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Noma, a neglected disease: a teaching article

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Abstract

Noma (cancrum oris) is a rapidly progressing infection of the oral cavity, associated with a reported 90% mortality rate within weeks after the onset of first symptoms, if left untreated. Noma mostly affects children aged between 2 and 5 years. Along with its high mortality rate, noma causes severe physical and mental health sequelae and affects those most vulnerable living in isolated communities with minimal access to healthcare. While the clinical manifestation and sequelae of noma in each case are unique, the infection invariably starts as an inflammation of the gums, which then leads to ulceration and the rapid destruction (within weeks) of the cheek, jaw, lip, nose and/or the eye. Noma is an ancient but neglected and poorly understood disease afflicting the most disenfranchised populations in the world. Noma has been reported in the literature for many hundreds of years; however, important gaps in our understanding of the disease remain. Future research should focus on determining the burden and distribution of disease, its true mortality rate, pathogenic cause(s) and the factors that influence prognosis and outcomes after treatment. The eradication of noma is possible with a global integrated approach. The addition of noma to the World Health Organization's list of Neglected Tropical Diseases would increase awareness about the disease and provide the impetus for increased research and funding for prevention and treatment programmes.



Fig 1. A 6-year-old child with WHO stage 4 defect of the right cheek and lip, note the exposure of the teeth and early scarring.

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Introduction

Noma is a rapidly progressing infection of the oral cavity, associated with a reported 90% mortality rate within weeks after the onset of first symptoms, if left untreated.¹ Along with its high mortality rate, noma causes severe physical and mental health sequelae and affects those most vulnerable living in isolated communities with minimal access to healthcare. Noma has been reported in the scientific literature for centuries, but many substantial gaps in knowledge about the disease exist. Despite its dire ramifications, noma is not recognized as an official World Health Organization (WHO) Neglected Tropical Disease (NTD). This short teaching article aims to offer an overview of what is currently known about noma and offer suggestions for the integrated control of noma.

Clinical progression and aetiology

Noma mostly affects children aged between 2 and 5 years.² While the clinical manifestation and sequelae of noma in each case are unique, the infection invariably starts as an inflammation of the gums (gingivitis), which then leads to ulceration and the rapid destruction (within weeks¹) of the cheek, jaw, lip, nose and/or the eye.¹ (Figs 1-3)

The WHO has classified noma into five stages:

- Warning sign: simple gingivitis;
- Stage 1: acute necrotizing gingivitis;
- Stage 2: oedema;
- Stage 3: gangrene;
- Stage 4: scarring;
- Stage 5: sequelae.¹

Deaths in noma patients are primarily due to starvation, aspiration pneumonia, respiratory insufficiency or sepsis.³ Noma is not reported to be contagious.¹ The pathogenesis of noma is poorly understood and the microbiology is debated. A range of organisms have been identified in the oral flora of noma patients but none have been consistently present, casting doubt on the role of a specific organism in the development of noma.¹

Risk factors

Reported risk factors for the development of noma include;

- age between 2 and 5 years;
- not being breastfed;
- comorbidities such as measles or human immunodeficiency virus, either at the time of noma diagnosis or in the 3 months leading up to diagnosis;
- a lack of access to quality healthcare including childhood vaccinations;
- poor oral hygiene practices;
- low socioeconomic status of the family.¹

Treatment

The current WHO guidelines for the management of the acute stages of noma in clinical settings include:¹ oral hygiene (chlorhexidine mouthwash 0.2%, 10 ml), antibiotic treatment (amoxicillin and metronidazole), nutritional support (high protein), wound cleaning (compresses soaked in diluted

hydrogen peroxide) and dressing (honey for local dressing and for antibacterial action and regeneration).¹ These treatments can reduce the duration and severity of the infection and the extent of tissue damage, thus reducing mortality and morbidity. Those who survive the early stages will often have severe facial disfigurements and multiple physical impairments such as difficulty eating, seeing and breathing.¹ Survivors often need complex surgical reconstruction to restore form and function.¹ Trismus (lockjaw) is one of the most disabling sequelae and can lead to complications such as aspiration, malnutrition, poor oral hygiene, speech deficits, a compromised airway and chronic pain.⁴ Physiotherapy is an essential part of noma treatment, especially to prevent or minimize trismus, and it can lead to improvements in eating, chewing and speaking.¹ Noma often leads to stigmatization and the resultant social isolation of the patients and their family members.² As such, the management of patients with noma invariably requires social and psychological support. Outcomes of noma treatment are difficult to ascertain due to inconsistent patient follow-up secondary to the remote locations of the home villages of patients and difficulties in accessing healthcare facilities for scheduled follow-up visits.



Fig 2. A 3-year-old child with WHO stage 5 noma who has been left with a significant defect affecting the left eye and cheek.

