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Letter to the Editor

A case of segmental arterial mediolysis with widespread involvement of cranial and abdominal arteries

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Dear Editor,

Segmental arterial mediolysis (SAM) is a very rare, nonatherosclerotic, non-inflammatory vasculopathy of unknown etiology characterized by non-inflammatory degeneration of the medial layer of muscular arteries [1,2]. Inflammatory vasculitis represent the main differential diagnoses. Although, most of the time, arterial involvement concerns only one anatomical site, it can spread to concomitant cerebral and abdominal vessels [3–5]. We report an unusual case of segmental arterial mediolysis with widespread involvement of cranial and abdominal arteries.

A 40-year-old man with a past medical history of migraine was referred for the assessment of a sudden diffuse abdominal pain and intense cervicalgia with stiff neck.

The patient did not suffer from recent neck trauma, and there was no family or personal history of hereditary disorders of connective tissue nor systemic autoimmune diseases. There was no history of fever, weight loss, chills, drug addiction, sweats, photosensitivity, rashes nor oral or genital ulcers. Physical examination revealed a temperature of 37,5 °C, no neurological focal signs but a diffuse abdominal tenderness with no rebound, guarding, distension nor masses. Skin examination was normal.

CT angiography of the supra-aortic and intracranial arteries revealed several dissecting aneurysms involving bilaterally the extracranial internal carotid and vertebral arteries, and multiple dilated areas alternating with narrowing of the blood vessels (Fig. 1A–C). Brain MRI showed no recent infarct whereas the axial spin echo FLAIR-Weighted sequence with fat suppression, demonstrated a recent left internal carotid artery dissection (Fig. 1D). CT angiography of abdomen and pelvis showed an aneurysmal dilatation of the celiac artery (15 mm), a right common iliac artery dissection, multiple irregularities of the mesenteric, splenic and renal arteries associated with an infarction of the upper pole of the left kidney (Fig. 1E-G). CT angiography of chest was unremarkable.

Extensive laboratory tests including cerebrospinal fluid examination showed no inflammation, blood cultures were negative and immunologic workup was normal. Transthoracic echocardiogram was also normal. The diagnosis of segmental arterial mediolysis (SAM) was suspected and the patient was managed conservatively with introduction of an angiotensin-converting-enzyme inhibitor and antiplatelet therapy, allowing a favourable clinical course.

SAM is a nonatherosclerotic, non-inflammatory vasculopathy of unknown etiology, originally described by Slavin and Gonzalez-Vitale in 1976 characterized by non-inflammatory degeneration of the medial layer of muscular arteries and sometimes adjacent veins [1,2] leading to aneurysms formation, dissections, dissecting aneurysms, arterial stenosis or occlusions [6,7]. It is an enigmatic disorder, poorly understood with many aspects remaining unclear. Although no clear familial or genetic predispositions have been demonstrated nowadays [3], some authors suggested a potential causal link between SAM, points mutations in the type 3 procollagen gene (COL3A1) and vascular Ehlers-Danlos syndrome [8]. The etiology is still unknown, but some authors suggest that arterial lesions could be in relation with vasospasm possibly ascribed to focal endothelial paracrine dysfunction. This is supported by the findings that arteries with chronic vasospasm have histologic features in common with SAM and that various disease states that provoke pathologic stimuli for endothelial mediated vasoconstriction occur in the immediate clinical background of patients with SAM (eg medical history of hypertension, shock, hypoxia, recent surgery, migraine, stroke or Raynaud's disease) [9,10]. Moreover, following animal experiments on greyhound dogs, it was suggested that SAM may be a disorder principally caused by iatrogenic or accidental exposure to alpha-1 adrenergic receptor agonists or Beta-2 agonists (such as ractopamine) able to release norepinephrine from the peripheral nervous system [11]. The role of catecholamine hyper-reactivity to stress has also been suggested. Consequently, an acute psychological stress could constitute an external trigger leading to SAM [12]. Clinical presentation is variable, ranging from abdominal pain to potentially life-threatening manifestations (sudden hemorrhage of abdomen, retroperitoneum or brain) [1,6]. In a systematic literature review of 85 patients with SAM between 1976 and 2012, it appears that mainly the celiac artery and its branches, and the superior mesenteric artery were involved, nonetheless, involvement of the cerebral or renal vasculature, as well as the inferior mesenteric artery has also been described [3]. Although, arterial involvement often concerns only one anatomical site, it can spread very rarely to concomitant cerebral and abdominal vessels [3–5] as in our case. A concomitant manifestation of SAM on neurovascular and abdominal arteries is even rarer and the explanation is unknown. Conventional angiography remains more sensitive than CT or MR angiography, and should be used if more conventional methods of

