PAMJ Clinical Medicine
Pan African Medical Journals

Case report

Diagnostic difficulty-an elusive case of Hansen´s disease mimicking sarcoidosis

Erere Otrofanowei[1,&], Olusola Olabisi Ayanlowo[1], Ayesha Omolara Akinkugbe[1], Vincent Onyekwelu[2]

[1]Department of Medicine, College of Medicine, University of Lagos, Lagos University Teaching Hospital, Lagos, Nigeria, [2]Department of Anatomy and Molecular Pathology, Lagos University Teaching Hospital, Lagos, Nigeria

[&]Corresponding author: Erere Otrofanowei, Department of Medicine, College of Medicine, University of Lagos, Lagos University Teaching Hospital, Lagos, Nigeria

Received: 11 Oct 2019 - Accepted: 16 Mar 2020 - Published: 19 Mar 2020

Domain: Dermatology, Infectious disease, Public health

Keywords: Hansen´s disease, leprosy, sarcoidosis, Nigeria

Abstract

Hansen´s disease (leprosy) is caused by a slow-growing type of bacteria called *Mycobacterium leprae (M. leprae)*. It is an age-old disease that has been around since biblical times yet cases still occur especially in Asia and Africa, despite concerted global efforts to eradicate the disease. Chemotherapeutic agents are available and effective once administered appropriately and adequately. We report an elusive case of Hansen´s disease wherein the only symptom was a nodule on the nasal bridge in an otherwise healthy woman for over one year. Repeated examinations and investigations in different centers were suggestive of sarcoidosis and she was left untreated for leprosy for the period. A skin biopsy was carried out for histopathology diagnosis. Histology confirmed borderline lepromatous leprosy. This case demonstrates the need for a definitive diagnosis of leprosy to reduce the spread of this contagious disease especially in tune with the concerted global efforts.

Case report | Volume 2, Article 122, 19 Mar 2020 | 10.11604/pamj-cm.2020.2.122.20622

Available online at: https://www.clinical-medicine.panafrican-med-journal.com/content/article/2/122/full

Introduction

Hansen´s disease-also known as leprosy is a chronic infectious disease caused by a bacterium *Mycobacterium leprae* and it is well known for its stigma and lasting sequelae. Eradication of this disease has been a target of the World Health Organisation (WHO) for decades and whilst successes have been recorded, a few largely populated countries still record high prevalence and incidence rates. This may be related to both late and difficult diagnoses. This case report highlights a typical example of the latter with an initial diagnosis of sarcoidosis and emphasizes the need for a highly specific and sensitive laboratory tests to achieve total eradication.

Patient and observation

A 68 year old female petty trader from the south-western parts of Nigeria was referred from another centre with a single non-itchy, non-painful rash on the bridge of her nose that had slowly increased in size to involve her upper lip over a 1 year period. The only associated symptom was rhinorrhea. There were no lesions on any other part of the body. There was neither a positive family history nor other contact history of similar lesions. On physical examination, she had a flesh-coloured plaque on the inferior end of the nasal bridge involving the tip of the nose and the philtrum, effectively flattening the septum and causing a saddle-nose deformity (Figure 1). There were few smaller flesh-coloured plaques on the malar region of the right half of her face. There was a non-tender solitary hyperpigmented plaque on the right nape posteriorly, about 6cm in size and was not scaly. All lesions had intact sensation and no loss of skin appendages. All other systems were normal, though blood pressure was elevated at 180/80mmHg for which she was taking Lisinopril tablets. Significant investigation results were elevated erythrocyte sedimentation rate of 55mm/hr. Westergren and Angiotensin converting enzyme (ACE) levels in serum of 82.2 (8-52IU/L). Serology for human immunodeficiency virus (HIV), hepatitis B virus (HBV) and hepatitis C virus (HCV) were all negative. Chest radiographs taken were also normal. Repeated skin slit and smear examination (on both normal and lesional skin) revealed no acid fast bacillus. Skin biopsy carried out initially suggested Lichenoid dermatitis- "mild acanthosis and downward elongation of the rete pegs, lymphoplasmacytic infiltration of the papillary dermis and destruction of the stratum basalis of these cells. Melanin incontinence and regurgitation is seen". She was given Prednisolone tablets before presenting at our facility with little improvement. With a high index of suspicion, a repeat skin biopsy of the lesions eventually revealed aggregates of granuloma composed of epithelioid histiocytic cells and foamy macrophages within the superficial dermis. These granulomas are displayed in an "onion skinning" fashion, with surrounding chronic inflammatory cells consisting of lymphocytes, plasma cells and multi-nucleated Langhans giant cells (Figure 2). There were also demonstrable acid fast bacilli (AFB) special stains (Figure 3). Features were in keeping with borderline lepromatous leprosy. She was eventually started on monthly rifampin and clofazimine as well as daily dapsone, clofazimine and noticed significant improvement in her lesions within a month.