Burden and distribution of disease

Due to the lack of robust estimates, the true global burden of noma and its distribution is unknown. Noma was commonly reported in Europe and North America until the beginning of the 20th century when living conditions in these regions improved.⁵ In recent years, noma has primarily been reported in low-and middle-income countries in Africa and Asia. In 1998, the WHO estimated that, globally, 140,000 new cases of noma occur each year, and 770,000 patients were living with noma sequelae at that time (based on expert opinion).¹ A 2018 Nigerian study estimated the community-based point prevalence in the northwest was 3300 out of every 100,000 children aged between 0 and 15 years, indicating the global burden could be substantially larger.⁶ The large variation is due to the different study designs and differing stages of noma included in the estimates.

Neglected noma

Noma has been reported for centuries in the scientific literature from a range of settings. However, case reports comprise much of the literature. There are very few robust studies that include primary data collection. The high mortality rate, rapid progression of the disease, difficulties in accessing healthcare and the lack of knowledge about noma amongst healthcare workers^{7,8} leading to misdiagnosis, make noma a difficult disease to study. These challenges mean that patients with noma and the disease itself remain invisible within their local communities, the health systems that are supposed to serve them and the global health community. This invisibility has led to neglect resulting in several substantial gaps in knowledge. The most important areas for future research include enumerating the global distribution and burden of disease, exploring the aetiology and microbiology of noma, the risk factors for progression to the later stages of disease, and the most effective prevention methods and messaging.

Prevention and control

The ideal form of control for noma is prevention. By improving living conditions and removing the main risk factors associated with noma development (low vaccination rates, malnutrition and poor oral hygiene) there would be a reduction in the incidence of noma and other childhood diseases. This could be achieved through job creation, the creation of a functioning vaccination system, improved access to appropriate nutrition, increasing access to quality healthcare, safe drinking water, basic sanitation and hygiene services.

Failing this, noma can be controlled with a move away from the vertical noma surgical programmes to integrated measures. The first would be to shift the focus of control from passive management to active surveillance, early diagnosis and treatment, a method that has worked well for the control of Buruli ulcer.⁹ An additional approach would be for oral screenings to be incorporated into routine primary healthcare assessments, vaccine coverage surveys, surveillance initiatives for other NTDs, or malnutrition surveys.

The above methods require a concerted campaign of awareness so that noma does not continue to be invisible. Increased awareness among healthcare workers would help ensure patients are diagnosed correctly and treated in a timely manner and increase the effectiveness of surveillance systems due to improvements in disease reporting. An initial step to achieve this goal is the inclusion of noma in infectious disease, medical and nursing training programmes. Awareness raising is also necessary at a community level to teach methods to modify risk factors (promote vaccine uptake, nutritional education and healthy oral hygiene practices), and educate community members on the early signs and symptoms of noma and the importance of seeking healthcare in the early reversible stages of the disease.



Fig 3. An approximately 3-year-old child with WHO stage 3 noma, note the defect of the lower lip and gum with near-complete necrosis at the centre.

These awareness campaigns are, by necessity, large, and require resources and attention for noma on a global scale. To garner the necessary support, noma needs to be included on the WHO list of NTDs. This recognition will place noma in the spotlight, highlight the need for more research into the disease, and be a catalyst to raise funds for prevention and treatment programmes.

Conclusion

Noma is a rapidly progressing infection that affects vulnerable young children, many of whom die. Noma is preventable and can be effectively treated if diagnosed early. There are many gaps in knowledge about the disease; it is relatively unknown by healthcare workers and frequently under-reported. The eradication of noma is possible with a global integrated approach. The addition of noma to the WHO list of NTDs would increase awareness about the disease and provide the impetus for increased research and funding for prevention and treatment programmes.

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QUIZ

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A 50-year-old farmer from Anuradhapura, Sri Lanka, presented with troublesome moist ulcerated verrucous skin nodules recurring on the face, limbs and trunk for the past 8 years (Figs 1 and 2).

At onset, he often had a 'stuffy nose' that he attributed to bathing in the farm pond every day after a hard day's work. The nodules around the nose and cheeks developed later and subsequently spread to distant sites including the arms and legs.

Question 1 - What is the diagnosis?

- a) Chromoblastomycosis
- b) Maduramycosis
- c) Atypical cutaneous mycobacterial infection
- d) Disseminated syphilitic gummata
- e) Disseminated rhinosporidiosis

Question 2 - This disease is most strongly associated with...

a) Poultry farming

- b) HIV
- c) Exposure to water in ponds and lakes
- d) COVID -19 infection
- e) Smoking

Question 3 – A skin nodule was biopsied. What is the histology likely to show?

a) Neutrophilic panniculitis

- b) Granuloma with brown annular structures ("copper pennies")
- c) Abscess with sulphur granules
- d) Granuloma with sporangia
- e) Fungal hyphae

Answers on page 31





Figs 1 & 2. Moist ulcerated verrucous skin nodules.

