Mycobacterial Diseases
ISSN: 2161-1068

Agbenorku and Saunderson, J Mycobac Dis 2013, 3:3
DOI: 10.4172/2161-1068.1000133

Review Article Open Access

Mycobacterium ulcerans Disease of the Face: The Fate of the Victims

Pius Agbenorku[1]* and Paul Saunderson[2]

[1]*Kwame Nkrumah University of Science and Technology, Kumasi, Ghana*
[2]*American Leprosy Missions, USA*

Abstract

Mycobacterium ulcerans disease (MUD), often called Buruli ulcer (BU), can result in severe disabling of affected persons. It affects all parts of the body with the upper and lower extremities as the most affected areas. BU causes disabilities such as scarring, limb contractures, disfigurements, as well as a great deal of distress if it affects the face. Patients with BU of the face encounter both physical and psychological problems that make their living difficult. The disease on the face can cause loss of eyes, nose, lips, repositioning of facial features and destruction of other soft tissues of the face. This study aims to discuss the fate of persons with MUD of the face. Patients lives MUD of the face should be given physical, social, economic and emotional support. This form of support should span from health care providers, families and friends to enhance their quality of life. Physiotherapy is also of great importance to both patients and health workers, to achieve a better physical appearance and psychological wellbeing.

Keywords: *Mycobacterium ulcerans* disease; Buruli ulcer; Face; Disability; Effects; Fate

Introduction

Buruli ulcer (BU) caused by *Mycobacterium ulcerans* (*M. ulcerans*), represents the third most common mycobacterial disease in the world after tuberculosis and leprosy [1]. It has emerged dramatically over the past two decades, particularly in Central and West Africa and has been confirmed by laboratory tests in 26 countries with reports in other countries around the world [2,3]. BU is a severe disfiguring and disabling disease, which affects primarily children less than 15 years of age in many tropical and subtropical countries [3-5]. Globally, about 35% of lesions occur on the upper limbs, 55% on the lower limbs and 10% on the other parts of the body [5].

BU lesions on the face have been reported in different countries, but there is little information on their frequency [3,6-11]. The difficulties in treating BU of the face and the importance of a conservative surgical approach to its management are essential factors in BU disability prevention [3,6-11]. This paper seeks to reveal the challenges of patients with BU of the face.

Epidemiology

M. ulcerans infection was first described in Australia in 1948 [12]. The disease was named after Buruli County in Uganda now called Nakasongola District, because of the many cases that occurred there in the 1960s [13,14]. The disease is endemic to humid, often rural, tropical climates and has been reported in 33 countries with tropical and subtropical climates [15-17]. The incidence of BU is highest in Africa especially in Benin, Cote d'Ivoire and Ghana, though cases also occur in Asia, South America, Papua New Guinea and Australia, where the disease is known as Bairnsdale ulcer [1,16-22].

Transmission

BU is found in many places around the world, often near water bodies but the exact mode of transmission of the germ from the environment to humans is not known [23]. However, it appears that different modes of transmission occur in different geographic areas and epidemiological settings [5]. BU has been observed more frequently during the rainy season in Africa and exposure may occur in muddy farming fields [1,24,25]. In March 2008, researchers announced the first isolation of *M. ulcerans* from the environment [26]. The available data suggests that the disease is transmitted from aquatic areas in the environment, rather than from person to person [6,19,27]. The disease is usually found in communities near rivers, swamps and wetlands [6,23]. The recent identification of *M. ulcerans* in certain water insects has raised the possibility of mechanical transmission of the infection [5,6,19].

Pathogenesis

Even though a particular vector of the disease has not been found, aquatic insects, notably naucoridae spp, adult mosquitoes or other biting arthropods may serve as vectors of *M. ulcerans* [5,28]. BU often starts as a painless nodule in the skin and infection often leads to extensive destruction of skin and soft tissue with the formation of large ulcers usually on the legs or arms [29,30] but no nodular stage was recorded on patients with BU of the head and neck region [30]. In early or pre-ulcerative lesions, *M. ulcerans* produces a lipid toxin, mycolactone, which is responsible for necrosis of the dermis, panniculus and fascia culminating in extensive ulceration [31]. The high prevalence of oedematous lesions among children may be attributed to multiple factors including immunity because children may have less effective protective immune response against *M. ulcerans* [1]; this is linked with the role that mycolactone plays in the pathogenicity of MUD [31-33]. Other factors may include the physical aspects of children since they have shorter stature and their entire body is nearer to the ground [10,34]. Delayed treatment may cause irreversible deformity and long-term functional disability but antibiotic treatment can be successful when patients seek treatment in the early stage of the disease [5].

