Case Report

Etiologic Dilemma and Challenges of Management of Nodular Vasculitis in Tertiary Health-Care Facility: A Case Report

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Abstract

Nodular vasculitis is a rare inflammatory disease of the skin and subcutaneous fat tissue, characterized by crops of small, tender, erythematous nodules on the legs, mostly on the calves and shins. We present a 17-year-old adolescent female who presented with a six-month history of cough; recurrent fever and bilateral lower limb multiple ulcerated nodules of 1-month duration. Clinical examination revealed generalized lymphadenopathy with bilateral pitting leg edema which had multiple nodules and discoid ulcers extending from the groin to the ankles and discharging purulent fluid. Tests for human immunodeficiency virus and tuberculosis were negative. Histology of nodule biopsy revealed extensive caseous and coagulative fat necrosis, granulomatous inflammation with epitheloid macrophages and multinucleated giant cells surrounding the necrosis, and lymphoid infiltration of vessel walls with fibrous thickening of the intima, typical of Whitfield-type erythema induratum. There was initial but very transient response to antibiotic treatment, with further deterioration and eventual death from overwhelming sepsis.

Keywords: *Erythema induratum, Mycobacterium tuberculosis, nodular vasculitis, panniculitis*

Introduction

Nodular vasculitis (NV) is an uncommon inflammatory disorder of the skin characterized by crops of small, tender, erythematous nodules on the legs, and lobular panniculitis with granulomatous inflammation, vasculitis, and focal fat necrosis on histology.[1] The term "nodular vasculitis" is often used interchangeably with erythema induratum, although both are historically considered different entities.[2]

Two variants of the nodular vasculitis complex have been described; erythema induratum of Bazin (EIB) associated with tuberculosis[2] and non-tuberculous erythema induratum (NV) described by Whitfield.[3] EIB is the commoner type and has been previously reported in African children. One of such reports was in a 10-year-old Kenyan child[4] and the other was in a 6-year-old Nigerian girl living in far-away Ireland.[5] In both cases, diagnosis was confirmed by biopsy and histology of skin lesions and both responded positively to anti-tuberculosis therapy.[4,5] On the other hand, reports on the Whitfield type in children remain rare in Africa and globally. Due to the diverse etiologies and overlapping morphological features, diagnosis of these conditions remains quite challenging,[1] We present this rarer form of non-tuberculous erythema indurtum (NV) in an adolescent female to highlight the diagnostic dilemma and the challenges of management of this rare disease.

Case Report

A 17-year-old female presented with history of cough and fever of 6 months; weight loss, multiple skin lesions, and swelling of both lower limbs of 1-month duration. Cough was productive of whitish sputum. The skin lesions initially were painful nodules, but subsequently ulcerated, oozing malodorous fluid. She reported a history of weight loss but there was no night sweats

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or contact with persons with tuberculosis or chronic cough. She received all scheduled vaccines at infancy. She belonged to the low socioeconomic class, lived in a single poorly ventilated room with her mother and three siblings, and predominantly engaged in subsistent farming with her mother.

Clinical findings were an adolescent female in painful distress, pyrexic, pale, with significant generalized lymphadenopathy involving the submental, submandibular, bilateral axillary (apical), and bilateral inguinal group of lymph nodes and bilateral pitting leg edema. Vital signs were temperature 40°C, pulse rate of 124 beats per minute, respiratory rate of 24 breath per minute, and blood pressure of 110/70 mmHg. Both lower limbs had multiple nodules of varying sizes (1 cm × 1 cm to 3 cm × 4 cm), multiple discoid ulcers extending from the groin to the ankles. Ulcers were discharging purulent fluid [Figure 1]. No abnormalities were detected on other systemic examination. A presumptive diagnosis of disseminated tuberculosis (pulmonary and cutaneous) was considered.

