Journal of Pakistan Association of Dermatologists. 2023;**33(3)**:1145-1150.

Case Report

Crusted scabies in an immunocompetent child: A rare neglected tropical disease

Larasati Budiyarto, Devi Arofah Mumtazah

Department of Dermatology and Venereology, Fatmawati Central General Hospital, Jakarta, Indonesia.

Abstract Crusted scabies is an uncommon and debilitating type of scabies that is distinguished by hyperinfestation of millions mites and the clinical hallmark of hyperkeratotic skin crust. This is a case report of crusted scabies in a 10-year-old immunocompetent female child. The case was first misdiagnosed at the previous healthcare center as ichthyosis vulgaris and was given prolonged topical corticosteroid with no improvement. Microscopic examination of skin scrapings revealed multiple Sarcoptes scabiei and their eggs. Diagnosis of crusted scabies was made. Patient's clinical outcomes improved after receiving combination therapy of oral ivermectin, topical permethrin 5% and topical keratolytic agents. Misdiagnosis of crusted scabies may pose risk of uncontrolled infestation and perpetuate an outbreak in society. It is also associated with complication of secondary bacterial infection. Therefore, it is important for clinicians to acknowledge the diagnosis and management of crusted scabies to improve clinical outcomes and control disease transmission.

Key words
Crusted scabies; Diagnosis; Treatment.

Introduction

Crusted scabies is a rare and severely debilitating form of scabies, an infestation of ectoparasite caused by the mite, characterized by an ectoparasitic infestation of millions of *Sarcoptes scabiei var. hominis* mites and the formation of a hyperkeratotic skin crust. It was initially reported in 1848 by Boeck and Danielssen in leprosy patients in Norway and was termed by Von Hebra as Norwegian scabies. Crusted scabies is prevalent in several isolated Aboriginal communities in Australia's Northern Territory (NT) with reported prevalence of up to 24/10.000.[1]

Address for correspondence
Dr. Larasati Budiyarto
Dermatology and Venereology Department,
Fatmawati Central General Hospital,
Jakarta, 12430, Indonesia
Ph: +6281223704604
Email: larasatib22@gmail.com

This type of scabies often affected individuals with dementia, disability in learning, immunosuppression, or HIV, also commonly associated with Down syndrome. Overcrowding, institutionalization (elderly in residential and nursing facilities), poor hygiene, poverty, sexual promiscuity, and undernourishment have been proposed as risk factors. Scabies also disproportionately affects children in low-income and middle-income nations and tropical regions.[2] In 2017, WHO listed scabies and other ectoparasites as Neglected Tropical Diseases (NTDs) to better develop control strategies and scabies outbreak response plans.[3]

Case report

A 10-year-old female child presented with generalized dry skin, hyperkeratotic scales, and thick yellow crusted plaques, accompanied with pruritus of the body. Two years ago, the lesion started as dry skin and scales on the lower back, which then slowly spread to other parts of the

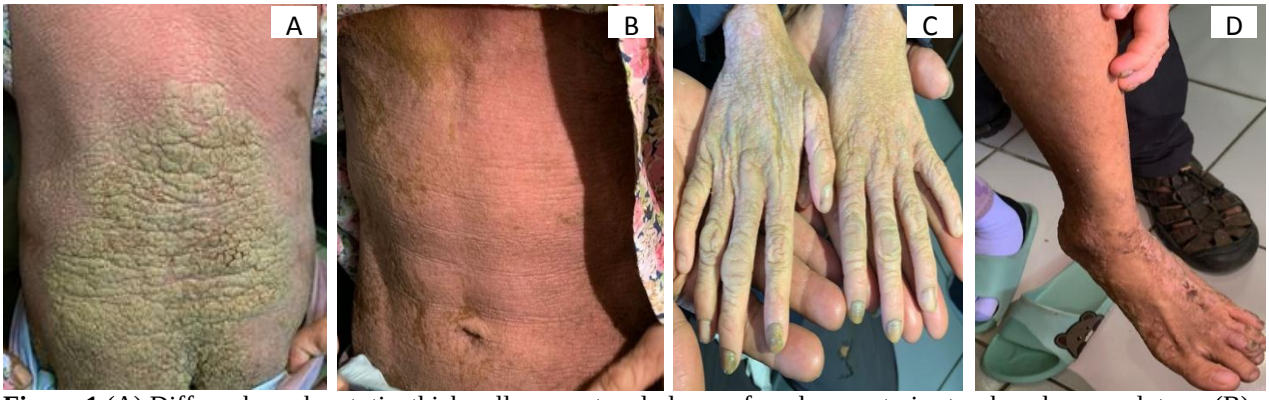

Figure 1 (A) Diffuse, hyperkeratotic, thick yellow crust and plaques found on posterior trunk and upper gluteus, (B) Anterior trunk showed generalized diffuse erythematous patches with fissures and lichenification, (C) Fingernails on the hand showed yellow discoloration, hyperkeratotic subungual and dystrophic changes, (D) Legs and feet presented diffuse erythematous patches with multiple hyperkeratotic plaque, fissures and lichenification.