CSH evaluation brief

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In March 2019, the CSH launched an evaluation to help determine how the journal is being used and ensure it is a relevant resource for healthcare workers around the world. The survey was promoted on the front page of four editions of the journal over 2 years. Paper questionnaires were also included as an insert in hard copies of the publication. Readers were able to submit responses via Survey Monkey, email or post. At the CSH, we greatly appreciate the valuable feedback our readership gave and are now happy to share some highlights!

Key findings from the 54 responses*

- 55% of respondents were healthcare workers specifically working in dermatology and venerology and 34% were general healthcare workers.
- 96% of respondents shared that they found the journal a useful resource.
- 96% of respondents said that the journal is beneficial to healthcare workers working in the field.
- 96% agreed that the journal provides information on how to better treat (or provide more support to) patients and the wider community.
- 62% indicated that once they have read the journal, they share it with colleagues.

The CSH has listened to feedback from readers on possible improvements to the journal and in future volumes we hope to:

- include more case reports and images;
- increase our disease coverage;
- provide more information regarding alternative drugs available in low-resource settings;
- research the potential of translating into new languages;
- include more articles from nursing perspectives;
- include more images of skin manifestations on different skin tones;
- provide further opportunities for readers to share their field experiences.

We would like to thank you for your valued contributions. Please reach out to us by email at csh@ilds.org if you would like to share further feedback on the CSH journal.

*The CSH recognizes that the low response rate for the evaluation survey limits our findings. We acknowledge that as a single hard copy of the journal is often shared among several people, this may have been a factor affecting responses.

Comments from our readers:

For general health workers, as skin diseases or manifestations of disease are so extremely common, having relevant, up to date information on diagnosis, management and follow-up is very helpful.

General Healthcare worker

There are some dermatological conditions which have been managed through the use of these journals. Cases presented in the journal have gone a long way in diagnosis of dermatological conditions encountered here.

Dermatology and Venereology Healthcare worker in Zimbabwe

It facilitates new knowledge, skills and practice in the dermatology field.

Dermatology and Venereology Healthcare worker

Articles are up to date and help me provide better dermatological healthcare in resource limited settings, and the articles I share provide knowledge that would otherwise not be available.

Dermatology and Venereology Healthcare worker in Haiti and Kenya



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Our grateful thanks to the following, who have generously given their time and expertise to supervise translation of Community Skin Health into the following languages:

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Postgraduate dermatology programmes in Africa – a synopsis, with permission, of the original article published in *Dermatologic Clinics*¹

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Key words: Africa, curricula, dermatology education/training, dermatovenereology, education/training.

Key learning points:

- Africa, comprising 55 independent nations, has a population of 1.2 billion and a population density of 30.5 people/km².
- The reported prevalence of skin disease in rural Africa ranges from 26.9% in Tanzania to 80.4% in Ethiopia, with new consultations for skin disease ranging from 6.9% for all levels of public healthcare in Mali to 13% for primary care in 83 villages in Cameroon, according to a World Health Organization (WHO) report.²
- The number of dermatologists per million population in each country varies (Fig 1). Most countries have a ratio of <10 dermatologists per million population, usually <1 per million population, except for North Africa.
- Of the 55 African countries, 30 have no training programmes, 10 have a regional programme, 10 have a national programme, and 5 have university programmes (Fig 2).
- Dermatology training programmes on the continent are of international standard and produce dermatologists with sound clinical acumen and understanding of the needs of the population they serve.
- Training programmes designed in the "global north" and transplanted into African universities emphasize practices, including latest therapies, which are not available, cost- effective, or based on evidence from Africa.



A synopsis of postgraduate dermatology training programmes and educational tools across Africa (Table 1)

- **Southern Africa:** The number of dermatologists per million population ranges from 0 to 3, with the highest number in South Africa (SA) (177). There are 3 training programmes: Angola, Mozambique and South Africa.
- North Africa: The greatest number of dermatologists (5000) on the continent is in Egypt with 49/million population. The number of dermatologists per million population ranges from 3 in Mauritania to 49 in Egypt. There are training programmes in Algeria, Egypt, Libya, Morocco and Tunisia.
- West Africa: The number of dermatologists per million population ranges from 0 to 3. Training programmes are found in Ghana, Mali, Nigeria and Senegal . All follow a harmonized regional programme for dermatology which is acceptable to most West African countries, even those without a training programme.
- East Africa: The range of dermatologists per million population is 1 to 7. There are established dermatology specialist training programmes in Ethiopia, Kenya, Madagascar, Sudan, Tanzania and Uganda. The Regional Dermatology Training Centre in Moshi, Tanzania offers the only Advanced Diploma in Dermatovenereology for clinical officers and medical assistants from sub-Saharan Africa.

These 290 diploma graduates are the backbone of dermatology services in sub-Saharan Africa.

• **Central Africa:** There are 0.1-5.5 dermatologists/million population in this region. There is a training programme in the Democratic Republic of Congo.

