Crusted (Norwegian) scabies treated with oral ivermectin: A case report and overview

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Abstract
Crusted (Norwegian) scabies is a severe and highly contagious form of infestation caused by the mite Sarcoptes scabiei var. hominis. It occurs in a subgroup of patients who are either immunocompromised or physically or mentally debilitated. Due to its atypical clinical presentation as well as difficulties associated with its management it presents a number of challenges both from a therapeutic aspect as well as from public health perspective. Failure to diagnose this condition may give rise to a massive epidemic when the patient is in an institutional setting.

We describe a case of crusted scabies which occurred in a nursing home leading to an institutional outbreak. She received a combination of oral ivermectin and topical scabicidal with good results.

This case highlights the importance of being alert to the possible diagnosis of crusted scabies and gives an overview of the condition as well as management strategies.

Keywords
Crusted scabies, Sarcoptes scabiei, ivermectin, institutional outbreaks

Introduction
Scabies is an inflammatory disease of the skin caused by the burrowing mite Sarcoptes scabiei var. hominis. It is a common worldwide problem affecting as many as 300 million people worldwide annually. Data collected by the Royal Infirmary of Edinburgh from 1815 to 2000 indicate a prevalence of about 5%, with rising incidence and prevalence during war-time. In a recent study carried out in the UK, the mean prevalence was 2.27 in 1000 for boys and men and 2.81 in 1000 for girls and women. It occurs in all age groups, races and socio-economic groups, although institutionalization, poverty, poor hygiene, overcrowding, undernourishment and sexual promiscuity have been proposed as significant risk factors. It has a history as long as mankind as has been described by Aristotle (384 BC – 322 BC) and in the Old Testament.

Crusted scabies was originally called Norwegian scabies because the condition was first described in Norway in 1848 among patients with leprosy. In fact, it may occur in any subject who is immunocompromised, malnourished, debilitated or has sensory impairment.

Case Report
A 92-year-old lady, resident in a home for the elderly, was admitted to Mater Dei Hospital because of an elevated INR greater than 10. On examination by the admitting firm she was noted to have a widespread, excoriated scaly eruption.

The patient had been bed-bound for several years and suffered from dementia and longstanding paresis, due to cerebrovascular disease. She was on long term bumetanide, warfarin, perindopril and iron supplements (Ferroglobin). Direct history taking was not possible because of her mental state. According to her carers the patient had had increasing skin scaling for several months, initially on the hands and scalp, gradually becoming more widespread. For some time she had been treated with topical steroid creams for presumed psoriasis with no improvement; more recently generalised erythema had appeared.

On examination there were large areas of warty crusting on the hands, feet and scalp with thickening and fissuring of the palms and soles (Figs 1, 2). The fingernails were thickened and discoloured with scaly debris evident in the distal subungual spaces. In addition, there was generalised erythema with widespread excoriated papules,
particularly over the breasts, axillae and abdomen. No typical scabetic burrows were identified. The patient was visibly uncomfortable and constantly attempting to scratch her skin, although she was unable to do so effectively because of her neurological state and flexural contractures.

A clinical diagnosis of crusted (Norwegian) scabies was made and one application of 0.5% malathion lotion (Derbac-M®) applied. In view of the extent and severity of skin involvement a decision was made to transfer the patient to the Dermatology ward at Sir Paul Boffa Hospital for further management. She was treated systemically with oral ivermectin (Stromectol) 6 mg (equivalent to 200µg/kg). She was simultaneously treated topically with 2 applications of 5% permethrin cream (Lyclear® dermal cream) applied, 3 days apart, to all the skin surface, including the scalp, and left on for 12 hours. She was also given a one-week course of oral flucloxacillin in view of the generalised erythema thought likely to be secondary to staphylococcal skin sepsis. The patient was nursed in a single room and members of staff who came in contact with her were instructed to wear gloves and long-sleeved gowns to avoid infection. The fingernails were trimmed and subungual debris removed. The generalised erythema subsided within a few days. However, since after 1 week significant crusting was still present, especially on the palms and scalp, the patient was retreated with oral ivermectin 6 mg (2 doses 2 days apart) and a further application of permethrin cream. The skin eventually cleared completely and she was discharged back to her nursing home 2 weeks after admission. She remained clear when reviewed at the home 6 weeks later.