Complete blood count showed neutrophilia, anemia, and elevatederythrocyte sedimentation rate of 105 mm/1 h. Chest X-ray showed no evidence of active or latent pulmonary tuberculosis. Gene Xpert for tuberculosis and tuberculin skin test were negative. Screening for retroviral disease, Hepatitis B and C screening were negative. Wound swab microscopy, culture, and sensitivity (m/c/s) were positive for gram-negative bacilli sensitive to Imipenem only.

Histopathology of tissue taken from the lesion revealed extensive caseous and coagulative fat necrosis confined to the lobules and sparing the septa with granulomatous inflammation. There were epitheloid macrophages and multinucleated giant cells of foreign body type forming broad zones of inflammation surrounding the necrosis, extensive, severe vascular changes, dense lymphoid infiltration of vessel walls with endothelial swelling and edema of vessel wall, and fibrous thickening of the intima [Figures 2 and 3]. The necrosis involved the dermis with subsequent ulceration. These features confirmed the diagnosis of erythema induratum of Whitfield. Autoantibody tests were not done as diagnosis of this condition is basically made on histology finding.

She received IV Imipenem for 2 weeks with remarkable improvement [Figure 4]. Subsequently, she was discharged to continue care as outpatient. Three weeks later, the skin lesions reappeared and she was readmitted. Imipenem was recommenced based on results of a repeat wound m/c/s. Her condition continued to wax and wane for 2 months until her demise.

Figure 1: Arrows show ulcerated nodules on presentation, discharging purulent matter

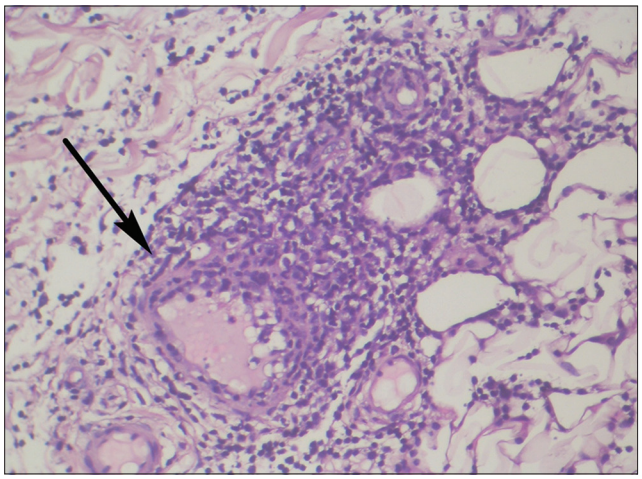

Figure 2: Arrow shows necrosis involving fat lobules with epitheloid macrophages and multinucleated giant cells surrounding the necrosis

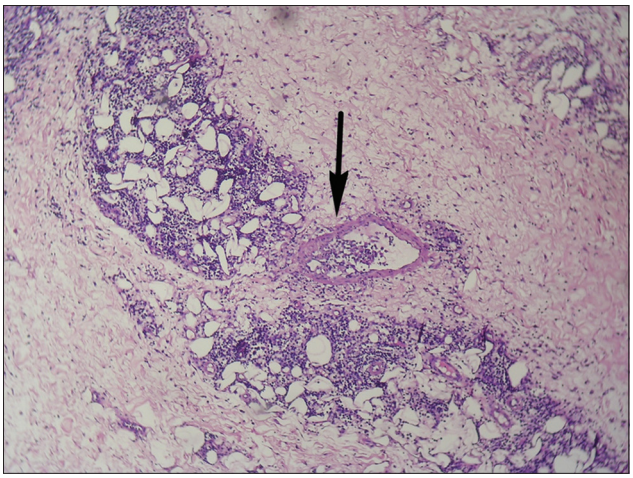

Figure 3: Arrow shows lymphoid infiltration of vessel walls and fibrous thickening of the intima

