

## Case Report

# The Buerger's Disease and the Type I Cryoglobulinemia, T Entities Expression of the Same Type III Hypersensitivity Reaction

Mancuso G\*

Department of Internal Medicine, EOC, "La Carità" Hospital of Locarno, Switzerland

### Abstract

The type I cryoglobulinemia vasculitis is a life-threatening disorder because of the severity of cutaneous and renal involvement and the underlying hemopathy and even a limited quantity of paraproteins can cause severe symptoms and the Type I cryoglobulinemia accounts for 10%–15% of patients with cryoglobulinemia. Thromboangiitis obliterans, (formerly called Buerger disease), is an uncommon cause of vasculitis, that affects the small to medium-sized arteries and veins of the extremities (upper and lower) and, less commonly, cerebral/visceral vessels with a nonatherosclerotic, segmental, inflammatory disease.

We describe a 53-year-old woman heavy cigarette smoker seen for extremity pain, claudication, recent complicated Raynaud phenomenon. She had a type I cryoglobulinemia while the other laboratory tests were normal and bone marrow histology revealed aspecific changes.

A causal relationship between TAO and cryoglobulinemia could not be proved and we suppose that the relationship between these two entities may be the expression of a unique dysregulation of type III hypersensitivity reaction.

The Thromboangiitis Obliterans (TAO), also called Buerger disease, an uncommon cause of vasculitis, is a nonatherosclerotic, segmental, inflammatory disease that affects the small to medium-sized arteries and veins of the extremities (upper and lower) and, less commonly, cerebral/visceral vessels [1].

\*Corresponding author: Mancuso G, Department of Internal Medicine, EOC, "La Carità" Hospital of Locarno, Switzerland, Email: mancuso.gaia@hsr.it

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The prevalence is estimated at 1/10,000 in Europe but the disease is found worldwide [2]. Patients present with clinical manifestations of an occlusive vascular disease characterized by distal arteriopathy: Raynaud's phenomenon, intermittent claudication at the beginning, rapidly followed by digital ischemia and/or gangrene. TAO may also involve temporal artery mimicking temporal arteritis [1,2]. The etiology of Buerger disease is unknown, but use or exposure to tobacco is central to the initiation and progression of the disease and the pathological mechanism may be delayed-hypersensitivity [3], a toxic angiitis or a cell-mediated sensitivity to types I and III human collagen, which are constituents of blood vessels [3]. TAO is characterized by highly cellular and inflammatory occlusive thrombus with relative sparing of the blood vessel wall the diagnosis is made with the Olin's criteria after exclusion of other entities that cause vascular occlusive disease [4]. The most important aspect of treatment is cessation of smoking (active and passive); the prognosis for those who succeed being generally very good [5]. Additional medical treatment options include the use of aspirin, Iloprost or streptokinase, with Iloprost (used intravenously), shown to be more effective than aspirin both in the short and long term [1,2].

A 52-year-old patient presented to the Immunologic Clinic because of the recent appearance of Raynaud phenomenon and digit painful ulcers. She complains claudication in the lower limbs with a walking autonomy of 100m.

She is a heavy tobacco smoker and otherwise well with no familiar history of immune rheumatological disease. On physical examination there were symmetrical digital ulcer on III and II digits, no puffy fingers, or cutaneous sclerosis were noted. The Allen test positivity on the both arm raised the suspicion of a small vessel disease. Capillaroscopic picture was characterized by poor visibility due to marked edema of the fundus with the capillary bed architecture apparently preserved with rare tortuosity and isolated ectasias but no megacapillaries or apical microhemorrhages. Preserved capillary density, in the absence of empty papillae or frank avascular areas. No neo angiogenesis phenomena are appreciated.

At that time we couldn't examine all digits due to marked hyperalgesia in the affected areas. The serologic work-up revealed aspecific and low ANA positivity (1:160) with granular pattern (AC4-5), no phospholipid antibodies were detected, the serologic evaluation for HIV, HBV, HCV was negative, as well as no flogosis was showed.

A cryocrit equal to 2% and an electrophoresis monoclonal component equal to 1.5g/L in the gamma zone of IgM lambda type equal to 2.1% of the total were revealed. The color Doppler echocardiogram was normal whereas the lower and upper limb color doppler ultrasound detected a small amplitude flowmetry of the radial and ulnar arteries bilaterally.

We recommend smoking cessation and therapy with iloprost infusion 0,5 ng/kg/min in 6 hours infusion on three days every 4 weeks, cilostazole (100mg twice daily), aspirin(100mg once a day), with rapid improvement of skin ulcer.

She underwent to bone marrow biopsy that showed a specific changes revealing a cellularity equal to 60%, slight and focal increase in the reticulin texture, with slightly reduced megakaryocytes. Rare CD20+ B lymphocytes and occasional CD3+ T lymphocytes even in reactive aggregates. Thus the hematological evaluation excluded the presence of a florid lymphoproliferative disease, but did not exclude a form of low aggressiveness and negligible neoplastic mass capable of maintaining a vasculitis with a dysreactive pathogenesis.

An attempt at immunosuppressive and anti-inflammatory treatment was judged as advisable: the patient started oral steroid therapy (prednisolone 1mg/Kg) and continued the infusions of iloprost, cilostazole and abstinence from smoking, with the complete tissue "restitution ad integrum" in three months where she present a dramatic improvement of the presenting symptoms showing a full disease recover.

Among differential diagnosis of TAO with other vasculitis there is the cryoglobulinemia vasculitis (CV). Vascular occlusion is more frequent in type I cryoglobulinemia, which is usually accompanied by high cryoglobulin concentrations, and can be associated with hyper-viscosity syndrome and cold-induced acral necrosis. Type I cryoglobulinemia accounts for 10%–15% of patients with cryoglobulinemia. Data on type I CV are scarce in the literature, therefore the characteristics and outcome are poorly known [6].

The type I CV is considered a life-threatening disorder because of the severity of cutaneous and renal involvement and the underlying hemopathy and even a limited quantity of paraproteins can cause severe symptoms [7]. The management of type I CV is based on empirical data in those patients without overt B-cell lymphoma, the treatment of vasculitis may include plasma exchange, corticosteroids, alkylating agents, rituximab, or iloprost [8].

A causal relationship between TAO and cryoglobulinemia could not be proved, until now we are aware that just one case has been described and we suppose that the relationship between these two entities may be the expression of a unique dysregulation of the same hypersensitivity reaction [9].

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