The case was notified to the Public Health Department. It transpired that other cases of scabies in residents of the same home had recently been reported. The home was visited by public health physicians and a dermatologist (MJB) and all residents, staff and contacts were treated. Ivermectin was prescribed to the most severely affected cases.

Discussion
Crusted scabies is a clinically distinct and highly contagious form of scabies. It presents several clinical challenges including diagnostic and therapeutic ones as well as posing a particular public health problem. Crusted scabies occurs when the host’s response to the mite is altered or inadequate (Table 1). It may occur in immunosuppression due to disease such as HIV or lymphoproliferative disorders as well as iatrogenic causes including post-transplant patients and those on drugs which modify the immune response such as long-term corticosteroids. Given its increased use in dermatology, rheumatology and gastroenterology it is pertinent to note that there has been a report of a case of scabies occurring during treatment with infliximab. Inappropriate use of potent topical steroids may predispose to this condition as well. In addition it may develop in the context of severe systemic illness or when there is neurological disease which either causes immobility and a reduced ability to scratch and thus dislodge the mites or gives rise to reduced sensation.

Table 1: Factors predisposing to crusted scabies

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<th>Example</th>
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<tr>
<td>Immunosuppression</td>
<td>Post-transplant patients</td>
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<tr>
<td></td>
<td>HIV/AIDS, HTLV-1</td>
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<td>Leukaemia, lymphoma</td>
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<td>Drugs e.g. corticosteroids, immune modulators</td>
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<td>Severe systemic disease</td>
<td>Rheumatoid arthritis or other severe arthropathy</td>
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<td>Systemic lupus erythematosus</td>
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<td>Epidermolysis bullosa</td>
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<td>Neurological disease</td>
<td>Paresis</td>
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<td>Down’s syndrome</td>
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<td>Sensory neuropathy</td>
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In these patients there is a reduced reaction to the mite due to failure of the skin immune system to mount an effective response. It has been reported that in such cases there is a predominance of infiltrating CD8+ T lymphocytes in the dermis with minimal helper T lymphocytes (CD4+) and an absence of any B cells. There may also be a reduced physical response to the mite (i.e. diminished scratching) or both these factors working in tandem. This results in absent or diminished symptoms (only about 50% of patients with crusted scabies have an itch) as well as giving rise to atypical and exaggerated clinical signs.

Due to the altered host response to the mite, the clinical presentation differs considerably and this may give rise to difficulty in diagnosis with resultant delay in proper treatment. In cases where the patient lives in an institution this poses a public health risk as this form of scabies is highly contagious due to the fact that there may be hundreds of thousands of mites present on the host, whereas there are usually only 10-20 females in classical scabies. Such patients shed large amounts of scale which contain vast quantities of mites.

Crusted scabies presents as a hyperkeratotic eruption particularly marked on the palms and soles which may be
fissured and/or warty. This may be accompanied by erythema and scaling on the face, neck, scalp and trunk which may become generalized. There is frequently an accumulation of scaly debris under the nails. It occasionally presents as erythroderma as in this case. Staphylococcus aureus colonizing burrows might play a part in initiating the erythroderma. Secondary infection may also occur with other organisms, commonly Streptococcus pyogenes and this leads to increased mortality in this subgroup of patients. Patients may also have generalised lymphadenopathy and frequently exhibit eosinophilia and elevated total IgG and extremely elevated IgE levels (up to ten times normal levels). As it mimics other common dermatoses such as psoriasis, eczema and contact dermatitis the diagnosis of scabies should always be kept in mind particularly when examining patients in the at-risk categories who, in addition, may be unable to give a proper history. It is frequently misdiagnosed as “senile pruritus” by primary caregivers and is treated with topical corticosteroids which aggravates the condition and leads to further delay in treatment.