***Corresponding author:** Pius Agbenorku, Reconstructive Plastic Surgery & Burns Unit, Komfo Anokye Teaching Hospital, School of Medical Sciences, College of Health Sciences, Kwame Nkrumah University of Science and Technology, Kumasi, Ghana, Tel: +233 20 630 0781; Fax: +233 51 22307; E-mail: pimagben@yahoo.com

Received October 09, 2013; **Accepted** November 21, 2013; **Published** November 27, 2013

Citation: Agbenorku P, Saunderson P (2013) *Mycobacterium ulcerans* Disease of the Face: The Fate of the Victims. J Mycobac Dis 3: 133. doi:10.4172/2161-1068.1000133

Diagnosis

The combination of polymerase chain reaction (PCR) and other methods such as histopathology, Ziehl-Nielsen (ZN) staining for acid-fast bacilli test and wound culture are necessary to make diagnosis [34-36]. Fine needle aspiration (FNA) is used for diagnosis of facial MUD patients with pre-ulcerative lesions, swabs are taken from early ulcerative lesions and specimens of tissue are obtained during debridement [34]. Punch or surgical biopsies from pre-ulcerative forms should be avoided as this may become the starting point of extensive ulceration [12,34]. Other diagnostic signs are based on clinical findings such as chronicity of the wound, typical undermined edges with central necrotic tissues, non-response to traditional wound management procedures and antibiotic therapy [30].

Effects: the fate of the victims

MUD in the face may lead to serious sequelae such as ulcerative destruction of the eyelids, loss of eyeball, loss of part of the ear, loss of part the nose, loss of part of the lower lips and deformity of the face in general [30,37]. The involvement of the maxillar and mandibular areas can also create functional problems for speaking, eating and drinking [34]. BU disabilities such as visual impairment and psychological effects due to facial scars associated with the disease and also surgeries on the face makes patients incapacitated in almost all aspects of their life, especially when the patient is a child [34,37,38]. Children are faced with psychological problems such as low self-esteem, shyness, and inferiority complex which could affect them in their education whilst adults may encounter problems in marriage, work and their social life as a whole [30].

The eyes

Scars on the face resulting from BU can cause the lower eyelid to evert (ectropion) and adhesions to adjacent structures can make it difficult for full eye closure (Figure 1).

It can cause the eyelids to invert (entropion), creating problems with eyelashes turning in and touching the cornea (trichiasis) [35]. Special surgical procedures should be given to patients whose eyes are been removed during the initial excision to ensure that a prosthesis can be fit in the future because, children especially develop asymmetric face when they are not fitted with prosthesis [35]. BU has permanently or partially disabled patients before and after the treatment procedures [24] by lose of eyeballs, eyelids, eyelashes and rendering patients totally or partially blind (Figures 2 and 3).

In some situations BU may affect the facial structures and yet may not lead to blindness. However, the consequence may be very bad scars in the face making the patient very uncomfortable in society, leading to withdrawal from social activities (Figures 4-6).

Figure 1: BU affecting the left eyelid. Source: Authors.

Figure 2: BU of the right eye, with complete destruction of the eye ball and extending into the surrounding soft tissue. Adapted from Agbenorku 2011 [30].

Figure 3: BU of eyes, the nose and other soft tissue of the face. Adapted from Agbenorku et al. 2012 [23].

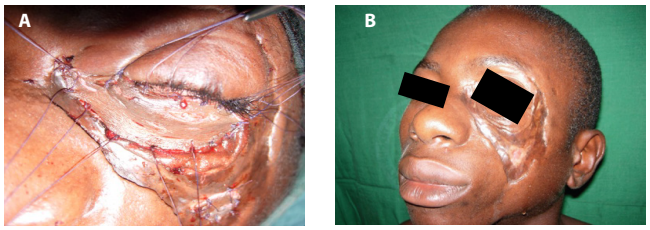

Figure 4: BUD of the left eye after excision and skin grafting. **A.** Intra-surgery on the face of a BU patient. **B.** Post surgery in Figure 4A-scarring of face. Adapted from Agbenorku 2011 [30].

