CASE REPORTS

Raynaud's phenomenon as initial manifestation of cutaneous polyarteritis nodosa

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Abstract

The case of a 45 year old man with cutaneous polyarteritis nodosa and Raynaud's phenomenon as initial manifestation of the disease is reported. Although peripheral vascular disease is a well characterised extracutaneous manifestation of cutaneous polyarteritis nodosa, to our knowledge this is the first reported case in which Raynaud's phenomenon was the initial and sole manifestation of the disease for a long time.

Cutaneous polyarteritis nodosa is a well characterised cutaneous vasculitis with occasional extracutaneous manifestations.^{1 2} Its relation with systemic polyarteritis nodosa is still unclear. Both entities share pathological features in the affected arteries, but the clinical expression, severity, and prognosis are remarkably different. Peripheral vascular disease has been reported among the extracutaneous manifestations of cutaneous polyarteritis nodosa.^{1 3-5} We present the case of a 45 year old man with Raynaud's phenomenon as the initial and sole manifestation of cutaneous polyarteritis nodosa.

Case report

A 45 year old white man was admitted to our hospital with a nine year history of triphasic Raynaud's phenomenon on both hands. Nine years earlier, classic triphasic Raynaud's phenomenon had appeared on both hands, with

a necrotic ulcer on the third finger of the left hand. He was studied at another institution. Clinical and analytical investigation did not disclose systemic disease. Because the lesion spread the finger was amputated; pathological examination of the finger removed showed no specific changes or vascular inflammatory signs. Despite this, high dose corticosteroid treatment was begun empirically, and the Raynaud's phenomenon clearly improved. No other drug to which the improvement might be attributed was being given at that time. Corticosteroid treatment was continued for years, and could not be suspended owing to worsening of the Raynaud's phenomenon on the second finger of the right hand and diffuse myalgia of the feet. An exhaustive clinical history did not suggest systemic or visceral disease. He did not smoke or drink coffee, tea, or cola beverages, and he had not been exposed to drugs, industrial freezing, or vibrating tools.

Examination showed an unaffected general condition, Cushingoid facies, amputation of the left third finger, and pallor on the ulnar aspect of the right hand as the only abnormal data. Peripheral pulses were normal. There were no skin lesions, clinically apparent peripheral nerve involvement, or arthritis.

Blood studies (haemoglobin, platelets, and leucocyte count) were normal. Erythrocyte sedimentation rate was 53 mm/h. Test results for liver enzymes, blood urea, creatinine, creatine kinase, and aldolase were normal, as were the coagulation profile and urine analysis findings. The protein profile, IgM, IgG, IgA, serum and urine electrophoretic analysis, latex fixation for rheumatoid factor, antinuclear and anti-DNA antibodies, cryoglobulins, cryoagglutinins, plasma viscosity, C3 and C4 complement components and Venereal Disease Research Laboratory test were normal or negative. Virus B serology findings (HBsAg, HBeAg, anti-HBcAg, anti-HBs, and anti-HBe) were negative. A radiographic examination showed that the chest, abdomen, hands, and cervical spine were normal. Arteriography of the hands, kidneys, and coeliac trunk disclosed, on the hand, a filling defect in the distal portion of ulnar artery and palmar ulnar arch, narrowing of the second interosseous artery, and absence of filling on the external collateral arteries of the second and third fingers (fig 1). There were no proximal abnormalities on the hands and abdominal arteriography was normal.

The patient was discharged with a provisional diagnosis of Raynaud's phenomenon, probably

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Figure 1 Arteriography of the right hand shows a narrowing of the second interosseous artery (arrow) and absence of filling on the external collateral arteries of the second and third fingers (arrowheads).

Figure 2 Photomicrograph of a skin biopsy specimen showing necrotising arteritis affecting medium sized arteries in the deep dermis. (Haematoxylin and eosin.)

secondary to distal inflammatory arteriopathy, though there were insufficient criteria for exact diagnosis at that time. He was treated with non-steroidal anti-inflammatory agents, nifedipine, and corticosteroids (prednisone 5 mg/day) with good control of symptoms, but steroid treatment could not be discontinued as Raynaud's phenomenon returned on the right hand.

One year after his initial admission he consulted for a flare of more than 50 reddish painful nodules, most with secondary ulceration, on both legs. Exhaustive clinical and analytical evaluation failed again to disclose general or visceral disease. A skin biopsy showed necrotising arteritis affecting medium sized arteries in the deep dermis (fig 2). Cutaneous polyarteritis nodosa with extracutaneous distal vascular disease was diagnosed. Skin lesions healed promptly after prednisone 20 mg/day was started; the doses were tapered to 5 mg/day after a few weeks. After a three year follow up the patient remains without Raynaud's phenomenon, but attempts to reduce prednisone doses below 5 mg/day result in its return on the right hand. There has been only one new flare of skin lesions and no evidence of visceral extension of the disease has developed.