Discussion

Leprosy and sarcoidosis are both chronic granulomatous diseases with similar cutaneous manifestations that have confused physicians the world over for many decades [1,2]. The WHO puts the prevalence of leprosy at 192,246 at the beginning of 2011 with a reduction to 189,018 cases at the end of first quarter 2013 [3]. Though global statistics show a reduction in prevalence, there are still high incidence rates in endemic countries and Nigeria is one of those. Theoretically, it should be easy to eradicate leprosy as the aetiopathogenesis is well known unlike the multi systemic inflammatory disease, sarcoidosis. Leprosy however poses a problem in the endemic

areas because of the indolent nature of the *M. leprae* causal organism, the stigma associated with the disease which deters early presentation, poor diagnostic facilities and a low level of suspicion of early lesions by healthcare givers amongst other factors. Sarcoidosis can present with only skin manifestations-cutaneous sarcoidosis of Boeck, or affect more systems in the Besnier-Boeck-Schaumann syndrome [4]. Skin lesions are seen in almost 25% of sarcoid patients and can be classified into specific or non-specific depending on the presence (or not) of non-caseating granulomas on histologic specimen [5,6]. The confusion between cutaneous sarcoid and leprosy is limited to tuberculoid leprosy as the lepromatous end of the spectrum has more distinct features both physically and histologically. There are actually some schools of thought that the mycobacterium causing leprosy may be an inciting agent of sarcoidosis [2,7], but this has not been generally accepted.

The diagnosis of leprosy is via a combination of compatible clinical features and the demonstration of characteristic caseating granulomas on histopathology carried out on well-developed skin lesions. The presence of acid fast bacilli in the dermis provides even more evidence but is not always seen especially in the tuberculoid end of the Ridley-Jopling spectrum [8]. Unfortunately, histopathology is not readily available in this part of the world and reliance on less sensitive tests as elevated ESR, skin slit and smears to demonstrate AFB (negative in 70% of cases) [9], lepromin tests, allows for delayed diagnosis and treatment which results in lepromatous presentation as cell-mediated immunity decreases. This is noted in the index case as initial tests carried out (when she had just a solitary hypopigmented patch on the face) were all negative. Serum ACE levels can be elevated in leprosy as well as sarcoidosis, but it is also seen in other diseases as histoplasmosis, Gaucher´s disease, HIV infections, lymphomas and so on. The need for early diagnosis of leprosy cannot be over emphasized and a new diagnostic tool for this is being developed by the U.S.-based Infectious Disease Research Institute (IDRI) and OrangeLife, a Brazilian medical products company [10]. It is said to yield results in less than 10minutes in a manner similar to a pregnancy test kit which makes it easy to use by community clinics and primary health care workers. If this is made available worldwide, especially to endemic countries, the world may well be on its way to eradicating this age-old disease neglected tropical disease.

Conclusion

The initial presentation of leprosy in the index patient was subtle enough to elude diagnosis both clinically and with investigations. This contributed to worsening of symptoms and the possibility of increased disease transmission. Recognition of early symptoms, rapid and reliable means of diagnosis are to be emphasized and promoted at the community clinics and primary healthcare centres to achieve a reduced incidence rate in endemic countries.

Competing interests

The authors declare no competing interests.

Authors' contributions

Otrofanowei Erere: initiation of case, biopsy and histopathology of lesion, detection of previous care and liaison with prior clinicians, follow up of patient and write up of case report; Ayanlowo Olusola: detection of case, follow-up of patient, participation of case write up; Akinkugbe Ayesha: follow up of the patient to date, participation of case write up; Onyekwelu V: repeated histopathology of biopsied specimen, participation of case write up. All the authors have read and agreed to the final manuscript.

Acknowledgements

We acknowledge the patient for her doggedness and patience in allowing us to reach a definite diagnosis.

