Configuration of Atypical Scabies as The Great Imitator

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Abstract: Scabies is still remaining a public health problem, especially in temperate regions tropical and subtropical. WHO includes scabies as a Neglected Tropical disease that needs large-scale control. There are various forms of atypical scabies that are difficult to identify which can lead to a misdiagnosis. This misdiagnosis also leads to the untreated well. The purpose of this article is to review the configuration of scabies as the great imitator. The atypical forms of scabies are scabies of cultivated, incognito scabies, nodular scabies, blousie scabies, animal transmitted scabies, bedridden scabies, scabies on AIDS patients, scabies with STD coinfection, scabies on baby and elderly, and crusted scabies. Among these forms, crusted scabies is the latest in diagnosis, and it often results in outbreaks at the endemic area.

Keywords: Scabies, Great Imitator, Neglected Tropical Disease

1. Introduction

Scabies is a skin disease caused by mites namely Sarcoptes scabiei (S.scabiei) hominis variety. This disease is a public health problem, especially in temperate regions tropical and subtropical. The number of people with scabies in the world is more than 300 million every year with figures that vary from any country. Because of that, WHO include scabies as Neglected Tropical Disease. (Hengge, 2006) In Indonesia, scabies is one of the most common skin diseases at the primary health care. The prevalence of scabies in health centers throughout Indonesia in 2018 was 5.6-12.9% and was the third most of skin disease. (Azizah, 2011) In the year of 2014, a research conveyed the prevalence of scabies in a islamic boarding school at South Jakarta was 68%. (Soedarman, 2014)

Itching sensation is the main clinical symptom of scabies. The initial period of mite infestation usually occurs at night (pruritus nokturna), hot weather, or when sweating. Itching felt all around lesions, but in chronic scabies itching can be felt up to whole body. Itching is caused by skin sensitization to excretions and mite secretions that are released when making tunnels. The incubation period from mite infestation to symptoms of itching around 14 days. (Hay, 2012)

Scabies is a skin disease with clinical manifestations often resembles other skin diseases so it is called the great imitator. There are various forms of atypical scabies that are difficult to identify which can lead to a misdiagnosis. This misdiagnosis also leads to untreated well. In Indonesia, there are still few articles that discuss atypical forms of scabies. The purpose of this article is to review the configuration of scabies as the great imitator.

2. Methods

A comprehensive search of the PubMed database was carried out in December 2021 to January 2022 uses the keywords 'Scabies' and 'Configuration' or 'Manifestation'. All results obtained manually reviewed for content, relevance and included in the review when deemed appropriate. Paper cited in the bibliography was also reviewed and included if deemed adequate. This article was manual excludes when there were duplication of script.

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3. Result

*S. scabiei* usually chooses thin epidermal locations for digging tunnels, for example between the fingers, wrists, hand, penis, areola mammae, peri-umbilical, breast fold, waist, lower intergluteal buttocks, thighs and axillary folds anterior and posterior. Tunnels dug by mites appear as the lesion is a fine line that is grayish-white throughout 2-15 mm, winding and slightly elevated compared to the surroundings. At the end of the tunnel are small papules or vesicles <5 mm in size where the mites are. In tropical climates, lesions are rare tunnel; even if there is a tunnel only about a short size 1-2 mm. These lesions are difficult to find because they are often accompanied by excoration due to scratching and secondary infection by bacteria. Nevertheless, tunnels can be in the hand, between the fingers, the wrist hands and ankles. Pustules without frequent tunnel lesions found in the external genitalia. In mild infestations, the location should be examined were between the fingers and external genitalia. (Golant, 2012)

In adults, scabies lesions are rarely found on the neck, face, hairy scalp, upper back, soles of the feet and hand; but in infants the area is often infested and even lesions can be found all over the body. Scabies lesions are usually absent on the head but in small children and babies can be found a pustul itch. Symptoms of scabies in children usually include vesicles, pustules, and node; the child becomes restless and has a reduced appetite. An overview clinical scabies in children is often difficult to distinguish from infantile acropustulosis and bullous vesicul dermatitis. Tunnel lesions are rare or not even found. (CDC, 2012)