The nose

MUD of the face can cause loss or repositioning of the nose of infected person making breathing difficult for such patients.

The ears

BU of the ear results in disabilities such as loss of earlobes. Further extensions of ulceration into the ear may affect the ear drum and cause loss of hearing and eventually may result in deafness in patients.

Other soft tissues in the face

Patients with MUD on the cheek and other soft tissues of the face (Figures 5 and 6) are faced with severe disability when the disease gets to a severe ulcerative stage. Severe ulceration of soft tissues may lead

Figure 5: MUD of the face. **A.** Lesion destroyed the left eye leading to its surgical enucleation, affecting the right eye as well as the bridge of the nose resulting in horrible scars in the face. **B.** Same patient as in Figure 5A: Long term post-surgery with the sequelae of disfiguring of the face. Source: Authors.

Figure 6: BU on the cheek extending to the chin. Adapted from Agbenorku 2011 [30].

to exposure of the patients' jaw bones, teeth and loss of lips. This may result in difficulty in speaking, drinking and eating. Scarring may cause significant disfigurement of the face of patients.

Patients with MUD of the face as a result of their disability encounter many challenges. They are socially, emotionally and economically affected. Socially, persons with the disability may feel shy and uncomfortable in socialising with people because they fear to be shunned by them; this makes it difficult for adults to get married and for children to build self confidence. As a result, their entire self image and confidence is crushed and patients become lonely and timid, creating a whole lot of psychological effects. Patients may also find it difficult to engage in any economic activities due to social and psychological factors and this renders them economically inactive making their lives difficult and a burden on their relatives.

Treatment

The Global Buruli Ulcer Initiative (GBUI) of the World Health Organization has recently recommended the initial use of conservative treatment particularly when lesions are located on the face, breast, and genitalia. The suggested approach for treatment of such patients is a combination of specific antibiotics with or without surgery and to follow an appropriate prevention of disability (POD) program assiduously [4,39].

Large ulcers or extensive edematous lesions generally require excision followed by skin grafting [23,30]. Ulceration that extends to the eyelid and base of the nose are treated by four surgical excisions, daily wound dressings and skin grafts.

Ulcers closer to the eye cannot have complete excision. They are dressed over long periods with normal saline or 2% acetic acid lotions and the excised ulcers are grafted due to the difficulty in achieving good hemostasis. Patients with eyelid destruction have reconstruction with an Indian forehead flap. Sharp debridement can also be used to treat BU of the face that has good healthy edges with hypertrophic granulation and the wound is covered with split-thickness skin grafts or local transposition flaps after meticulous hemostasis and dressed with Vaseline gauze and tie-over dressing [30]. Surgery is combined with antimycobacterial chemotherapy (Rifampin and Streptomycin) for 8 weeks and the treatment demands long term hospitalization and follow-ups after discharge [13,30,38,40,41]. Preservation of vision is very important, so special attention must be given to the eyes. To prevent corneal dryness, ulceration and infection, the cornea must be lubricated with artificial tears and with ointment at night. The eyes must also be protected during the day with hats and glasses, and an eye shield at night to reduce dryness from exposure, and to protect the eye from dust and other foreign objects. Frequent exercise to close the eye helps lubricate the cornea, strengthen weakened muscles and maintain full movement for eye closure. Gentle massage over the scar area softens it, stretches it and limits spread of adhesions to adjacent structures, which can otherwise limit movement [35]. Interventions to prevent disability should start before excision and continue after excision and skin grafting in order to prevent soft tissue contractures. Prevention of disability and rehabilitation are only possible with the active participation of those affected by BU, their families, the community, and the health-care team [23]. The use of laser therapy in the treatment of facial scars has been reported [42]; however, none of the patients who have developed scars as a result of the disease as shown in the figures above has undergone laser therapy.

Conclusion

Severity of sequelae following *M. ulcerans* infection in the face causes significant deformities hence there should be early presentation of patients to health facilities for treatment. The surgical team should be extra cautious when operating on patients with MUD of the face to avoid disfiguring permanent scars and associated disabilities. Physiotherapy procedures for patients should be of high priority to both the patient and the health team. Patients should also be supported physically, emotionally and economically by relatives before and after surgery to ensure healthy living, emotional fitness and economic wellbeing.