**Treatment**

Crusted scabies presents a treatment challenge for several reasons. The huge burden of infestation usually requires several treatments and relapse is common. Although treatment with a combination of topical and oral treatment is generally recommended the presence of a compromised skin barrier function may lead to a greater risk of toxicity through increased skin absorption. In addition local side-effects such as skin irritation and a burning sensation may be exacerbated in this situation. The Centers for Disease Control and Prevention (CDC) currently recommends combined treatment with a topical scabicide and repeated treatment with oral ivermectin 200μg/kg on days 1, 2, 8, 9, and 15. Additional treatment on days 22 and 29 might be required for severe cases. Ivermectin should be combined with the application of either 5% topical benzyl benzoate or 5% topical permethrin (full body application to be repeated daily for 7 days then 2 times weekly until release from care or cure). This is in contrast to classical scabies which usually only requires 2 applications, one week apart, of a topical scabicide.

Ivermectin binds selectively to receptors in the peripheral motor synapses of the mite, blocking chemical transmission of g-aminobutyric acid (GABA)-gated chloride channels localized in the CNS. This stimulates the discharge of GABA at the nerve endings of endoparasites, increasing the affinity of GABA in the receptor at synapses and causing interruption of the nerve impulses, producing paralysis and death of parasites. Ivermectin is relatively safe. Reported adverse reactions include fever, headache, chills, arthralgia, rash, eosinophilia, and anorexia. However, many of these symptoms are thought to result from the death of parasites rather than as a reaction to the drug.

There are several advantages to using oral ivermectin over standard topical scabicides. These include high efficacy, ease and rapidity of application and avoiding the irritation caused by topical treatments. It also mitigates to some extent for improper/inadequate application of topical agents (this may apply in particular to institutional outbreaks where staff may have to deal with many cases concurrently). It is thus convenient for persons who are bedridden and immunocompromised.

In crusted scabies, topical scabicides should be applied from head to toe including the scalp, face, ears and flexures and left in contact with the skin for the recommended amount of time (usually 12 hours for permethrin and 24 hours for malathion). Several treatments are usually needed. Special attention should be paid to the nails as they are frequently a haven for mites and thus a source of recurrence/treatment failure. Nails should thus be clipped short and any subungual debris removed. Topical agents should then be applied to the subungual space and nailfolds.

Secondary bacterial infection should be treated with appropriate antibiotics as this is a major cause of morbidity and even mortality.

Since topical treatments tend to cause irritation and xerosis of the skin they should be followed by the liberal use of emollients. A sedating antihistamine may be necessary for a few days to control the itching and allow for undisturbed sleep.

**Other control measures**

In cases where an institution is involved, environmental control measures should include patient isolation until therapy is completed, barrier precautions with long-sleeved gowns and gloves, and screening and synchronous treatment of infected caregivers and close contacts. Visitors should be limited to reduce opportunity for spread.

The bedding and clothing used by a person with crusted scabies should be collected and transported in a plastic bag and emptied directly into washer to avoid contaminating other surfaces and items; all items should be machine-washed and dried using hot water and high heat cycles (temperatures in excess of 50°C for 10 minutes will kill mites and eggs); laundry personnel should use protective garments and gloves when handling contaminated items. Items that cannot be washed should be sealed in a plastic bag for 72 hours.

Public health officials should be involved as soon as the diagnosis is made to guide the institution regarding best practice in this situation as well as to monitor the outbreak.
and make sure all contacts are traced and treated. Moreover, when the patient has been harbouring the infestation for many months (as in this case) it is essential to trace any staff that may have left for another job in the meantime. This is important both for the individuals involved as well as to avoid transfer of the infestation to other institutions. Similarly, this should also be applied to any residents that may have recently left the facility.

Conclusion
Crusted scabies is a distinct form of scabies affecting patients who are debilitated and it is a severe form of infestation with the mite *Sarcoptes scabiei* var. *hominis*. It often poses a challenge as it is frequently misdiagnosed for other dermatoses. Once diagnosed there are also pitfalls in the treatment due to the number of organisms present as well as to general conditions affecting this subgroup of patients. A combination of topical and systemic treatment is necessary for adequate control and this must be accompanied by public health measures. This case clearly demonstrates the consequences of a delay in diagnosis as well as the highly contagious nature of crusted scabies as it resulted in a large outbreak affecting many of the residents as well as members of staff and took several months to be completely eradicated. It is therefore very important for healthcare staff to be aware of the possibility of such a diagnosis and maintain a high index of suspicion, if necessary referring early for specialist dermatological review so that timely treatment may be instituted.

*Figure 1: Crusty plaques on scalp.*
Fig. 2: Hyperkeratotic, fissured hands

References