Scabies causes intense itching so that sufferers often scratching and abrasions followed by infection secondary bacteria by *Group A Streptococci* (GAS) and *S. aureus*. The infection causes pustules, excoriation and enlargement lymph gland. In secondary infection by *S. aureus* can raised bullae so it is called bullous scabies. In tropical countries often secondary bacterial infection occurs with pustular lesions or crusts in the predilection area for scabies and in children the lesions are on the face. These secondary infectious lesions are similar to impetigo. Scabies with secondary infection should be treated first before giving a scabisida. (CDC, 2012)

The severity of scabies depends on the number of mites and management. If diagnosis and treatment are delayed, then the number of mites increases and the symptoms become more severe. Weight the degree of skin damage depends on the degree of sensitization, duration infection, individual hygiene, and previous treatment history. In the chronic stage, scabies results in thickening of the skin (lichenification) and darker in color (hyperpigmentation). (Hicks, 2009)

Scabies of cultivated usually found in people with good hygiene. Scabies sufferers complain of itching in the predilection area of scabies like between the fingers and wrists. Itching feeling usually not too heavy. Manifestations of scabies in clean people is a lesion with a small number of papules and tunnels so it is difficult to identify and often misdiagnosed due to atypical symptoms. From the tunnel of 1000 sufferers scabies of cultivated, only 7% of the tunnels were found. (Sungkar, 2016)

Incognito scabies often shows no clinical symptoms usual, atypical distribution, extensive and similar lesions to other diseases. Form of incognito is found in scabies treated with corticosteroids so that the clinical symptoms and signs improve, but the mites remain and can still transmit scabies. On the other hand, treatment long-term topical steroids make the lesions worse due to decreased cellular immune response. The use of corticosteroids applied to the skin is capable affect the immune system such as decreased inflammatory response and suppressed cellular immune response. Topical corticosteroids inhibits the production and release of cytokines such as interleukins IL-1,2,3,6 and tumor necrosis alpha (TNFa). Use of steroids topical inhibits phagocytosis and stabilization of the cell membrane of phagocytes lysosomes so that the pro-inflammatory and phagocytic response is inhibited and ultimately unable to control scabies infestation. (Karaca, 2009)
4. Discussion

Karaca et al reported a woman aged 75 years who came to an outpatient clinic with complaints of itching and reddish plaques accompanied by resilience in the scalp area. Sufferers have been given antihistamines and clobetasol propionate ointment for three weeks. The sufferer was initially diagnosed with xerosis and pruritus but after being given the treatment the complaint getting great. On physical examination, there was a lesion of shape annular, serpiginous, vesicles, erythematous pustules, excretion due scratching marks, as well as excoriated papules around the armpits and body. (Karaca, 2009)

Blood laboratory examination will show normal results but microscopic results of reddish plaque scrapings were found S. scabiei and their eggs in various fields of view. Sufferers diagnosed as incognito scabies and treated with ointment keratolytic 2x / day and 5% permethrin lotion. One of the manifestations of incognito scabies is subcomenal pustular dermatosis-like eruption characterized by eruption pustular in normal skin or slightly reddish. Pustular eruption typically found in the flexor and proximal areas of the limb, cause itching and irritation. Pustules are located on the surface skin, easily breaks and gives rise to superficial, shaped crusts annular like a circle or serpiginous pattern. Immunosuppressive therapy can obscure the clinical picture scabies and gives an atypical clinical picture like incognito scabies. In infants, the elderly, and immuno compromised host, all skin surfaces can be infested including the scalp and face. In incognito scabies can be found lesions on the scalp without the itching sensation that distinguishes it with classic scabies manifestations. (Karaca, 2009)

