Necrotizing Vasculitis Due to a Severe Type II Leprosy Reaction

Moussa Diallo M¹*, Diack N. D², Diatta B. A¹, Ndiaye A¹, Dieng M. T¹

¹Department of Dermatology, Teaching Hospital Aristide LeDantec, Dakar, Senegal.
²Department of Internal Medicine, Teaching Hospital Aristide LeDantec, Dakar, Senegal.

*Corresponding author: Moussa Diallo

Abstract

Introduction: Despite the wide spectrum of the clinical manifestations of leprosy, necrotizing cutaneous vasculitis has rarely been reported as a manifestation of this disease. We report a case of cutaneous vasculitis due to a type II leprosy reaction, secondary to lepromatous leprosy. Case report: A 38-years-old Gambian man was admitted for necrotic and infiltrated purpura of acral distribution (limbs, ears and nose), which was associated with multiple subcutaneous inflammatory nodules, epistaxis, polyarthralgia, fever and a poor general health status. Mycobacterium Leprae was isolated from the nasal mucosa (bacillary index 3 +) and the biopsy of the purpuric lesions revealed a leukocytoclastic vasculitis. A multidrug therapy concurrently with oral corticosteroids was started with good clinical results after 4 weeks. Discussion: Cutaneous necrotizing vasculitis has been described as a rare manifestation of erythema nodosum leprosum in medical literature. We think, in endemic areas, erythema nodosum lepromatous can be added to the list of the possible etiologies of cutaneous vasculitis.

Keywords: Mycobacterium Leprae, Leprosy, Cutaneous Vasculitis.

Introduction

Leprosy is a chronic infection caused by the Mycobacterium leprae with a wide range of clinical manifestations depending on the immune status of the patient [1]. Vasculitis is a nonspecific term that encompasses a large and heterogeneous group of disorders that are characterized by inflammation of blood vessels. Systemic vasculitis occurs in a heterogeneous group of primary disorders or it can be a manifestation of infection, an adverse drug reaction, malignancy or a connective tissue disease [2].

In our regions, infections with hepatitis B and HIV viruses are more commonly implicated. Apart from the Lucio's phenomenon, the occurrence of systemic vasculitis in Hansen's disease has rarely been described in medical literature [3, 4, 5]. We report a type II leprosy reaction which manifested as cutaneous necrotizing vasculitis secondary to lepromatous leprosy.

Case report

A 38-years-old Gambian man, without any significant past medical history, was admitted in our department for purpuric and necrotic lesions on the extremities which were associated with polyarthralgia, fever and a significant deterioration of the general health status one month prior to presentation.

Three years ago, the patient had experienced painful subcutaneous nodules on both upper and lower limbs, associated with fever, evolving by relapses and remissions. Additionally, there was a history of epistaxis, a serous rhinorrhea and peripheral arthritis.

Physical examination at admission revealed the necrotic and infiltrated purpura with acral distribution, involving the limbs (Fig.1), ears and nose (Fig. 2) which were associated with disseminated subcutaneous inflammatory nodules, predominantly in the upper limbs (Fig. 3). Crusted nasal erosions and infiltrated papules on ears and forehead were also observed. The neurological examination showed a bilateral and symmetrical decrease in touch sensation in lower limbs. Signs of orchitis were noticed in genitourinary examination.
The temperature was at 38.3 °C, with a heart rate of 104 / min. The rest of the physical examination was within normal ranges. Laboratory investigations showed a non-specific inflammatory syndrome with an ESR of 100 mm/hr, hyperfibrinogenemia of 6.6 g/l, microcytic anemia with a Hb of 8.3g/dl, a thrombocytosis of 541000/mm3 and leukocytosis at 16000/mm3 with neutrophilic predominance. Serological tests for hepatitis B, C and HIV, as well as antinuclear antibody and ANCA were negative.

Figure 1: Infiltrated and necrotic purpura of legs

Figure 2: Infiltrated purpura on ears

Figure 3: Infiltrated subcutaneous nodules of limbs

Mycobacterium Leprae was detected in slit nasal smears with a bacillary index of 3+. Biopsy of skin purpuric lesions revealed a leukocytoclastic vasculitis of small superficial dermal vessels.

The histopathologic examination of subcutaneous nodules showed a polymorphous granulomatous infiltrate involving mainly interlobular septa of subcutaneous fat tissues, which contained acid-fast-bacilli in Ziehl-Neelsen staining.

The patient underwent treatment with multidrug therapy (Rifampicin, Dapsone, Clofazimine) and systemic corticosteroid therapy with methyl prednisolone 1mg/kg/day.

After four weeks of treatment the temperature returned to normal, the purpuric lesions and subcutaneous nodules were cleared with post inflammatory hyperpigmentation.
Discussion

We report a case of erythema nodosum leprosum (ENL) which clinically presented as cutaneous necrotizing vasculitis. The infiltrated papular lesion of the earlobes and living in an endemic area were our clues to raise the diagnosis of leprosy which was confirmed by histological and bacteriological evidences.

The type II leprosy reaction or ENL is a hypersensitivity reaction occurring in 5-25% of cases of lepromatous forms of leprosy (LL) [1, 6]. In 90% of cases it follows the introduction of the treatment, but it can also occur suddenly, as it was the case for our patient. The pathogenesis of type II leprosy reaction is bacterial lysis due to the treatment, which causes the formation of circulating immune complexes depositing in the interlobular septal vessels of subcutaneous fat tissues, resulting clinically in the occurrence of subcutaneous nodules [3, 6]. However, a clinical presentation of this process as a cutaneous necrotizing vasculitis has very rarely been reported in the medical literature [7].

In general, the occurrence of a systemic vasculitis in Hansen's disease is very rare [7]. What is more reported is the Lucio's phenomenon which manifests as necrotizing and ulcerated lesions [4, 8]. It is more frequent in Latin America and is very rare in African patients. The delayed diagnosis in our patient was, in part, explained by the unusual clinical presentation. Indeed, the nasal and articular manifestations made other differential causes of vasculitis such as Wegener's granulomatosis and peri arteritis nodosa, more likely initially, which were finally ruled out by the negativity of hepatitis B and ANCA serological tests as well as histopathologic evidences. On the other hand, this delayed diagnosis may also explain the severity and the dissemination of the necrotic lesions, as well as the severe deterioration of the general condition in our patient.

According to WHO, the treatment of ENL is based on systemic cortico therapy, plus standard anti-leprosy multidrug therapy [1, 9]. The efficacy of this protocol was proven in our patient with a favorable outcome within a month.

Conclusion

The clinical presentations of Hansen's disease are variable and some of them such as cutaneous necrotizing vasculitis can be misleading. Our observation emphasizes the importance of raising the diagnosis of erythema nodosum leprosum as an underlying cause of systemic vasculitis in endemic areas.

References