Regions that have dermatology or dermatovenereology training programmes following regional, national or university curricula (Fig 2)

- Regional programme, harmonized curriculum and country accreditation: West Africa; Anglophone Ghana and Nigeria; Francophone Benin, Burkina Faso, Côte d'Ivoire, Guinea Conakry, Guinea Bissau, Mali, Senegal and Togo.
- National programme, curriculum and accreditation: Algeria, Angola, Egypt, Ethiopia, Libya, Morocco, South Africa, Sudan, Tanzania (RDTC) and Tunisia.
- University programme, curriculum and accreditation: Democratic Republic of Congo (University of Kinshasa), Kenya (Moi University), Madagascar (Antananarivo University), Mozambique and Uganda (Mbarara University).

There is little sharing of curricula and training programmes between the various universities except for West Africa, highlighting the need for pooling of expertise and resources so that training can be strengthened regionally, and curricula developed that are relevant to local African needs.

Table 1. Summary of training programmes by region and country							
Region	Country	Training Programmes					
	Angola	4-year specialist diploma issued by the Ministry of Health which has a national curriculum and examination by Colegio Angolano de Dermatologia e Venereologia College of Dermatologists of SA					
Southern Africa	Mozambique	5-year diploma (3 in dermatology). Specialist in Dermatology and Venereology					
	South Africa	4-year specialist fellowship, FF Derm, (SA), which has a national curriculum and examination by College of Dermatologists of SA. For the university based MMed masters a thesis is required. To register as a specialist dermatologist both the FF Derm (SA) and MMed are required					
	Egypt	5-year master's programme with MD/PhD					
North Africa	Libya & Tunisia	4-year master's programme with exit examination					
	Algeria & Morocco	Details unavailable at time of publication					
		Training programmes in Nigeria, Ghana, Senegal and Mali A regional harmonized curriculum is widely followed Programmes are adapted to local needs and have curricula, logbooks and research components					
West Africa	Ghana	5-year fellowship (3 in dermatology). Fellowship of the West African College of Physicians or the Fellowship of the Ghana College of Physicians and Surgeons					
	Mali	4-year programme					
	Nigeria	5-year fellowship (3 in dermatology). Fellowship Internal Medicine subspecialty Dermatology					
	Senegal	4-year diploma (Specialist in Dermatology)					
	Ethiopia	National curriculum since 2006. Diploma (Specialty Certificate in Dermatovenereolgy)					
	Madagascar	Resident training (4 years) since 2000. Fellowship with certificate French Diplôme d'Études de Formation Spécialisée (DEFS) and Attestation de Formation Spécialisée (AFS)					
East Africa	Sudan	4-year MD. 500 dermatologists since training started in 1995					
	Tanzania	RDTC and WHO collaborating centre trains various healthcare workers from sub-Saharan countries. Both a 4-year MMed residency programme and 2-year Advanced Diploma in Dermatovenereology (ADDV) are offered.					
	Uganda	3-year programme (MMed) which has graduated 13 dermatologists since the programme started					
Central Africa	Democratic Republic of Congo	5-year diploma (Doctor Specialist in Dermatology) at the University of Kinshasa					

Postgraduate dermatology programmes in Africa – a synopsis, with permission, of the original article published in *Dermatologic Clinics*¹ ...continued

What is evident is that many curricula are developed in high-income countries which dominate the global knowledge space setting the metrics for success to their benefit and to the detriment of low-income to middle-income country knowledge systems (epistemicide). These curricula are "transplanted" into universities on the continent with minimal consideration of their relevance for Africa or its peoples. Calls for the decolonization of global health education and acknowledgement of the legacy of former colonial relationships and their influence on global health initiatives are increasing.³

Most of the continent has <10 dermatologists/million population; only North Africa has a higher number of 8–49 dermatologists/million population. Most African dermatologists are private practitioners in urban areas with only a minority in academic and public service resulting in limited or absent skin healthcare for large sectors of the population.⁴ More detailed information about the numbers of dermatologists, types of training programmes available and details of curricula by region and by country has been published recently.¹

A previous attempt to describe dermatology training in Africa which focused mainly on anglophone countries noted that training was limited.⁵ Some solutions put forward to improve the delivery of skin health include task shifting (with short courses and diplomas for the training of generalists, nurses, medical assistants, clinical officers and traditional healthcare providers) and technology, particularly teledermatology.

The educational materials available for African training programmes are predominantly driven by sources published in Europe, America and the United Kingdom. The applicability of this material to those with pigmented skin in different global regions and the ability of dermatologists to diagnose and manage skin disorders in people of colour has not been evaluated.

In 2006, Ebede and Papier undertook a survey of educational material dealing with pigmented skin in the United States of America.⁶ The dermatology textbooks they evaluated contained low percentages of images and textual content focused on ethnic skin (Bolognia 19%; Freedberg 15%; Rook 12%; Fitzpatrick 5th edn. 11%; Fitzpatrick 4th edn. 10%; Sauer's 9%; and Habif 4%). From 1996 to 2005 the percentage of teaching events at the American Academy of Dermatology annual meetings that focused on skin of colour has remained static at 2%. They recommended that more photographic coverage and textual information describing common and serious skin diseases in people of colour should be incorporated into educational resources. This highlights the inadequate educational material currently preferred and recommended for African residents.⁶



Fig 2. Regions that have dermatology or dermatovenereology training programmes follow regional, national or university curricula.