Scabies nodularis was first reported in 1923 by Ayres and Anderson. It is called scabies nodularis because of the lesions in the form of reddish-brown, itchy nodes in areas covered by clothing. The formation of nodes is caused by a skin hypersensitivity reaction against S. scabiei and its products. Nodular lesions occur in 7-10% scabies sufferer. The nodes have a diameter of 5-20mm and are tunnels usually found early in the formation of nodes. Mites are rare found in the node. (CDC, 2012)

In the classification of diseases, scabies nodularis is a part of pseudo cutaneous lymphoma, along with persistent nodular arthropodite reactions and lymphomatoid contact dermatitis. Pseudolymphoma cutaneous is a heterogeneous group of lymphoproliferative processes of T lymphocytes or B lymphocytes which can be caused by various conditions and resemble cutaneous lymphoma both clinically and histologically. Cutaneous pseudolymph is divided into two, namely cutaneous pseudolymphoma cells T lymphocytes and B lymphocytes. Nodular scabies includes pseudolymphoma cutaneous T lymphocyte cells which mean histologically the inflammatory infiltrate arises in nodular scabies dominated by T lymphocyte cell components. In nodular scabies, superficial and deep dermis demonstrated moderate perivascular and interstitial nodular infiltrates to solid consisting of lymphocytes (predominantly T lymphocyte cells), histiocytes, plasma cells and eosinophils. In addition, large atypical mononuclear cells which resembles reed-stemberg cells can also be found. Overall nodular scabies may mimic Hodgkin's lymphoma or non-hodgkin. (Ploysangam, 1998)

The predilection for scabies nodularis is in the penis, scrotum, axilla, wrists, elbows, areola mammae, and stomach. After therapy, the appearance of the skin is similar to the healing condition of an eczematous eruption. Scabies nodes can last for several months or even several years despite having been given anti scabies drugs. Cause persistent nodes are not known with certainty but are suspected as a result of a slow-type hypersensitivity reaction to the components scabies mite. Because anti-rabies drugs are not effective for scabies nodularis, then the treatment is by injecting corticosteroids intralesional. Despite this, scabies nodes can persist for as long several months even up to a year even though it has been given scabicides and corticosteroids. (CDC, 2012)

Scabies infesting infants and immunocompromised individuals are more likely to develop bullous scabies. Bullae that are formed are similar to bullae in bullous pemphigoid namely a skin disease characterized by large blisters. Although bullous scabies is similar clinically and histopathologically to
pemphigoid bullous, they are not similar when examined by immunofluorescence either directly or indirectly. Another difference between bullous scabies with bullous pemphigoid is the location of the lesion, symptoms, and age of the patient. Bullous scabies is usually spread between the fingers, hands, wrists, and genitals while pemphigoid is bullous spread over the body and extremities. (Maan, 2015)

Symptoms of bullous scabies are nighttime itching and itching positive family history of scabies while sufferers of bullous pemphigoid usually complains of itching all day long and no family suffering from scabies. Bullous scabies can affect all ages whereas pemphigoid bullous is more common in older people advanced and rare in children and adults. Scabies bullae itching and the walls of the bullae may become tense or loose. In the moon can be found blood; bullae can also crusted. Scabies predilection bullosa is the same as typical scabies. (Maan, 2015)

Bullal lesions in typical scabies are rare. The cause of its formation bullae are superinfection of mites by S. aureus bacteria as in bullous pemphigoid. Auto-mediated bullae formation antibodies may result from exposure to basement membrane zone antigens as a result mechanical wounds caused by mite bites or enzyme lysis. Another possibility is the cross-reaction between mite antigens and basement membrane zone antigen. Bullae may also form due to the scabid process, namely the body's reaction to mites. Scabies diagnosis bullous should be considered in all cases of skin disease have bullae accompanied by papules and itching that is resistant to steroid treatment. (Aulia, 2012)

