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Neonatal Norwegian scabies: three cooperating causes

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Sarcoptes scabiei, scabies, topical

Abstract

Background: Norwegian or crusted scabies is seldom reported in infancy, usually in immune deficient patients.

Main Observations: We report a case of an infant affected by atopic dermatitis since birth. The patient was ineffectively treated with topical and systemic steroids for several weeks for the insurgence of cutaneous xerosis and erythema. Clinical inspection and optical microscopic examination of skin scraped scales led to the diagnosis of crusted scabies. The physiological inability to scratching reaction, the immunological profile deriving from atopy and finally iatrogenic immunodepression cooperating to conduce to crusted scabies.

Conclusion: In our case iatrogenic immunosuppression, atopic dermatitis and the absence of skin scraping reaction because of the very young age might have contributed to the unusual presentation of scabies. This case suggests considering possible alternative diagnosis of scabies in the failure of the treatment for atopic eczema.

Introduction

Human scabies is a mite infestation caused by an obligate human parasite of the phylum Arthropoda, *Sarcoptes scabiei* var. *Hominis*, that is directly transmitted between individuals by skin to skin contact or seldom through an indirect transmission¹ inducing a marked, predominantly nocturnal, itching with few to some papules in typical locations, such as interdigital folds, cubital margins of the hands, anterior side of the wrists, anterior axillae, around nipples and navel, external male genital organs and internal side of the thighs. It affects over 300 million individuals per year all over the world,² in some countries with prevalence among the general population ranging between 4% and 27%.³ However, in developed countries the incidence is much lower, although sexual behaviour, immigration, scarce hygiene and indigence are responsible of some epidemics, especially in adults and elderly. Norwegian or crusted scabies is a highly infectious form with a large number of mites infesting the epidermis as a result of the failure of the host immune response⁴ or of the lack of scratching response to pruritus⁵ that conduces to mite removal and burrow destruction. Absence of itching and unusual clinical presentations may sometimes lead to misdiagnoses.

Case report

A three month-old baby girl was admitted to our Department showing lesions affecting trunk, head, limbs, palms and soles, consisting of papules and small vesicles arising upon diffuse xerosis and erythroderma.

The baby was affected by atopic dermatitis since birth. In the last month a worsening in the cutaneous xerosis and erythema was reported and topical treatment with gentamicin and betamethasone was started to decrease atopic eczema rash. Because of the persistence of the skin lesions topical steroid daily schedule was increased and, in the last week, a systemic steroid (betamethasone 0.5 mg/day) was introduced because of the unresponsiveness to the topical therapy. The further worsening of the dermatitis (Fig. 1), together with the increased insomnia and agitation, induced the mother to seek a specialized medical advice.

The presence of monomorphic vesicles and the observation of some papules with linear shape draw the physician's attention to consider the possibility of skin infestation as possible differential diagnosis.

Dermoscopy was inconclusive (Fig. 2), but optical microscopic examination of scraped scales from some lesions

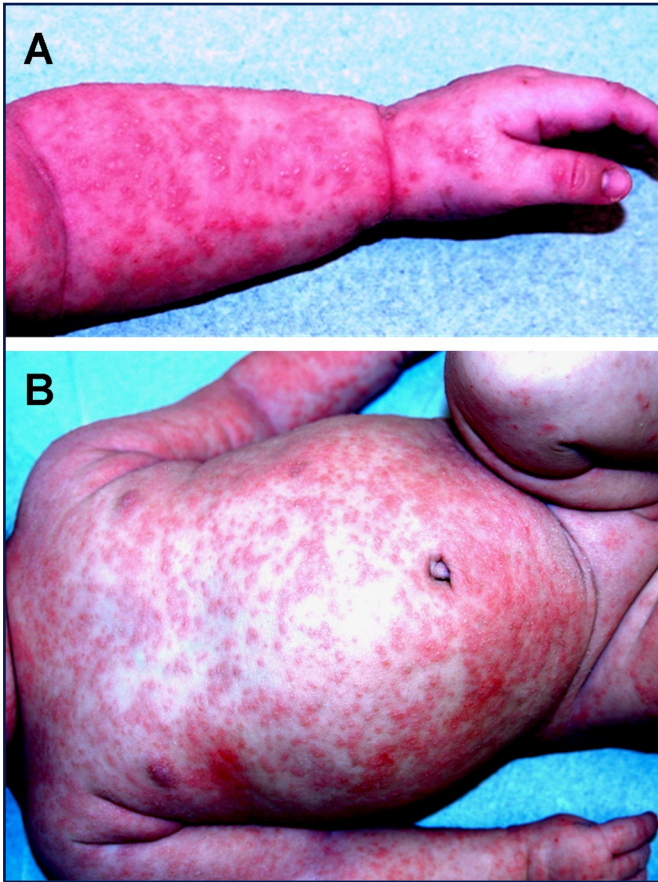


Figure 1
Clinical appearance of skin lesions (A and B).