There is need for the development of programmes that provide education and resources that are more culturally, ethnically, and geographically appropriate.

Continent and region-wide surveys of dermatologists trained, their distribution in countries, their retention profiles and their contribution to both undergraduate and postgraduate dermatovenereology training and curriculum decolonization should be undertaken. Also, of importance would be surveys of how this contribution of dermatovenereology to the health burden is recognized and funded throughout the continent, both with respect to clinical services to the population and the funding of training.

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A distorted presentation of steroid dermatitis resembling rosacea: a case report

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Key words: Steroid dermatitis resembling rosacea, topical corticosteroids, rosacea.

Abstract

Steroid dermatitis resembling rosacea manifests clinically with erythema, papules and pustules. In some patients, atrophic changes such as telangiectasia, developing from the repeated application of moderate-to-high potency topical corticosteroids, are also present. We present a 31-year-old female patient who had steroid dermatitis resembling rosacea. She had severe erythema, papules, nodules, and scales on the face. Our therapeutic approach, which consisted of tapering topical corticosteroids and initiating tinidazole and topical tacrolimus, resulted in notable improvement within 2 months.

Introduction

Rosacea is a common chronic inflammatory facial dermatosis. Rosacea is characterized by flushing, background erythema, telangiectasia, inflammatory papules, pustules, oedema, phymatous changes, and ocular features.¹⁻³ It mostly affects white patients with light skin phototypes and is poorly documented in black patients. This under-reporting may be the result of under-recognition due to obscurity of typical rosacea features (e.g. background erythema and telangiectasia) and the innate hyperpigmentation in people with skin phototypes V and VI.¹⁻³ One of the causes of a rosacea-type appearance is the overuse of topical corticosteroids (TCS).

From 1951 to 1974, many dermatologists tried to come up with a proper name for steroid-induced dermatosis. It is in 1974 that Leyden et al.gave it the name of "steroid rosacea".⁴ On the other hand, considering that rosacea is already a welldefined term, it was thought that the best name for it would be steroid dermatitis resembling rosacea (SDRR). SDRR is a common symptom of TCS overuse. Although discontinuation of TCS is necessary to stop SDRR, a rebound reaction may occur if the discontinuation is too abrupt.⁵ There are three types of SDRR, classified according to localization: perioral, centrofacial and diffuse-type.⁶ They clinically manifest with erythema, papules and pustules. In some patients, atrophic changes, such as telangiectasia, develop from the repeated application of moderate-to-high potency TCS.7 In patients of skin phototypes IV-VI, erythema is often not visible with the naked eye making the clinical diagnosis of SDRR difficult. Dermatoscopy may be a valuable tool to assess erythema in dark skin types.⁸ Rosacea can be mistaken for acne, an allergic reaction or other conditions that present with skincoloured papules.³ The first step in the management of SDRR is to discontinue all the TCS. The second important factor to consider is the suppression of bacterial infection in hair follicles with systemic antibiotics. The selection of antibiotics should consider the presence of the Gram-negative bacteria, *Staphylococcus* spp., *Streptococcus* spp., and overgrowth of *Propionibacterium acnes*, resulting from the overuse of TCS.⁶

Case

A 31-year-old female patient complained of itchy erythematous papules and patches on the face. She reported she had experienced the same lesions for almost 4 years and she had been treated with several topical steroids and antifungal creams. During her last consultation, she was treated for tinea incognito with miconazole cream 2%, which made her lesions worse with severe signs of irritation.



Fig 1. Erythema, papules, nodules and scales.

A distorted presentation of steroid dermatitis resembling rosacea: a case report...continued

On examination she had severe erythema of all affected facial areas, upper chest and back. The lesions consisted of papules, nodules and scales. She also had some comedones and telangiectasia on the forehead and malar region. A few terminal hairs were noticeable on the chest as well as on the mandibular area and chin (Fig 1).

Laboratory studies revealed a normal hormonal profile. On microscopy, no demodex or fungal elements were seen.

The case was diagnosed clinically as steroid dermatitis resembling rosacea. Oral tinidazole 500mg twice a day and hydrocortisone 1% cream twice daily were started. From the second week, the hydrocortisone 1% cream was tapered to once daily, then to three times a week for the third week, then twice a week for the final week before it was stopped. Tacrolimus 0.01% cream was started on the days hydrocortisone was not applied. After 1 month of treatment, she still had erythema, but it was mild, and papules were remarkably decreased (Fig 2). On the third visit, there was complete resolution of papules and scales (Fig 3). A maintenance dose of metronidazole gel 1% was given to apply twice daily. For the first and last visits, dermoscopy images were also captured and revealed a remarkable reduction of erythema (Fig 4 and Fig 5).



