Cholesterol Crystal Embolization

Cutaneous pseudovasculitis represents a diverse spectrum of disorders that might clinically mimic cutaneous vasculitis and can be broadly classified into diseases that produce hemorrhage or vessel occlusion (1). Pigmented purpuric dermatitis, scurvy, senile purpura, idiopathic thrombocytopenic purpura, and viral or drug induced eruptions are some of the pseudovasculitic disorders that are the result of dermal hemorrhage due to vessel wall incompetence. Apart from the above mentioned disorders, diseases like cholesterol embolism, antiphospholipid syndrome, thrombotic thrombocytopenic purpura or livedo vasculopathy cause vascular injury due to infarction secondary to luminal occlusion (livedo, cyanosis, ulcers, digital necrosis) (1). Skin biopsy, as a crucial step in making the correct diagnosis, allows separation of these entities from true vasculitis. A histological hallmark of cutaneous vasculitis is the presence of inflammatory infiltrate within or around the vessel walls accompanied by fibrin deposition (fibrinoid necrosis) (2). The lack of histologic criteria required for the diagnosis of authentic vasculitis should always direct evaluation and diagnosis towards pseudovasculitis. Cholesterol crystal embolization, as one of these vasculitis mimics, is a severe complication of atherosclerosis originating from ulcerated atherosclerotic plaques of the aorta or its major branches (3). Since its first description in 1945, it has been described in almost every major organ system, often mimicking multisystemic disease resembling systemic vasculitis and being underdiagnosed as leading to multiorgan failure and death (4). It can occur spontaneously or following invasive vascular procedures, anticoagulant and thrombolytic therapies. When an atheromatous plaque is disrupted, its components are released into the bloodstream occluding small arterioles. Clinical manifestations, mostly those of renal and skin involvement, reflect ischemia of the affected organs. The kidney, because of its proximity to the aorta and direct blood supply from that vessel, is the main target organ with progressive renal failure developing in almost all patients affected (4,5). Livedo reticularis is usually the first and most common sign of skin involvement, although cyanosis, ulceration and gangrene might be seen as well (6,7). Livedo is generally bilateral, involving legs and feet. Examination of the patient in upright position has been suggested to increase the likelihood of detecting it. Some reports point to the fact that it can be missed when examining the patient while lying down (6). The timing of clinical findings varies from days (sometimes hours) to weeks. Demonstration of cholesterol clefts in deep dermal vessels from areas of livedo reticularis in skin biopsies is a prerequisite for the diagnosis. Histologically, initial lesions can be associated with perivascular infiltrate of leukocytes, lymphocytes and eosinophils, whereas older lesions will have multinucleated giant cells and fibrosis of the vessel wall (1). There still are no standard treatment modalities for cholesterol embolization. Corticosteroids have been proposed by some authors while others stand for pentoxifylline and the statins, which stabilize cholesterol-rich plaques and prevent recurrence of cholesterol embolization (7).

We might expect that the prevalence of the aforementioned disease will increase because the incidence of atherosclerotic disease is higher daily, and interventional procedures as well as anticoagulation therapy are being ever more widely used. Cholesterol embolization should be taken in consideration on the differential diagnosis of necrotic skin lesions or livedo reticularis in patients having undergone invasive vascular interventions, anticoagulant therapies, or in those with acute renal failure.

References

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