References

1. Portaels F, Silva MT, Meyers WM (2009) Buruli ulcer. Clin Dermatol 27: 291-305.
2. World Health Organization (2001) Buruli Ulcer. Diagnosis of Mycobacterium ulcerans Disease. Portaels F, Johnson P, Meyers WM, eds. Geneva: World Health Organization, WHO/CDS/CPE/GBUI/2001.4.
3. Janssens P, Pattyn SR, Meyers WM, Portaels F (2005) Buruli ulcer: a historical overview with updating to 2005. Bull Seances Acad R Sci Outre Mer 51: 265-299.
4. World Health Organization (2004) Buruli ulcer disease. Mycobacterium ulcerans infection: an overview of reported cases globally. Wkly Epidemiol Rec 79: 194-200.
5. WHO Buruli ulcer (Mycobacterium ulcerans infection) updated on June 2013.
6. World Health Organization (2000) Buruli Ulcer-Mycobacterium ulcerans Infection. Asiedu K, Sherpbier R, Raviglione M, eds. Geneva: World Health Organization, /CDS/CPE/GBUI/2001.1.
7. Ouattara D, Aka GK, Meningaud JP, Sica A, Kaba L, et al. (2003) [Facial localizations of Buruli ulcers: two cases]. Rev Stomatol Chir Maxillofac 104: 231-234.
8. Kouame K, Ecra E, Cisse M, Gbery I, Kacou E, et al. (2008) [Buruli ulcer involving the head: outcomes and therapeutic aspects in 8 cases observed at the University Hospital Center in Abidjan, Cote d'Ivoire]. Med Trop (Mars) 68: 643-644.

9. Noeske J, Kuaban C, Rondini S, Sorlin P, Ciaffi L, et al. (2004) Buruli ulcer disease in Cameroon rediscovered. Am J Trop Med Hyg 70: 520-526.

10. Debacker M, Aguiar J, Steunou C, Zinsou C, Meyers WM, et al. (2004) Mycobacterium ulcerans disease: role of age and gender in incidence and morbidity. Trop Med Int Health 9: 1297-1304.

11. Phanzu DM, Ablordey A, Imposo DB, Lefevre L, Mahema RL, et al. (2007) Short report: edematous Mycobacterium ulcerans infection (Buruli ulcer) on the face: a case report. Am J Trop Med Hyg 77: 1099-1102.

12. Johnson PD, Veitch MG, Leslie DE, Flood PE, Hayman JA (1996) The emergence of Mycobacterium ulcerans infection near Melbourne. Med J Aust 164: 76-78.

13. Agbenorku P, Kporku H (2001) Buruli ulcer: a poverty disease? Indian J Clin Pract Int 3: 23-25.

14. Guédénom A, Zinsou C, Josse R, Andélé K, Pritze S, et al. (1995) Traditional treatment of Buruli ulcer in Bénin. Arch Dermatol 131: 741-742.

15. MacCallum P, Tolhurst JC, et al. (1948) A new mycobacterial infection in man. J Pathol Bacteriol 60: 93-122.

16. Sizaire V, Nackers F, Comte E, Portaels F (2006) Mycobacterium ulcerans infection: control, diagnosis, and treatment. Lancet Infect Dis 6: 288-296.

17. (1969) BCG vaccination against mycobacterium ulcerans infection (Buruli ulcer). First results of a trial in Uganda. Lancet 1: 111-115.

18. Merritt RW, Benbow ME, Small PLC (2005) Unraveling an emerging disease associated with disturbed aquatic environments: the case of Buruli ulcer. Frontiers in Ecology and the Environment 3: 323-331.

19. Oluwasanmi JO, Solankee TF, Olurin EO, Itayemi SO, Alabi GO, et al. (1976) Mycobacterium ulcerans (Buruli) skin ulceration in Nigeria. Am J Trop Med Hyg 25: 122-128.

20. Portaels F, Aguiar J, Debacker M, Guédénon A, Steunou C, et al. (2004) Mycobacterium bovis BCG vaccination as prophylaxis against Mycobacterium ulcerans osteomyelitis in Buruli ulcer disease. Infect Immun 72: 62-65.