body. Patient also complained of severe nocturnal pruritus accompanied by crawling sensation underneath her skin. Patient's mother and brother had an erythematous papular eruption with intensified pruritus at night, affecting the interdigital web spaces and lateral aspects of fingers without any presence of crusted lesions. History of cognitive impairment, immunodeficiency disorders, use of immunosuppressant drugs, sensory and mobility impairment, and other chronic medical diseases were denied.

One year before consultation patient sought for medical care from dermatologist at the previous hospital. Biopsy of the skin was done and revealed skin tissue lined with stratified squamous epithelium, hyperkeratosis, acanthosis, hypergranulosis, epidermolysis and intraepidermal blister. Subepithelial layer contain fibrous connective tissue. Biopsy result concluded hyperkeratotic epidermolysis with no signs of malignancy. Patient was initially diagnosed with ichthyosis vulgaris and was prescribed with topical corticosteroid treatment (alternating between mometasone furoate 1% cream and clobetasol 0,05% cream) and oral antihistamine for one year. Itchy sensation was slightly reduced but the scales on the skin progressed and thickened. Persistence of lesions prompted a referral for this patient to our hospital.

Physical examination revealed unremarkable general condition. Patient had normal nutritional status (25kg of weight with 125cm of height). Extensive generalized diffuse erythematous hyperkeratotic patches with fissures and lichenification were observed on trunk, arms, forearms, legs, hands, and feet. Diffuse, hyperkeratotic, thick yellow crust and plaques were found on posterior trunk and upper gluteus. Some of the lesions on feet were excoriated. Fingernails on the hand showed yellow discoloration, subungual hyperkeratotic and dystrophic changes. Face, scalp, and neck were spared from lesions. Lymphadenopathy was not found on examination.

Microscopic examination from the skin scraping with sample taken from interdigital web spaces of hand and thick crust on posterior trunk dissolved in 20% potassium hydroxide (KOH) revealed the presence of multiple *Sarcoptes scabiei* and their eggs. Blood tests showed hypereosinophilia with total eosinophil count = 2310 (normal value range: 62-200) and significantly elevated total IgE= 43.292 IU/mL

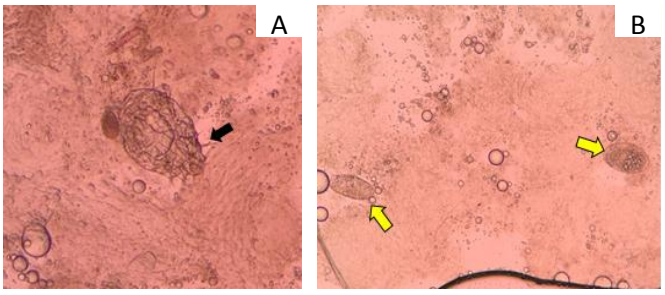

Figure 2 Microscopic examination of skin scrapings from interdigitalis and thick crust from posterior trunk dissolved with 20% KOH revealed (A) Adult *Sarcoptes scabiei* (black arrow) laying egg and scybala (B) Eggs of *Sarcoptes scabiei* (yellow arrows).

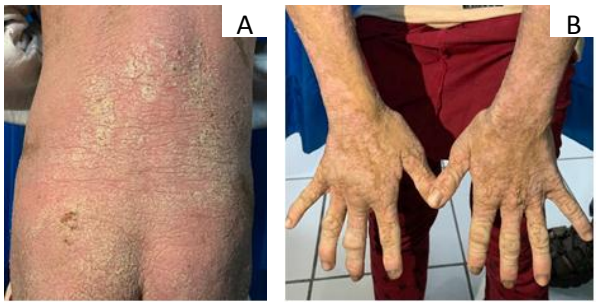

Figure 3 One week follow up consultation after therapy showed that thick yellow crust previously found became thinner on (A) posterior trunk and upper gluteus, and (B) arms and back of hands.