The treatment for bullous scabies is the same as the treatment for scabies in general, namely using 5% permethrin cream, krotamiton 10%, benzyl benzoate, 6% sulfur or ivermectin. Although the mechanism bullae formation is mediated by the immune system, bullous scabies is not response to glucocorticoids and immunoglobulins. (Maan, 2015)

Scabies can infect animals such as dogs, horses, goats, rabbits, monkeys, and others. The main source of scabies in animals in America is a dog. The causes of scabies in animals are similar with those that infest humans but different strains. Human can transmit scabies to pets, but more so often is a cross-infestation from pets such as dogs to human. The symptoms of scabies that are transmitted through animals are different from classic scabies in humans. Animal scabies is absent tunnel, does not attack between the fingers and external genitalia. Location the lesions are usually at the place of contact when cuddling a pet ie the arms, chest, abdomen, and thighs. (CDC, 2012)

The animal scabies mite causes erythematous and pruritic papules. Mites live in deep human skin a short period of time but in immunodeficient patients the clinical manifestations of animal scabies in humans can be more severe. The mode of transmission of animal scabies is easier and the incubation period shorter. The lesions are temporary (4-8 weeks) and can heal itself because S. scabiei animal varieties cannot continue the cycle his life in humans. Prevention is done by preventing contact with the cause animal, treat the infected animal and bathe him cleanly and regularly. Two human populations that are more susceptible to contracting zoonotic scabies is a person who works with domestic animals and that keeping dogs. Transmission of domestic animal scabies to humans for example in pig breeders and animal slaughterers. (Ljunggren, 2005)

Scabies in people who are bedridden often found in people who suffer from chronic diseases or elderly people who lie in bed for a long period long time. The lesions in Bedridden scabies are only limited. The diagnosis of scabies in elderly patients is often delayed because the clinical manifestations are similar to other skin diseases so the diagnosis hard to define. To solve the difficulty of diagnosis, Katsumata et al sought an alternative test for diagnosing scabies in elderly sufferers who are bed rest for a long time. Katsumata et al used adhesive tape (duct tape) as a tool to find mites by attaching them to skin lesions which is a predilection for scabies, for example between the fingers. After being affixed, the tape is suddenly removed and then affixed to the object glass. The object glass is dripped with KOH and then examined
under a microscope. Detection mites with adhesive tape can be applied to people aged continued because it has thin and dry skin. (Katsumata, 2006)

In people with AIDS, atypical scabies and pneumonia are common Pneumocystis carinii. Atypical scabies diagnosis can be used as an indication of opportunistic infection-AIDS. HIV is a disease with a high prevalence in the world also with scabies. Fernandez-Sanchez et al made observations against cases of crustous scabies in HIV patients aged 28 years. Based on these observations it was concluded that scabies crustosa in immunocompromised patients who are being treated with antiretrovirals are part of the immune reconstitution spectrum inflammatory syndrome (IRIS). Atypical presentation of scabies in patients who are undergoing it previously did not appear to have antiretroviral treatment skin disease is an unmasking crusted scabies-associated IRIS. In this situation an infection appears that was not initially marked with excessive inflammation and atypical or present clinical presentation accelerated. This incident represents a restoration of specific immunity. Most cases of IRIS were associated with antiretroviral drugs with bacterial, viral and fungal infections but the infection can spread in parasitic infections such as leismaniasis, strongiloidiasis, schistomiasis, and toxoplasmosis. So crustous scabies in people with HIV in the treatment of ARVs can be considered a parasitic infection contribute to IRIS as well as a spectrum of IRIS. (Fernandez, 2012)

Scabies can be accompanied by other sexually transmitted diseases (STD) such as syphilis, gonorrhea, genital herpes, pediculosis pubis, and so on. Therefore, scabies lesions are found in the genital area a further examination is necessary in the form of a culture for gonorrhea and serologic testing for syphilis in persons with high risk.6 In typical scabies the tunnels and papules are frequent found in the glans penis, scrotum, and penis. (Buechner, 2002)