Discussion

Historically, erythema induratum has been described as

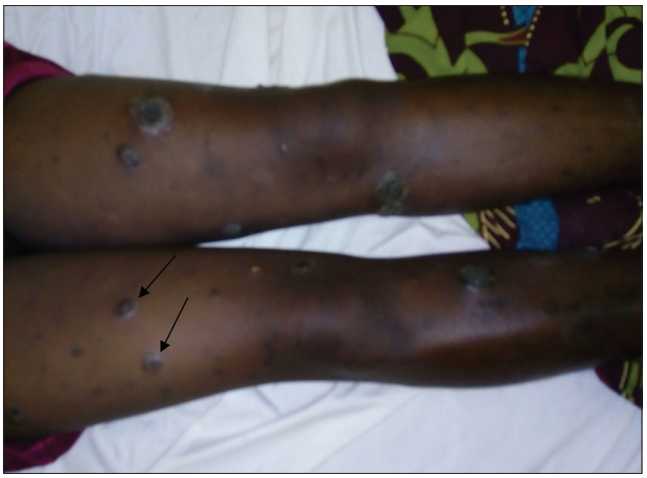

Figure 4: Arrows show healing nodules following antibiotic therapy

a disease characterized by chronic nodular eruption that usually occurs on lower legs of young or middle-aged women strongly associated with *Mycobacterium tuberculosis* infection.[2,6] Cases of EI have also been reported in adolescent males[7] and in older women with comorbid conditions.[8]

Although EI was initially associated with pulmonary tuberculosis, Whitfield[3] and Galloway[9] in early 1900s distinguished cases of EI unrelated to *M. tuberculosis* infection as a separate clinical entity. Years later, Montgomery and colleagues[10] described this entity as "nodular vasculitis."

The Bazin type was first described in 1855.[8] EIB, regarded as a tuberculid, is a cutaneous hypersensitivity reaction to an antigen of extracutaneous focus.[2] This type is commoner in women less than 25 years old.[3] Diagnosis of EIB is based on cutaneous characteristics, a positive Mantoux test, evidence of tuberculosis and histopathological findings.[2] Often times, it is clinically difficult differentiating EIB from erythema induratum from other non-tuberculous causes. At first, the index case posed a diagnostic challenge to both pediatricians and dermatologists. It is known that panniculitis poses a diagnostic challenge to clinicians and histopathologists due to its variable histopathologic appearance and overlapping morphological features, which most times depends on the duration of the lesions, the sites from which biopsy specimens are taken and dynamic nature of the lesions.[1] The histopathological findings without the evidence of tuberculous infection informed the diagnosis of NV in the index case.

Vasculitis in the West African subregion is precipitated by infections and attributable to climatic and ecological conditions.[11] Socio-economic status and environmental factors which are important factors in the occurrence of skin diseases contribute to this condition. Overcrowding and poor hygiene prevail in most suburban and rural housing. The index patient shared a single poorly ventilated room with her mother and three siblings. Amer and Metwalli[11] reported that rural workers were susceptible to skin injuries that often become infected.

Atypical mycobacterium such as *Mycobacterium chelonei* and *Mycobacterium monacense* has also been implicated in causation of EI.[6] Although, *M. chelonei* is best known for its pathogenicity in immunocompromised hosts and has been reported secondary to traumatic implantation, Campbell *et al.*[6] reported a case of EI caused by *M. chelonei* in an immunocompetent middle-aged woman with no history of trauma which responded to antibiotic therapy. The index case predominantly engaged in farming with her mother but there was no history of skin injuries prior to the onset of the skin lesion.

NV runs a chronic course, not readily amenable to treatment. Prognosis is better when the patient presents early without ulcerations, and the actual cause of the disease is known as in the case of tuberculosis-associated erythema induratum.

A major challenge in management in the index case was late presentation (1 month after ulceration of lesions was noticed), as well as difficulty in arriving at the primary cause of the disease.

Conclusion

This case highlights the diagnostic and management challenges associated with NV. Proper evaluation of the patient is required to make accurate diagnosis in a such difficult circumstance.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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