Fig 2. Mild erythema and few papules.

Discussion

Only a few days of TCS treatment is sufficient to suppress the signs of many primary dermatoses. TCS have antiinflammatory characteristics and vasoconstrictive effects, which especially eliminate erythema, though they do



Fig 3. Complete resolution of papules and scales.

not treat the cause of the disease. Discontinuation of TCS often causes the exacerbation of the primary dermatosis, leading many persons to continue to use TCS long term.⁶

It is challenging to appreciate erythema with the naked eye on skin phototypes IV up to VI. This complicates the diagnosis of rosacea in these skin types as erythema may be the main diagnostic feature. In darkskinned individuals, when erythema is seen with a history of TCS usage, it is often mistaken for tinea incognito, acne or allergic reactions.³

Our patient was initially diagnosed to have an allergic



Fig 4. Erythema seen on dermoscopy on the first visit.



Fig 5. Reduced erythema seen on dermoscopy on the second visit.

reaction then later tinea incognito. Her long-term management with TCS followed by topical antifungal caused much irritation along with SDRR. After we examined her dermoscopically, she was clinically diagnosed to have SDRR. After tapering down the TCS and initiating tinidazole 500mg twice daily together with topical tacrolimus 0.01%, our patient showed marked improvement. We used metronidazole twice daily for her maintenance therapy.

Conclusion

Long-term usage of TCS can result in SDRR. We recommend that the diagnosis of SDRR based on dermoscopy findings should be considered for a patient who used TCS when presenting with erythema and other dermatoses.

Consent

The patient consented to participate in this study and have her data published.

Acknowledgement

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DERMATOLOGY ATLASES (free access)

There are several online picture galleries of skin conditions, many including illustrations of lesions in skin of colour.

www.atlasdermatologico.com.br

One of the most extensive created by Dr Samuel Freire de Silva.

https://dermnetnz.org

The New Zealand Dermatology Society offers authoritative advice on management, with a link to conditions in skin of colour.

https://www.dermatlas.net

Drs RP Usatine and BD Madden have produced an interactive dermatology atlas.

www.dermoscopyatlas.com

Dermoscopy images are available here.

https://www.cmecde.com/download-atlasof-clinical-dermatology-3rd-edition-pdf-free/

To download a pdf of Anthony du Vivier's Atlas of Dermatology.

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Links to other online resources are available.

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Post-elimination scenario of leprosy in a tertiary care hospital in South India

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Key words: Leprosy, elimination, multibacillary.

Key learning points

- 1. Leprosy continues to be a major health problem in low-income countries despite its "elimination" status.
- 2. The total numbers of multibacillary cases and new cases among children are high in our study, which points to active transmission in the community.
- 3. The presence of deformities severely affects the quality of life in individuals with leprosy and will potentially affect their livelihood.
- 4. Lower smear positivity status reflects the need for improved training of health workers in the technique, and in the use of other diagnostic modalities like histopathology or polymerase chain reaction (PCR), so as to increase the diagnostic yield.
- 5. Public health education, increased contact tracing and training of healthcare workers are essential in order to achieve leprosy eradication.

Abstract

Background: Leprosy or Hansen's disease is a disabling condition, with widespread prevalence. It continues to be a major public health problem despite efforts by the World Health Organization (WHO) and governmental agencies to curb its spread.

Objectives: This study aimed to describe the clinical and epidemiological profile of patients with leprosy attending a tertiary care institute in southern India, spanning a period from 2005 to 2020.

Methods: Medical records of patients with leprosy were analysed retrospectively, over a period ranging from 2005 to 2020. This study was undertaken at the Department of Dermatology, Venereology and Leprosy, Father Muller Medical College, Mangalore, India. Data were obtained using a specific questionnaire and entered into the database system.

Results: A total of 520 newly diagnosed cases were seen during the study period. Borderline tuberculoid leprosy was the commonest clinical type. Smear positivity was low, despite multibacillary cases being higher in number. The occurrence of visible deformities and lepra reactions which contribute to the morbidity, continue to be a cause for concern.

Conclusion: This study revealed that even in the postelimination era, the presence of active transmission in the community continues to occur. Hence, more stringent and focused health education and diagnostic methods are the need of the hour.

