Comment on: Vasculitis, fibromuscular dysplasia or hereditary aneurysms? by Marvisi *et al.*

Sirs,

With great interest we have read the recent letter by Marvisi *et al*. reporting on a patient with vascular lesions of different aetiologies (1). The authors provided an overview of disorders that should be considered in the differential diagnosis of large- and medium-vessel vasculitis, including fibromuscular dysplasia and hereditary aneurysm formation. In patients with evidence of vessel wall aneurysms or dissection, it is also important to consider segmental arterial mediolysis (SAM) (2). SAM is a rare condition of unknown aetiology that causes a non-atherosclerotic, non-inflammatory, non-hereditary arteriopathy. Disruption of the arterial medial layer is the main histological finding, which causes the vessel to be susceptible to aneurysm formation and dissection (3).

It is essential to recognise this disorder to avoid unnecessary immunosuppressive treatment. Kalva *et al*. have developed criteria to establish a non-invasive diagnosis of SAM based on the a) clinical presentation, b) exclusion of other vascular disorders, c) laboratory results, and d) imaging criteria for when a tissue sample is not available (4, 5). The clinical presentation is typically determined by the vascular areas that are involved. SAM mainly targets medium-sized arteries within the abdominal cavity, where it may particularly resemble polyarteritis nodosa and fibromuscular dysplasia (5). Symptoms may be acute in nature, including abdominal pain or chest pain with involvement of the mesenteric or cardiac vessels, respectively, or hypotension in case of aneurysm rupture or dissection. Chronic symptoms such as hypertension and haematuria may arise due to renal infarctions. Other clinical features such as constitutional symptoms, arthralgia, and cutaneous manifestations, which are often present in polyarteritis nodosa, are absent in patients with SAM (6).

It is important to rule out congenital vascular abnormalities associated with connective tissue disorders, fibromuscular dysplasia, and vasculitis in all patients with suspected SAM. It may be particularly challenging to distinguish SAM from fibromuscular dysplasia, and some even consider it to be a precursor of fibromuscular dysplasia (7, 8). Fibromuscular dysplasia typically affects young women and has a predilection for the renal arteries (6). SAM may affect individuals of any age, has no gender predilection, and typically involves the splanchnic vasculature (6). Inflammatory markers are often within normal limits and anti-nuclear antibodies (ANA) and anti-neutrophil cytoplasmatic antibodies (ANCA) should be absent in patients with SAM (4). However, inflammation may be transiently present due to associated haemorrhage or tissue ischaemia, which often triggers the initiation of corticosteroids for presumed vasculitis. Imaging may show evidence of dissection, fusiform aneurysm formation, vessel occlusion, a beaded vessel appearance, or vessel wall thickening with or without organ infarction. In addition, it may also reveal perivascular cuffing or inflammation caused by reactive changes secondary to the vessel wall abnormalities (9). In our experience, this cuff may even capture 18F-fluorodeoxyglucose (18F-FDG) on 18F-FDG PET imaging. These changes may also be seen in patients with vasculitis, which further contributes to the challenging diagnosis.

We fully agree with Marvisi *et al*. that it is crucial to consider the non-inflammatory arteriopathies in patients with suspected vasculitis. Considering the overlap in clinical and radiological presentation, in particular when the mesenteric vessels are involved, SAM is best included on the list of differential diagnoses of large- and medium-vessel vasculitis.

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