Discussion

We describe a patient with cutaneous polyarteritis nodosa and longstanding Raynaud's phenomenon as the initial manifestation of the disease. Skin biopsy confirmed cutaneous polyarteritis nodosa and arteriography showed distal arteriopathy on the right hand. Exhaustive clinical and analytical studies failed to discover more extensive disease and a three year follow up has shown no new organs to be affected.

In this patient other causes of Raynaud's phenomenon were ruled out. The major involvement of right hand and positive arteriographic findings do not support a diagnosis of

idiopathic Raynaud's phenomenon. He was not a smoker and arteriography did not disclose the typical corkscrew image of thromboangitis obliterans. Also, there were no arteriosclerotic lesions on proximal arteries. Clinical and radiological examination did not disclose data suggestive of thoracic outlet syndrome. There was no exposure to drugs, polyvinylchloride, vibratory tools, or industrial freezing. Clinical and analytical evaluation did not find evidence of scleroderma, CREST (calcinosis, Raynaud's phenomenon, oesophageal dysmotility, sclerodactyly, telangiectasia) syndrome, mixed connective tissue disease, polymyositis-dermatomyositis, systemic lupus erythematosus, rheumatoid arthritis, or occult neoplasm. Several findings supported an inflammatory origin of Raynaud's phenomenon in this patient, such as the high erythrocyte sedimentation rate, close relation between changes in steroid treatment and disease manifestations, and compatible arteriographic findings. Raynaud's phenomenon has been found to accompany vasculitis, particularly essential mixed cryoglobulinaemia associated with persistent virus B antigenaemia,⁶⁻⁸ though probably it can appear in other medium and small sized vasculitis, and it may even be the initial and sole manifestation of the disease, as our case shows.

Cutaneous polyarteritis nodosa is a necrotising vasculitis confined to dermis and hypodermis, though mild extracutaneous disease is often seen. Since the series of Borrie in 1972¹ and Díaz-Pérez and Winkelmann in 1974² cutaneous polyarteritis nodosa is a well characterised cutaneous vasculitis. The fact that systemic polyarteritis nodosa affects skin in about 40% of cases⁹ and, on the other hand, cutaneous polyarteritis nodosa presents mild extracutaneous symptoms, like fever, general malaise, diffuse myalgia and arthralgia, frank arthritis, sensitive peripheral neuritis, and distal arterial involvement of hands and feet, in about 60% of cases¹⁻² has been a source of confusion. Both entities have common pathological features: necrotising vasculitis affecting medium sized arteries that, in cutaneous polyarteritis nodosa, are located typically in hypodermis and deep dermis without visceral involvement. The clinical course and prognosis of cutaneous and systemic polyarteritis nodosa differ essentially: visceral extension is not associated with cutaneous polyarteritis nodosa and fatal outcome has not been reported.

Distal arteriopathy was reported in the initial series by Borrie.¹ Two of their patients had incipient gangrene of the toes; one toe was affected in one case and all in the other. Toes were cold, grossly cyanosed, and extremely painful. Other authors have reported distal arteriopathy associated with cutaneous polyarteritis nodosa, manifested by ischaemic pain, pallor, coldness, necrotic ulcers, or gangrene.³⁻⁵ Our case is, to our knowledge, the first report of distal arterial disease associated with cutaneous polyarteritis nodosa with a clinical presentation as Raynaud's phenomenon.

The arteriographic findings of distal arteriopathy in cutaneous polyarteritis nodosa have been described by several authors⁴⁻⁵ as distal

arteritis with narrowing of the vessel lumen and absence of filling; it is noteworthy that proximal lesions of larger vessels were not noted. This is useful for the differential diagnosis with other forms of vasculitis or arteriosclerosis with distal involvement due to embolism from proximal lesions.

Our case suggests some interesting questions about the clinical spectrum of medium sized necrotising vasculitis. Extracutaneous disease in cutaneous polyarteritis nodosa usually accompanies flares of cutaneous lesions. This case is distinctive since Raynaud's phenomenon preceded cutaneous manifestations of the disease by years. Furthermore, the two manifestations had an independent course during follow up. Probably, our patient has a limited form of polyarteritis nodosa confined to skin and distal arteries, and should be placed in the other well known group of limited polyarteritis nodosa with involvement of gallbladder or appendix.^{10 11} Cutaneous polyarteritis nodosa should be included in the differential diagnosis of Raynaud's phenomenon, and Raynaud's phenomenon should be added to the clinical

picture of extracutaneous disease of cutaneous polyarteritis nodosa.

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