

Catastrophic multiple arterial dissections revealing concomitant polyarteritis nodosa and vascular Ehlers-Danlos syndrome

Sirs,

Multiple arterial dissections are rare and usually complicate arterial fibromuscular dysplasia, pregnancy or disorders of the connective tissue (1, 2). We report here the case of a 31 year-old woman presenting with fatigue and fever evolving for 10 days, associated with abdominal and chest pain and a painful swelling on her left forearm. Physical examination revealed articular and cutaneous hyperlaxity and a Marfan-like phenotype. Neutrophil count, C-reactive protein and serum creatinine levels were elevated. Tests for antineutrophil cytoplasmic antibodies and antinuclear antibodies yielded negative results. CT-scan showed bilateral carotid artery dissection, left carotid-cavernous sinus fistula, right renal artery dissection with partial renal infarction, left renal artery dissection with a partially thrombosed aneurysm (Fig. 1A), superior mesenteric artery dissection and primary iliac artery ectasia (Fig. 1B). The patient rapidly worsened due to left arm compartment syndrome caused by a ruptured ulnar artery aneurysm. Rapidly, she went into hypovolemic shock revealing new left lumbar, iliac and femoral artery dissections and an extended psoas and mediastinal haematoma. Emergency multiple arterial embolisations associated with the surgical ligation of the left ulnar artery were performed. By day 4, the pathological results of the left ulnar artery showed a necrotising panarteritis with fibrinoid necrosis highly suggestive of the diagnosis of polyarteritis nodosa (PAN). Due to the patient clinical history, morphology and the presence of a carotid-cavernous sinus fistula, vascular Ehler-Danlos syndrome (EDS) was also suspected and confirmed a few weeks later by the presence of the COL3A1 mutation. The patient worsened until she received intravenous methylprednisolone (15mg/Kg) and intravenous cyclophosphamide (CYC) (700 mg/m²). After 9 months of prednisone and CYC bolus, azathioprine (100 mg/d) and corticosteroids were prescribed as maintenance therapy. Treatment was fully stopped at 30 months. The patient did not experience a relapse of the vasculitis during the 6 years follow-up.

Vascular EDS is a rare autosomal dominant genetic collagen disorder with an estimated prevalence of approximately 1:150,000 inhabitants. Mutations in the COL3A1 gene (5) encoding for the pro- α chain of type III collagen explain the extreme vessels and tissue fragility and is detected in no more than 61% of cases. Arterial lesions

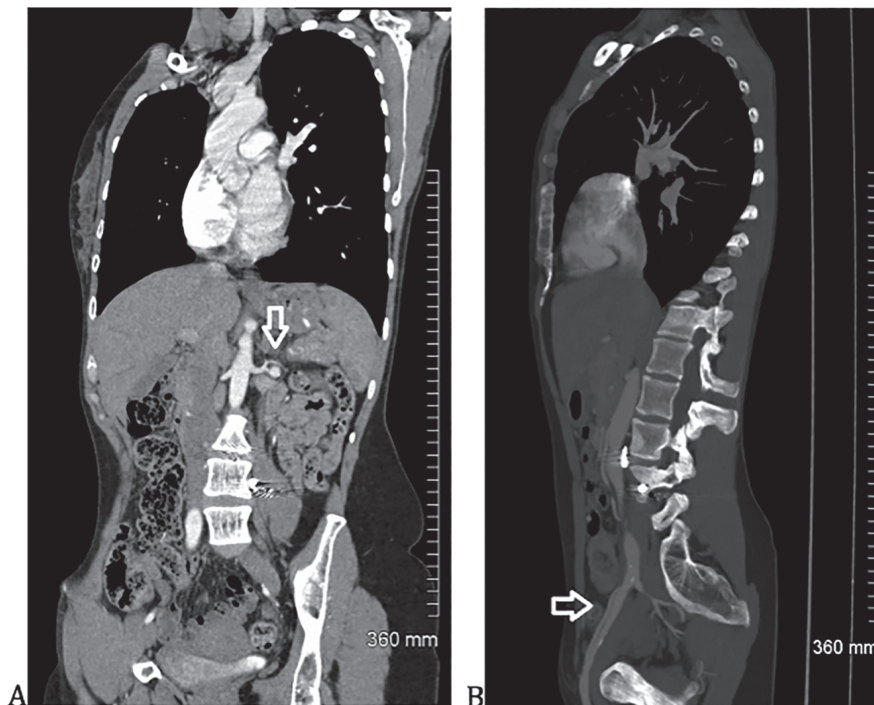


Fig. 1. A: Left renal artery dissection. B: Primary iliac artery ectasia.

suggestive of the disease include an excessively bulbic internal carotid; aorta-, iliac-, and visceral branch-dissecting aneurysms; spindle-shaped aneurysms of the splenic artery, and early onset non-direct carotid traumatic events. The presumptive vascular EDS diagnosis is based on consensual, yet not formally validated clinical criteria and compatible non-invasive arterial imaging (6,7). Management is complex and requires multiple specialists who can respond to and manage the major complications. PAN is a rare systemic necrotising vasculitis that predominantly affects medium-sized arteries. Its estimated incidence is approximately 7:1,000,000 inhabitants in France, in decline (8). Vasculitic lesions are caused by the deposition and/or the in situ formation of immune complexes which activate the complement membrane attack complex and neutrophils resulting in fibrinoid necrosis of the media (9). Treatment is based on corticosteroids and immunosuppressive therapy.

To the best of our knowledge, we report the first case of catastrophic multiple arterial dissections revealing the association of PAN and vascular EDS. PAN is exceptionally associated with arterial dissections whereas arterial dissections are present in 5 to 10% of case of vascular EDS (2-4). In the present case, the vasculitic process resulted in additional vascular stress, probably facilitated catastrophic multiple dissections in a young patient already suffering from undiagnosed vascular EDS.

We suggest searching for vasculitis in vascular EDS patients presenting with multiple arterial dissections in a context of poor

general condition, myalgia, and biological inflammation. PAN requires an early diagnosis in this context as if corticosteroid and immunosuppressive therapies are delayed, the clinical course may be catastrophic.

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