Figures

Figure 1: flesh coloured plaque on philtrum, depressed nasal septum and flesh coloured plaques on the right malar area (left picture); right picture shows resolution of plaque with minimal residual hypopigmentation on the philtrum and right malar area of the same patient on completion of multi drug therapy for Hansen´s disease; the nasal septum is however still damaged

Figure 2: micrograph showing the granulomas at x400 magnification; "onion-skinning" The Langhans giant cells, epithelioid histiocytes and surrounding chronic inflammatory cells are better seen

Figure 3: micrograph showing red rods of acid fast bacilli against the blue background; Ziehl-Nielsen special stain

References

1. Tatem GB, Bash B, Morris ZQ. Leprosy mistaken as sarcoidosis in large referral sarcoidosis clinic. CHEST Journal. 2005;128(4):424S-424S. **PubMed** | **Google Scholar**

2. Wade H. Leprosy and Sarcoid the Kveim Test in Leprosy Patients and Contacts1. Journal of Investigative Dermatology. 1951;17(6):337-347. **PubMed** | **Google Scholar**

3. Organization WH. WHO Expert Committee on Leprosy. World Health Organization technical report series. 2012;(968):1. **PubMed** | **Google Scholar**

4. Irgang S. Sarcoid of Boeck: Report of a Case of Generalized Cutaneous Distribution and Pulmonary Involvement, with Clinical Cure with Tuberculin. Archives of Dermatology and Syphilology. 1939;40(1):35-44. **PubMed** | **Google Scholar**

5. Mangas C, Fernández-Figueras MT, Fité E, Fernández-Chico N, Sàbat M, Ferrándiz C. Clinical spectrum and histological analysis of 32 cases of specific cutaneous sarcoidosis. Journal of cutaneous pathology. 2006;33(12):772-777. **PubMed** | **Google Scholar**

6. English J, Patel PJ, Greer KE. Sarcoidosis. J Am Acad Dermatol. 2001;44:725-743. **PubMed**

7. Kissmeyer A, Nielsen J. Etiology of Boeck's Sarcoid. Arch Dermat & Syph. 1934;30(July):116.

8. Ridley D. Histological classification and the immunological spectrum of leprosy. Bulletin of the World Health Organization. 1974;51(5):451. **PubMed** | **Google Scholar**

9. Britton WJ, Lockwood DN. Leprosy. Lancet (London, England). Apr 10 2004;363(9416):1209-1219. **PubMed**

10. The Global Health Biotech. **The need**. Accessed January 8 2015.

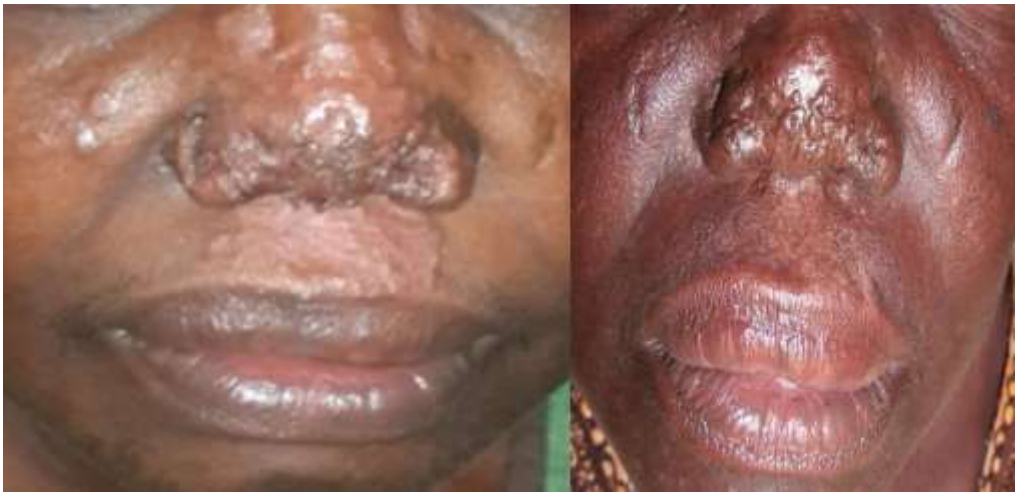

Figure 1: flesh coloured plaque on philtrum, depressed nasal septum and flesh coloured plaques on the right malar area (left picture); right picture shows resolution of plaque with minimal residual hypopigmentation on the philtrum and right malar area of the same patient on completion of multi drug therapy for Hansen´s disease; the nasal septum is however still damaged