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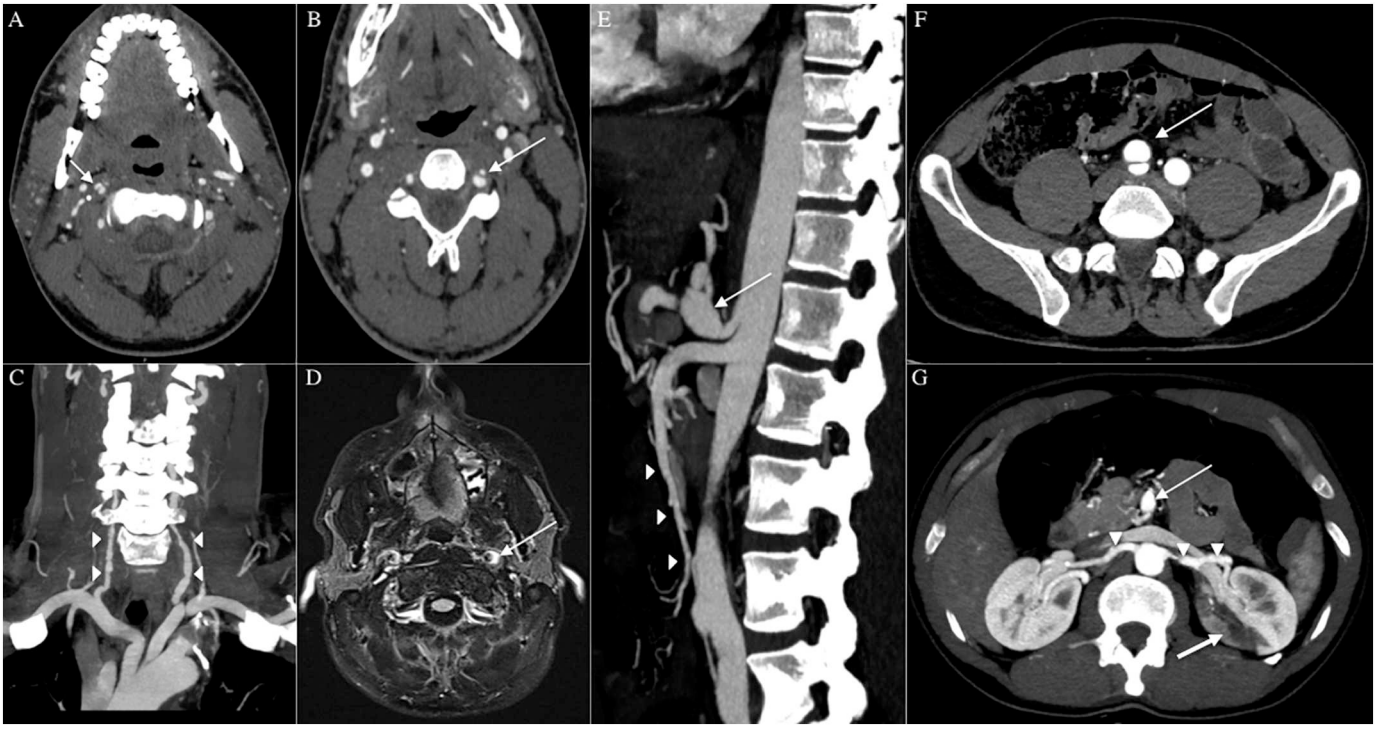

Fig. 1. Imaging of the supra aortic trunks, intracranial and abdominal vessels. CT angiography of the supra-aortic trunks revealed dissecting aneurysms involving multiple arterial segments especially the right carotid artery (A, arrow), the left vertebral artery (B, arrow), the right vertebral artery (not shown) associated with multiple areas of stenoses involving especially the vertebral arteries (C, arrowheads). Brain MRI with axial spin echo FLAIR-Weighted sequence with fat suppression showed the left extracranial internal carotid artery intramural hematoma corresponding to a recent dissection (D, arrow).
CT angiography of abdomen disclosed the 1.5 cm aneurysmal dilatation of the celiac artery (E, G small arrows), a right common iliac artery dissection (F, arrow) and multiple irregularities involving the superior mesenteric artery (E, arrowheads) and renal arteries (G, arrowheads) associated with infarction of the upper pole of the left kidney (G, large arrow).

imaging are unrevealing [1]. Histologic diagnosis is obtained in a minority of cases in the literature, most often when the patient management needs the exeresis of a pathological arterial segment [3]. Initial treatment is most often conservative. However, treatment of symptomatic aneurysms (in the acute phase, often as a bailout measure) is performed most of the time with endovascular techniques as coil embolization, which appear safer than an open chirurgical approach [3]. Differential diagnoses are numerous and include mainly systemic vasculitis of large or medium vessels (polyarteritis nodosa, Kawasaki disease, Behcet's disease, giant cell arteritis, Takayasu's disease), infectious causes such as mycotic aneuryms as a consequence of infective endocarditis, but also non inflammatory diseases such as fibro muscular dysplasia, hereditary disorders of connective tissue (type IV Ehlers-Danlos syndrome, pseudoxanthoma elasticum, Marfan's syndrome), neurofibromatosis or atherosclerosis [1]. In our case, there were no anamnestical, clinical or biological arguments for an inflammatory or infectious vasculitis. Our patient did not present marfanoid habitus, and normal echocardiography and CT angiography of the chest were not suggestive of Marfan's or related disorder. In the same way, fibromuscular dysplasia generally affects younger females with a predilection for renal vascular involvement, causing premature hypertension. SAM has no gender predisposition, affects mainly the celiac artery and its branches and arterial dissections are much more common in SAM than in fibromuscular dysplasia [1]. To conclude, SAM diagnosis should be considered in case of acute onset of abdominal pain or headache especially in the presence of medium and large vessels abnormalities such as dissections, aneurysms, stenoses or occlusions even if they affect many anatomical sites and after having ruled out differential diagnoses.

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Conflicts of interest

All authors report no conflicts of interest.

Authors contributions

Dr Elodie Delafosse, MD: Study concept and design.

Dr Guillaume Armengol, MD: critical revision of the manuscript for important intellectual content.

Dr Aude Triquenot-Bagan, MD: critical revision of the manuscript for important intellectual content.

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