(normal value range <200 IU/mL). Other routine laboratory examination were within normal limits.

A diagnosis of crusted scabies (Norwegian scabies) was made. Treatment was initiated with 200 µg / kg oral ivermectin prescribed for day 1, 2, 8, 9, and 15 of therapy. Topical permethrin 5% cream was instructed to be applied daily for the first week and twice a week on the following week. Topical keratolytic (salicylic acid 7% combined with menthol 0.5%) was prescribed to be applied twice a day. Household members and other close contacts of the patient were advised to undergo topical therapy of permethrin 5% cream once at the same time. To avoid transmission and re-infestation of disease, patient's family was instructed to clean their house. Bedding, clothes, and other similar goods must be laundered in hot water or stored in plastic sealed containers for 10 days. A week after therapy, pruritus has significantly improved and thick hyperkeratotic scales became thinner.

Discussion

Crusted scabies, also known as Norwegian scabies, is a clinically distinct and extremely contagious subtype of scabies. It poses various clinical issues, including challenges in diagnostic and treatment, as well as a specific public health issue.[4] Crusted scabies develops when hyperinfestation of *Sarcoptes scabiei* occurs and the host's immune response to the mite is insufficient, such as in immunosuppressed individuals, patients with chronic disease, sensory dysfunction of the skin and physical impairment. Immunosuppression may occur due to use of drugs (topical and systemic corticosteroids, cytostatic agents, biological agents and immunosuppressants), HTLV-1 or HIV infection, congenital immune deficiencies, lymphoma, graft-versus-host disease, leukemia, and Down syndrome. Chronic diseases including diabetes mellitus, end-stage renal failure, liver disease, undernutrition and malnutrition may pose risk on developing crusted scabies. Patients with skin sensory dysfunction (leprosy, spinal cord injury, and sensory neuropathy) and physical impairment (paraplegia, paresis, epidermolysis bullosa, severe arthropathy) may also raise the risk of crusted scabies.[5]

This case portrays a crusted scabies infestation in an immunocompetent child with no history of other medical conditions nor drugs which may compromise skin sensory function and physical mobility. Based on studies, children may have a greater frequency of developing this disease due to bed sharing, overcrowding, high reinfestation rates, and underdiagnosed illness.[6]

Clinical presentations in this case suit the major

manifestations of crusted scabies, except for the severe nocturnal pruritus. Despite a high mite burden of up to 4700 mites per gram of shredded skin, itchy sensation is frequently absent. The defining hallmarks of crusted scabies include formation of hyperkeratotic skin scales, which can be loose, scaly, and flaky or thick and adherent. An inflammatory response imbalance in the dermis results in elevated levels of IL-4, which is responsible for skin hyperkeratosis. Skin flakes containing thousands of mites can be shed into bed linen or floors on a regular basis. Although the hands and feet are the most usually affected locations, the distribution is typically widespread, including the neck, scalp, and face, as well as the trunk and limbs, particularly the knees and elbows. Thick debris and mite deposits build beneath the nails, which are frequently dystrophic and thickened. In certain situations, crusting may be restricted to one or more limbs, the back of the fingers and hands, or only the buttocks. The skin of the trunk is often very xerotic. Fissures and secondary bacterial infection are prevalent, as well as regional lymphadenopathy in severe cases. Crusted scabies can occasionally show as erythroderma.[6–9]

We performed skin scraping for microscopic examination added with KOH 20% which revealed presence of multiple *Sarcoptes scabiei* and their eggs, typically seen in high numbers. This provides a conclusive diagnosis and remains as the gold standard examination. Potassium hydroxide may improve inspection of the material as it dissolves excess keratotic debris. Blood tests showed hypereosinophilia and significantly elevated total IgE. Peripheral blood eosinophilia is prevalent but not always present, and serum immunoglobulin E (IgE) levels are frequently exceedingly high.[6-10]

Skin biopsy is frequently unnecessary and only performed to rule out other possible diagnoses.[11] Histopathological investigation from skin biopsy of crusted scabies may reveal a severely thickened scale crust. Numerous *Sarcoptes scabiei* mites were able to be seen in stratum corneum, as well as acanthosis and a mixed inflammatory infiltrate (lymphocytes and histiocytes with or without neutrophils and eosinophils) in the papillary dermis.[12,13]