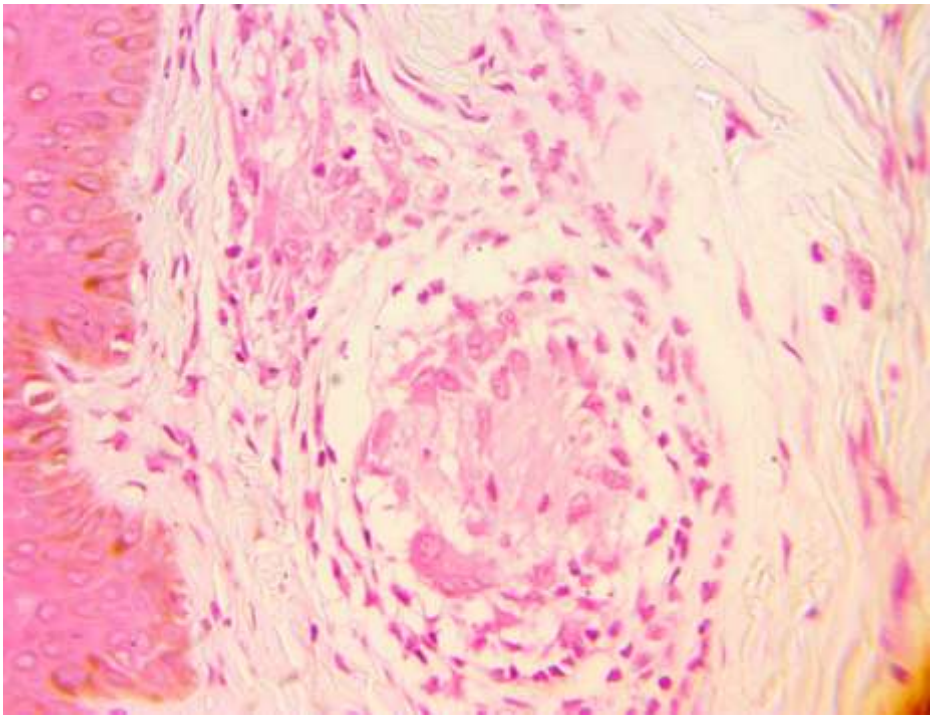

Figure 2: micrograph showing the granulomas at x400 magnification; "onion-skinning" The Langhans giant cells, epithelioid histiocytes and surrounding chronic inflammatory cells are better seen

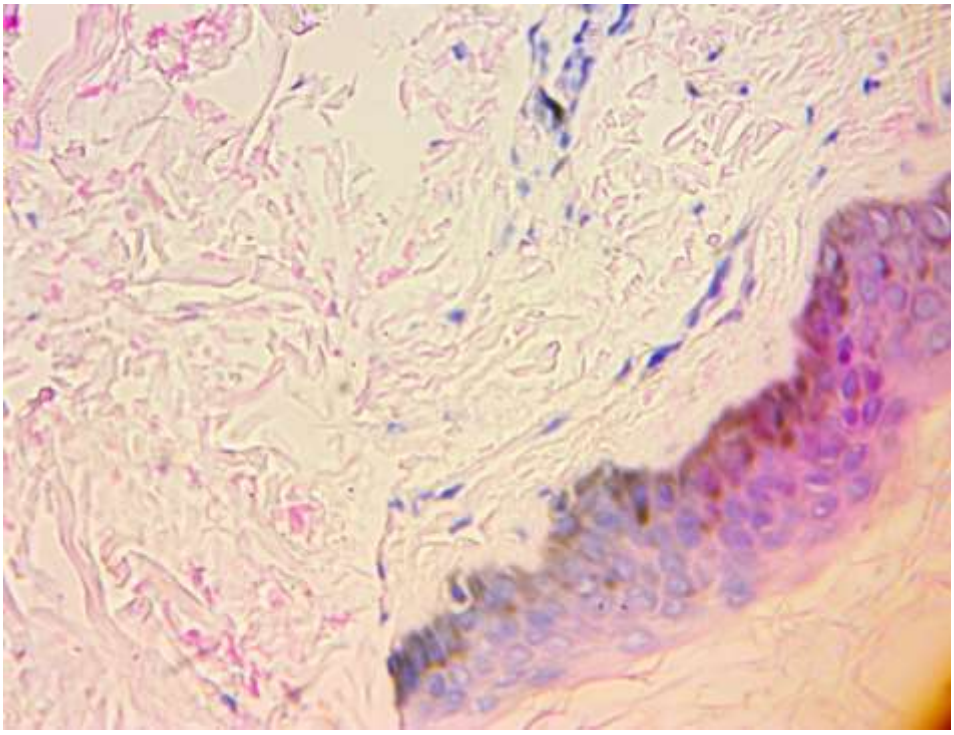

Figure 3: micrograph showing red rods of acid fast bacilli against the blue background; Ziehl-Nielsen special stain