed small vessel masses in 30% of the excised lesions, which were nearly all (90%) of the arteriovenous type.[4] Clinically, these lesions are easier to follow than their counterparts in the heart, and we noticed that microvascular growth can occur episodically, also later in life, and is often associated with symptoms such as pain and swelling. Altogether, these observations may lead to the notion that arteriovenous malformations with intramuscular small vessels are not so harmless as suggested by the uneventful 30 months' follow-up of the patient presented by McCammond, and this is further illustrated by the fatal outcome of our two cases. We agree with the strategy of "watchful waiting", as is advocated by the authors, which includes regular imaging of the vascular mass and electrophysiological surveillance. However, to our opinion, this strategy should be continued also in the long term, likely throughout life, because of the unpredictable biological behaviour of arteriovenous malformations with a small vessel component.

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