21. Portaels F, Elsen P, Guimaraes-Peres A, Fonteyne PA, Meyers WM (1999) Insects in the transmission of Mycobacterium ulcerans infection. Lancet 353: 986.

22. Rapini RP, Bolognia JL, Jorizzo JL (2007) Dermatology: 2-Volume Set. St. Louis: Mosby. pp. Chapter 74.

23. Agbenorku P, Edusei A, Agbenorku M, Diby T, Nyador E, et al. (2012) Buruli-ulcer induced disability in ghana: a study at apromase in the ashanti region. Plast Surg Int 2012: 752749.

24. Revill WD, Barker DJ (1972) Seasonal distribution of mycobacterial skin ulcers. Br J Prev Soc Med 26: 23-27.

25. Rook A, Wilkinson DS, Ebling FJG (1979) Textbook of dermatology, Vol.1 (3rdedn), Oxford: Blackwell Scientific Publications.

26. Amofah GK, Sagoe-Moses C, Adjei-Acquah C, Frimpong EH (1993) Epidemiology of Buruli ulcer in Amansie West district, Ghana. Trans R Soc Trop Med Hyg 87: 644-645.

27. Barker DJ (1973) Epidemiology of Mycobacterium ulcerans infection. Trans R Soc Trop Med Hyg 67: 43-50.

28. Marsollier L, Robert R, Aubry J, Saint André JP, Kouakou H, et al. (2002) Aquatic insects as a vector for Mycobacterium ulcerans. Appl Environ Microbiol 68: 4623-4628.

29. Leonardo J (2000) PSEF International Scholar Sees Allies In Buruli Ulcer Battle. Plastic Surgery News.

30. Agbenorku P (2011) Multicenter study of buruli ulcer disabilities in the head and neck region. Plast Surg Int 2011: 647418.

31. Asiedu K, Sherpbier R, Raviglione M (2001) Buruli ulcer–Mycobacterium ulcerans Infection, vol. 1, World Health Organization, WHO/CDS/CPE/GBUI/2001.

32. En J, Goto M, Nakanaga K, Higashi M, Ishii N, et al. (2008) Mycolactone is responsible for the painlessness of Mycobacterium ulcerans infection (buruli ulcer) in a murine study. Infect Immun 76: 2002-2007.

33. Silva MT, Portaels F, Pedrosa J (2009) Pathogenetic mechanisms of the intracellular parasite Mycobacterium ulcerans leading to Buruli ulcer. Lancet Infect Dis 9: 699-710.

34. Phanzu DM, Mahema RL, Suykerbuyk P, Imposo DH, Lehman LF, et al. (2011) Mycobacterium ulcerans infection (Buruli ulcer) on the face: a comparative analysis of 13 clinically suspected cases from the Democratic Republic of Congo. Am J Trop Med Hyg 85: 1100-1105.

35. Herbinger KH, Adjei O, Awua-Boateng NY, Nienhuis WA, Kunaa L, et al. (2009) Comparative study of the sensitivity of different diagnostic methods for the laboratory diagnosis of Buruli ulcer disease. Clin Infect Dis 48: 1055-1064.

36. Centers for disease control and prevention (2010) Buruli Ulcer: Technical Information.

37. Agbenorku P, Donwi IK, Kuadzi P, Saunderson P (2012) Buruli ulcer: treatment challenges at three centres in ghana. J Trop Med 2012: 371915.

38. Lehman L, Simonet V, Saunderson P, Agbenorku P (2006) Buruli Ulcer: Prevention of Disability (POD). World Health Organization, Geneva, Switzerland.

39. (2008) Buruli ulcer: progress report, 2004-2008. Wkly Epidemiol Rec 83: 145-154.

40. Etuaful S, Carbonnelle B, Grosset J, Lucas S, Horsfield C, et al. (2005) Efficacy of the combination rifampin-streptomycin in preventing growth of Mycobacterium ulcerans in early lesions of Buruli ulcer in humans. Antimicrob Agents Chemother 49: 3182-3186.

41. Agbenorku P, Agbenorku M, Saunderson P, Lehman L (2006) The benefits of a combination of surgery and chemotherapy in the management of Buruli ulcer patients. Journal of Science and Technology 26: 6-10.

42. Carniol PJ, Meshkov L, Grunebaum LD (2011) Laser treatment of facial scars. Curr Opin Otolaryngol Head Neck Surg 19: 283-288.