Introduction

Leprosy, a chronic infectious disease which affects the skin, eyes and peripheral nerves, is caused by *Mycobacterium leprae* and is chiefly transmitted by close contact with individuals with the disease or asymptomatic nasal carriers by droplet spread. The organism can also persist in the environment. Although it is one of the oldest diseases known to humankind and the first infectious disease to have its aetiological agent discovered, it still remains a public health problem especially in low-income countries.¹ Following the introduction of multidrug therapy (MDT) in 1982, the reported global prevalence of leprosy had reduced from over 5 million cases to less than 200,000 by the end of 2016. The situation in India too saw a drastic change from a prevalence rate of 57.8/10,000 in 1983 to less than 1/10,000 in 2005, when elimination was declared in India as per World Health Organization (WHO) criteria.² However, among the African, South American and South East Asian countries that report high numbers of cases, India still remains one of the leaders in terms of numbers. A total of 120, 334 new cases and 9227 childhood cases were detected by the end of the year 2018 in India, indicative of active transmission.³

The epidemiological distribution of leprosy cases traditionally has been clustered in tropical climates which have high temperatures and abundant rainfall. However, temperate and cold climates too have shown high numbers in some instances; leprosy is a disease of poverty.⁴ We undertook this study to describe the epidemiological profile of cases in a tertiary care hospital in South India which predominantly has the former climate. Heavy industrialization and migration of populations from other endemic areas have added to the burden of leprosy in our region. We studied the prevalence of leprosy cases over a 15-year period, ranging from 2005 to 2020, in order to assess the prevailing control programmes and devise methods to improve the situation in future.

Materials and methods

This was a retrospective, observational, descriptive study in which consolidated data on leprosy cases were analysed. Data were obtained from the medical records of the Department of Dermatology, Venereology and Leprosy, Father Muller Medical College, Mangalore, India. Patient records for a period of 15 years, from April 2005 up until March 2020 were analysed. All these patients included were thus on the prescribed WHO MDT. Data recorded in the patients' profiles at the time of diagnosis were retrieved using a specific questionnaire, and then entered into a database system. The following variables were collected:

- total number of new cases;
- new cases among children;
- number of patients with multibacillary (MB) and paucibacillary (PB) disease;
- presence of grade 2 or visible deformity;
- presence of reactions;
- smear positivity from routine sites. The data were processed using Microsoft Office Excel 2007 in the construction of tables and graphs.

Results

A total of 520 new cases were seen during the study period (Fig 1). This included 47 (9.0%) newly affected children aged 4 to 15. Among the cases, 331 (63.7%) were classified as MB and 189 (36.3%) were deemed PB after smear and clinical examination (Fig 2). Borderline tuberculoid leprosy was the most common type at the time of diagnosis (261, 50.2%), followed by tuberculoid leprosy (98, 18.9%). Mid-borderline leprosy was the least commonly encountered subtype (16, 3.1%) (Table 1). A total of 102 (19.6%) cases presented with reactions during treatment and follow up. Furthermore 45 (8.7%) of patients had grade 2 or visible deformities; some, such as clawing of hands or neuritis were related to reactions during treatment, although the precise proportion was







Post-elimination scenario of leprosy in a tertiary care hospital in South India...continued

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Clinical	types	of	new	leprosy	cases	

Clinical type of leprosy	Number (%)
Indeterminate	19 (3.7%)
Tuberculoid	98 (18.9%)
Borderline Tuberculoid	261 (50.2%)
Mid-borderline	16 (3.1%)
Borderline lepromatous	49 (9.4%)
Lepromatous	53 (10.2%)
Pure neural	24 (4.6%)

unavailable. Of the total number of cases, smear positivity was seen only in 115 (22.1%).

Discussion

In April 2016, the WHO launched a 5 year "Global Leprosy Strategy 2016-2020" which was titled "Accelerating towards a leprosy free world". It revolved around early leprosy detection and disability reduction. It was based on the agenda of elimination of leprosy at the subnational level which has been far from satisfactory, and hence the challenge remains. A specific focus was on women and children, increasing contact tracing, strengthening referral systems, assessing the efficacy of chemoprophylaxis and monitoring for drug resistance. At the end of the year, a slight decrease in prevalence was noted worldwide but many endemic developing and underdeveloped countries failed to send their data, so the accuracy of the data is limited.² Thus, it is important that in diseases like leprosy with a global burden of cases that accurate data are collected.

Among the patients we studied, MB cases (63.7%) were more common in comparison with PB cases. A high number of MB leprosy cases and associated disabilities may reflect national deficits in the control programme as regards the epidemiological surveillance methods and health education tools being used, which result in a late diagnosis.⁵ Furthermore, the high proportion of MB cases indicates ongoing active transmission of leprosy in the community.²

Although the percentage of MB cases was 63.7%, smear positivity was seen in only 22% of the total cases. The yield from smears was disappointingly low and there is a need to improve the training of health workers in taking, using specific stains and interpreting smears. It is important that smears are not stained for TB but adapted for leprosy. Use of TB ZN stain is often the cause of low readings. PCR may be needed (multiple sites from one patient can be pooled to reduce cost). Although histopathology is valuable, it samples only one or a few sites and it requires a higher level of training.