Scabies lesions in infants and the elderly can develop in palms, soles of the feet, face, and scalp. In people elderly mite infestations will become more severe. Skin lesions in scabies it is usually typical and gives intense itching especially at night but in infants, young children and people elderly the appearance of scabies can be atypical. Frequent atypical lesions resembling seborrheic dermatitis, eczematous dermatitis, impetigo, insect bites, and Langerhans cell histiocytosis (LCH). (Yang, 2015)

In a case report in Korea, a baby with scabies was diagnosed as LCH because of similar clinical and histopathological manifestations. In the report, a 6 months old baby girl had papules and reddish nodes on the body for one month. Baby gets corticosteroid cream therapy because it was initially diagnosed as contact dermatitis however the lesion extends rapidly to the area hands and feet with erythematous squama and plaque so an abdominal punch biopsy was performed. Histopathological examination showed cell infiltrates in the upper dermis with a perivascular distribution consisting of histiocytes, lymphocytes, and eosinophils. Histiocytes in the dermis above showed positive results with CD1a and S-100 staining so that the baby was diagnosed with LCH. Sometime later the baby's mother complained about the appearance lesions between the fingers and the accompanying periumbilical area itching feeling. Furthermore, the mother was carried out a dermoscopyassisted skin scraping on the palms and periumbilical. On Dermoscopy examination found eggs and scabies mites in both lesions so that the sufferer was diagnosed as scabies instead of LCH. Furthermore, the patient was treated with 10% crotamiton cream and recovered after one week of treatment. (Park, 2011)

The manifestations of infantile scabies resemble the lesions in disease other skin so that the diagnosis is difficult to establish on the basis alone clinical manifestations. The histopathological findings of scabies are sometimes not specific and do not support the diagnosis such as eczematous reactions in epidermis, perivascular area and inflammatory cell infiltrates in the dermis. The infiltrate cell usually shows positive histiocytes on CD1a stain and the S-100 which is often found in LCH. Based on these, clinicians must be able to distinguish infantile scabies from LCH through careful history, especially risk factors for scabies and examination supports such as CD30 stain, electron microscopy for find birbeck granules which are a distinctive sign of LCH and dermoscopy-assisted skin scraping for the diagnosis of scabies. (Park, 2011)
Crustous scabies is characterized by a crusted lesion wide, generalized scale and thick hyperkeratosis. Scabies crustosa was first reported by Danielsen and Boeck on 1848 in a Norwegian citizen who experienced Hansen morbus (leprosy) so that scabies crustosa is called also Norwegian scabies. Crustous scabies is common in people with retardation mental, senile dementia, and other neurological diseases. Besides crustous scabies is also often suffered by people with leukemia, patients who receive immunosuppressant therapy for example sufferers autoimmune or sufferers undergoing an organ transplant, and people with HIV-AIDS. Based on this, crustous scabies often associated with conditions of the less immune system (immunocompromised host). (Sungkar, 2016)

In crustous scabies, patients generally have deficiency immunology so that the immune system is unable to inhibit it proliferation so that the mites reproduce easily and fast. Crustous scabies almost always affects people who are are immunocompromised like the elderly, people with AIDS, mental retardation, lymphoma, and all other conditions can decrease the effectiveness of the immune system. The main symptom of classic scabies is intense itching especially felt at night. It's different from scabies. Classically, the itching of scabies crustosa is usually mild even none at all so that sufferers do not feel complaints which resulted in a late diagnosis. In late diagnosis scabies crustosa often results in outbreaks because the number of mites that infest sufferers is very large so it is very contagious. (David, 2013)

5. Conclusion
There are various forms of atypical scabies that are difficult to identify which can lead to a misdiagnosis. These forms are scabies of cultivated, incognito scabies, nodular scabies, bulose scabies, animal transmitted scabies, bedridden scabies, scabies on AIDS patient, scabies with STD coinfection, scabies on baby and elderly, and crusted scabies. Among these forms, crusted scabies is the most late in diagnosis, and it often results in outbreaks at endemic area.

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