Crusted scabies should be differentiated with other diseases with similar presentations such as hyperkeratotic eczema, psoriatic erythroderma, contact dermatitis, ichthyosis, dyshidrotic eczema, atopic dermatitis, seborrheic dermatitis, palmoplantar keratoderma, Darier's disease, pityriasis rubra pilaris, Sézary syndrome, and erythrodermic mycosis fungoides.[14,15] Many of those differential diagnoses are treated with steroids (both topical and systemic), which may further delay the accurate diagnosis and management. This case was first misdiagnosed as ichthyosis vulgaris at previous hospital and was treated with topical corticosteroid for a year. Prolonged use of topical steroid may lead to an atypical presentation of scabies, also known as 'scabies incognito' and may cause iatrogenic immunosuppression which resulted in the conversion of classic scabies to crusted scabies.[16,17] Misdiagnosis poses risk of uncontrolled infection and transmission, and also associated with secondary bacterial infections, most notably *S. aureus*.[16]

Crusted scabies can be difficult to cure due to high concentration of scabies mites in the epidermis and hyperkeratotic skin.[15] To reduce the mites and penetrate thick scale, crusted scabies requires frequent simultaneous oral and topical treatments. The Centers for Disease Control and Prevention (CDC) currently advises treatment with a combination of oral ivermectin (200 ug/kg on days 1, 2, 8, 9, and 15, and possibly days 22 and 29) and topically applied 5% permethrin or 25% benzoyl benzoate (daily

for 7 days, then twice weekly until lesions healed). Ivermectin is solely acaricidal and not ovicidal; thus, should be repeated after 7 to 14 days to kill all larvae that were still unhatched at the time of the initial treatment. Furthermore, topical keratolysis containing urea 10% with lactic acid 5% or 5-10% salicylic acid ointments as an adjunct treatment is considered necessary to aid in the penetration of scabicides and removal of scales. Severe hyperkeratosis may need to be physically removed through debridement surgery.[5,6,9,18]

Another study proposed a simple clinical grading scale to guide the steps in treating patients with crusted scabies. Body surface areas, depth of skin crusting, prior episodes, hospitalizations, degree of skin cracking, and pyoderma have all been used to establish grading scales for crusted scabies. Each domain is rated 1 for mild until 3 for severe, and the scores are added together to generate an overall score: grade 1 (4-6), grade 2 (7-9), and grade 3 (10-12).[9,19] In this case, crusts formed on upper body, forearms, lower legs, buttocks, and trunks, estimated 10-30% of BSA. The depth of crust approximately exceeded 5 to 10 mm with moderate skin shedding. Patient has never been hospitalized due to crusted scabies before. Superficial skin cracking and multiple sores were found on both of lower legs and back of feet. This case can be concluded as moderate crusted scabies (grade 2). The study suggested oral ivermectin 200 ug/kg for 5 doses given on days 0,1,7,8,14, which was in line with proposed treatment by CDC.[19]

Avoiding direct skin-to-skin contact with an infested individual and their personal belongings can help prevent scabies transmission. Transmission by fomites may be more common in crusted scabies, where mites are more abundant and may persist in shedded scale. Mites in crusted scabies can survive for up to seven days by feeding on sloughed skin. Objects, linens, and clothings used by patient during this time should either be placed in a plastic bag for at least 72 hours until 10 days (in crusted scabies) or machine washed in hot water (at least 60 ^{o}C) and machine dried or dry-cleaned. To avoid spreading crusted scabies, the infested individual must be isolated, hospitalization may also be considered. Regardless of symptoms, simultaneous scabies treatment is indicated for both patients and their close contacts.[6,9,10,20]

Scabies infestation is also frequently worsened by subsequent bacterial infection due to scratching-induced skin damage. In crusted scabies, systemic complications include sepsis due to Gram-negative organisms such as *Pseudomonas aeruginosa* which may develop alone or as a polymicrobial sepsis in conjunction with *S. pyogenes* and/or *S. aureus*. Secondary bacterial sepsis including acute poststreptococcal glomerulonephritis and systemic sepsis has a significant death rate, especially in untreated crusted scabies.[6,7]

Conclusion

In conclusion, we reported a case of crusted scabies in an immunocompetent child which was misdiagnosed earlier as ichthyosis vulgaris and was treated with prolonged topical steroid therapy. Early, accurate diagnosis and treatment should be conducted for crusted scabies as clinical progress of the disease may result in multiple complications. Crusted scabies is also known to perpetuate an outbreak in society due to its high load of mites; therefore, prevention of transmission should also be highlighted in its management.