The prevalence in children under the age of 15, was 9.0% in our study, in concordance with previous reports and the national average.^{6,7} Detection of cases in this group is taken to be an indicator of the greater severity of endemicity of the

disease. Children may be the contacts of cases undetected by the healthcare system. Childhood leprosy is a serious problem, as the younger the age of affliction with the disease, the higher the possibility of developing disability, which can be counterproductive in a developing economy as affected patients would be of limited economic productivity in the future.⁸

The development of physical disabilities in leprosy is always a concerning issue. We found 8.7% patients with disabilities, which points to deficiencies in early case detection. It also highlights the need for specialized centres for deformity management, which if tackled early in the course of the disease, could drastically reduce the associated morbidity. The fact that the study was performed in a tertiary care referral hospital could also indicate higher referral of patients with early deformities.⁹ Notwithstanding, it is of paramount importance that early detection of reactions be made a priority, in order to reduce disabilities.

As this study was retrospective, the availability of data was limited. Histopathological data have not been included although histopathology was used in the confirmation of diagnosis. History of contacts of each case would have added more value to the study and helped in contact tracing.

Conclusion

Although the WHO's strategy is of accelerating towards a leprosy-free world, our study shows ongoing active circulation of lepra bacilli in the community. The high prevalence of MB cases and visible deformities indicates that the main hurdles to this acceleration are from late diagnosis, probably due to delayed reporting or lack of awareness of the signs and symptoms of the disease. Hence, in accordance with the global strategy of 2016–2020, we need to increase contact tracing and strengthen referral systems and, most importantly, the focus should be on health education of the general public and improved training of health workers, including the detection and treatment of reactions.

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New ILDS Executive Director announcement

We are pleased to announce that Arpita Bhose will be the next International League of Dermatological Societies (ILDS) Executive Director.

After serving as Executive Director for 5 years, Jo Groves passes on a strong foundation to Arpita for the implementation of the ILDS 2020–2023 Strategy and beyond. Please join us in thanking Jo for her dedicated leadership and strategic direction in strengthening the development of the ILDS. We look forward to an exciting 2022 with Arpita leading the Secretariat, as we prepare for the 3rd ILDS World Skin Summit from 13 to 15 October 2022 in Peru and the 25th World Congress of Dermatology from 3 to 8 July 2023 in Singapore.





GLODERM Trainee Committee Webinars

The GLODERM Trainee Committee have launched a new educational webinar series for 2021-2022! These free monthly online webinars are delivered by expert speakers and trainees from around the world on a range of different dermatology topics. Previous webinars discuss skin conditions including albinism, psoriasis, pigmentary disorders and deep fungal infections.

This series hopes to deliver educational events focusing on clinical dermatology, research, leadership and management skills. These webinars are aimed at trainees, practising dermatologists and healthcare practitioners providing dermatologic care in low-resource settings. The webinars also serve as a virtual platform in which trainees or individuals interested in learning about dermatology can meet other dermatologists from around the world and engage in dialogue about skin health. To access free recordings of our latest webinars when they are released, visit and subscribe to the GLODERM YouTube channel here: https://www.youtube.com/c/GLODERM

If you would like to be the first to hear about upcoming webinars and Trainee activities, please sign up to the GLODERM Trainee mailing list here: http://eepurl.com/hHvI_L

We look forward to seeing you at future GLODERM webinars!

QUIZ ANSWERS

1. e) Disseminated rhinosporidiosis

- 2. c) Exposure to water in ponds and lakes
- 3. d) Granuloma with sporangia

Discussion

Rhinosporidiosis is a chronic granulomatous parasitic infection caused by *Rhinosporidium seeberi*, an aquatic parasite that has been recently placed in the class Mesomycetozoea after much debate. Predominantly an infection of the mucous membranes of nose and nasopharynx, rhinosporidiosis



Figs 1 & 2. Moist ulcerated verrucous skin nodules.

is endemic in parts of South Asia, South America and Africa. Infection occurs through trauma-induced direct inoculation and there is often a history of contact with stagnant water. In Sri Lanka and neighbouring countries, farmers work barefoot in muddy paddy fields.

Clinical presentation to the dermatology clinic is with soft cutaneous nodules that resemble polyps at onset and later acquire a verrucous form. Dissemination, with cutaneous nodules at multiple sites, and internal organ involvement, as in this patient, is recognized but rare. There is no association with immunodeficiency.

The diagnosis is confirmed by the typical appearance of *R. seeberi* sporangia seen in scattered granulomata (Fig 3).

Solitary or well-defined lesions respond very well to surgical excision but may recur. Dapsone appears to be effective in early disease (e.g. lesions limited to the nose or solitary skin nodules). In disseminated disease, systemic therapies are mostly ineffective; dapsone, given in prolonged courses, is the preferred treatment; it may reduce the risk of recurrence after surgery. However, recurrences are very common.



Fig 3. Thick-walled *Rhinosporidium* sporangia (arrowed) containing endospores.

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The International Foundation for Dermatology (IFD) was created in 1987 to carry out the global health dermatology activities of the ILDS. Today, the IFD supports projects in Africa, AsiaPacific and South America. CSH is the official journal of the IFD.



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