Figure 2
Dermoscopy of skin lesions (20x).

showed numerous mites (Fig. 3). In the mother and the sister skin hitching and distinctive papules and burrows were revealed. The diagnosis of scabies was also confirmed by optical microscopic examination in the relatives.

Before treatment, blood examination of the baby showed increased serum IgE (400 KUI/L) and hyper eosinophilia (38.1%).

For the non availability of permethrin in our country and for the consideration of the systemic toxic effects of



Figure 3
Mite in light microscopy.



Figure 4
Clinical appearance of skin after therapy.

ivermectin in infants, a regimen of topical application of Benzyl benzoate 10% b.i.d. for 5 consecutive days, together with low potent steroid, was chosen, obtaining a complete healing of the papules and vesicles with a residual mild erythema and xerosis after 10 days (Fig. 4). A complete resolution was also obtained in the affected relatives by means of topical benzyl benzoate 30% b.i.d. for five days.

Discussion

Atopic dermatitis is the most common skin disease in infancy and childhood.⁶ The hallmark of the disease is pruritus localized in characteristic areas, depending on the age of presentation, skin xerosis and chronic relapsing course. Because of the pathogenetic role of the immune

system and the presence of compromised epidermal barrier, the non-etiological treatment is usually based on immunomodulating drugs and emollients. In our case the infant was treated with topic and systemic steroids with the clinical and anamnestic hypothesis of atopic dermatitis. However, it should be considered that in our geographical area crusted scabies is very seldom reported in infants. It occurs mostly in adults^{7,8} especially those suffering of immune deficiencies, which reduce humoral and cell-mediated immune responses⁹⁻¹¹ and allow the uncontrolled burrowing of the female mite in stratum corneum. Other predisposed adults are those presenting physiological or pathological conditions, such as Down syndrome or neurophysiological impairment, which preclude scratching response to pruritus and the mechanical mite removal.^{12,13} Patients chronically treated with immune suppressive drugs, i.e. post renal-transplant patients, or patients affected by HIV^{14,15} infection^{12,13} are also prone to develop this infestation. Also the lack of normal scratch response to pruritus leads to the inability to remove the mite and destroy the burrows. It has also been reported that mental retardation or long period application of topical corticosteroids could predisposed to crusted scabies.

In our case both the absence of skin scraping reaction for the very young age, both the iatrogenic suppression of the immune response, led to the unusual presentation, resulted deceptive for more than one month. Moreover, the presentation in the form of crusted scabies could also be due to the underlying skin conditions of the patient, since skin atopy is characterized by a preponderant Th2 lymphocyte response and increased interleukin 4 (IL-4) production, which it was reported to contribute to the mite proliferation in the epidermis.¹⁶⁻¹⁷ Recent evidence¹⁸ indicate that crusted scabies develops largely as a result of a Th 1/Th 2 imbalance, with a cytotoxic T-cell type 2 response in the skin, high levels of antibody in the blood, and uncontrolled growth of the parasite. Other studies display a significant increase of IgE, IgG, IgG1 and IgG4 in serum of affected patients in comparison with normal population,^{19,20} and these antibodies appear to play no role in protection. At the same time IgE response is usually accompanied to production of IgG4 in allergen specific disease as Atopic dermatitis as a consequence of production of Th2 cytokines IL-4 and IL-13.^{21,22}

In conclusion, the atopic dermatitis immunological pattern seemed to be the predisposing substrate on which the physiological inability to scratch and the iatrogenic immunodeficiency promoted the uncontrolled mite proliferation. This case also shows the importance to always consider all the differential diagnosis versus atopic dermatitis, taking care of an accurate skin examination especially in the failure of steroidal therapy.

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