References

1. Glennie M, Gardner K, Dowden M, Currie BJ. Active case detection methods for

crusted scabies and leprosy: A systematic review. *PLoS Negl Trop Dis*. 2021;**15(7)**:1-17 .
2. Griffiths CEM, Bleiker TO, Creamer D, Ingram JR, Simpson RC. Rook's Dermatology Handbook [Internet]. Wiley Blackwell; 2022. Available from: www.wiley.com
3. Mitra AK, Mawson AR. Neglected tropical diseases: Epidemiology and global burden. *Trop Med Infect Dis*. 2017;**2(3)**:36; https://doi.org/10.3390/tropicalmed2030036
4. Apap C, Piscopo T, J.Boffa M. Crusted (Norwegian) scabies treated with oral ivermectin: A case report and overview. *Malta Medical J*. 2013:**4**;49.
5. Sunderkötter C, Wohlrab J, Hamm H. Epidemiologie, Diagnostik und Therapie der Skabies. *Dtsch Arztebl Int*. 2021;**118(41)**:695–704.
6. Thomas C, Coates SJ, Engelman D, Chosidow O, Chang AY. Ectoparasites: Scabies. J Am Acad Dermatol. 2020;**82**: 533–48.
7. Currie BJ, Hengge UR. Scabies. In: Tropical Dermatology: Second Edition. Elsevier Inc.; 2016. p. 377–86.
8. Arora P, Rudnicka L, Sar-Pomian M, Wollina U, Jafferany M, Lotti T, *et al*. Scabies: A comprehensive review and current perspectives. Vol. 33, Dermatologic Therapy. Blackwell Publishing Inc.; 2020.
9. Richards RN. Scabies diagnostic and therapeutic update. *J Cutan Med Surg*. 2021;**25(1)**:95-101. doi: 10.1177/1203475420960446.
10. Leung AKC, Lam JM, Leong KF. Scabies: A Neglected Global Disease. *Curr Pediatr Rev*. 2019;**16(1)**:33–42.
11. al -Tarawneh AH, Shamasneh A. Crusted (Norwegian) Scabies Mimicking Psoriasis: A Case Report and Literature Review. *Bahrain Medical Bulletin*. 2021;**43(3)**:648-50.
12. Sánchez-Borges M, González-Aveledo L, Capriles-Hulett A, Caballero-Fonseca F. Scabies, crusted (Norwegian) scabies and the diagnosis of mite sensitisation. Vol. 46, Allergologia et Immunopathologia. Elsevier Doyma; 2018. p. 276–80.
13. Wang MK, Chin-Yee B, Lo CKL, Lee S, El-Helou P, Alowami S, *et al*. Crusted scabies in a renal transplant recipient treated with daily ivermectin: A case report and literature review. *Transpl Infect Dis*. 2019;**21(3)**: e13077.
14. Talaga-Ćwiertnia K. Sarcoptes infestation. What is already known, and what is new about scabies at the beginning of the third decade of the 21st century? *Pathogens*. 2021;**10(7)**:868; https://doi.org/10.3390/pathogens10070868.
15. Widaty S, Rachmawati Y. Erythrodermic Manifestation due to Hyperinfestation of Scabies. *Periodic Dermatol Venereol*. 2021;**33(2)**:141-4.
16. Aukerman W, Curfman K, Urias D, Shayesteh K. Norwegian Scabies management after prolonged disease course: A case report. *Int J Surg Case Rep*. 2019;**61**:180–3.
17. Zorbozan O, Gerçeker Türk B, Acar A, Oraloğlu G, Ünver A, Töz S, *et al*. Comparison of Skin Scraping and Standard Superficial Skin Biopsy in the Laboratory Diagnosis of Scabies. *Turkiye Parazitol Derg*. 2020;**44(3)**:164–7.
18. Talty R, Micevic G, Damsky W, King BA. Erythrodermic scabies in an immunocompetent patient. *JAAD Case Rep*. 2022:112–115.
19. Davis JS, McGloughlin S, Tong SYC, Walton SF, Currie BJ. A Novel Clinical Grading Scale to Guide the Management of Crusted Scabies. *PLoS Negl Trop Dis*. 2013;**7(9)**:e2387.
20. Aždajić MD, Bešlić I, Gašić A, Ferara N, Pedić L, Lugović-Mihić L. Increased Scabies Incidence at the Beginning of the 21st Century: What Do Reports from Europe and the World Show? *Life*. 2022;**12(10)**:1598; https://doi.org/10.3390/life12101598