

Qual o Seu Diagnóstico?

DERMATOSE COMUM NO PASSADO, MAS QUE AINDA DESAFIA OS DERMATOLOGISTAS - QUAL O SEU DIAGNÓSTICO?

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RESUMO – Os autores apresentam um caso de prurigo de Hebra, com o quadro típico numa criança com o mesmo perfil socioeconômico e de fatores predisponentes descritos por Hebra em Viena no século XIX. Essa doença se manifesta por múltiplas pápulas pequenas e extremamente pruriginosas. Devido à escoriação, ocorrem erosões e liquenificação e infecções secundárias, evoluindo para cicatrizes hiper ou hipopigmentadas.. As crianças afetadas geralmente são desnutridas e têm adenomegalias inguinal, axilar ou cervical. Normalmente, são crianças atópicas com hipersensibilidade cutânea a picada de insetos, num contexto de higiene inadequada e má nutrição.

PALAVRAS-CHAVE – Prurigo/diagnóstico, Doenças da pele.

Dermatology Quiz

A VERY COMMON DISEASE IN THE PAST THAT STILL CHALLENGES DERMATOLOGISTS - WHICH IS YOUR DIAGNOSIS?

ABSTRACT – This paper presents a case of Hebra's prurigo, with the typical picture in a child under similar socioeconomic conditions and predisposing factors described by Hebra in Vienna, in the nineteenth century. This disease is an eruption of multiple extremely itchy small papules. Because of scratching, there are abrasions or lichenification, frequent secondary infection and progression to scars with dyschromia. Affected children are usually malnourished and have increased inguinal, axillary or cervical lymph nodes. They are generally atopic children with hypersensitivity to insect bite, in a context of inadequate hygiene and poor nutrition.

KEY-WORDS – Prurigo/diagnosis; Skin diseases.

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CASE REPORT

A 6-year-old boy, from the countryside of Amazonas - Brazil, with a history of allergic reactions to insect bites since he was 6 months old, had little improvement on topical corticosteroids and oral antihistamines. The child has marasmus and disseminated highly pruritic rash composed of small erythematous papules with excoriation in recent lesions and lichenification with dyschromia in the older ones. Lesions predominated in extensor surface of limbs and face. He had painful lymphadenopathy in the axillary and inguinal region, and loss of weight (Fig. 1).

WHICH IS YOUR DIAGNOSIS?

- A. Persistent scabious nodules
- B. Hyde's prurigo (Nodular prurigo)
- C. Hebra's prurigo
- D. Atopic dermatitis

DIAGNOSIS - C. HEBRA'S PRURIGO

This is a case of Hebra's prurigo, presenting with the typical picture in a child living under similar socioeconomic conditions as those described by *Hebra* in Vienna in the 19th century. This rare disease consists of an eruption of grouped dried extremely itchy papules of 2-3mm, localized in the whole body, mainly in the extensor surface of upper limbs and face. Because of scratching, there are abrasions or lichenification, leaving scars with dyschromia. Secondary infection of the lesions may occur. The child affected is usually malnourished and has increased inguinal, axillary or cervical lymph nodes. They are generally atopic children with hypersensitivity to insect bite (experimentally induced) and positive skin tests to insect extracts.¹

Scabious nodules are persistent lesions in patients with evidence of current or previous scabies, and are not recurrent lesions after treatment of the underlying disease, although they may persist for months after treatment.²

Histologically, the lesions of Hyde's prurigo (nodular prurigo) and Hebra's prurigo may be identical. But the



Fig 1 - Hebra's prurigo. Multiple papules associated with signs of excoriation, lichenification and scaling, some with brownish or yellowish crusts, especially on the extensor surface of limbs and face.

key clinical features of Hyde's prurigo are large papules and more-or-less symmetrical nodules, 1-3 cm in diameter, with a raised, warty surface. Cases occur at all ages, but mainly from 20 to 60 years. Both sexes are equally affected and patients have intense, often distressing pruritus. The lesions may arise as a secondary event in a wide range of cutaneous and systemic diseases, but patient may have an idiopathic nodular prurigo.²

Atopic dermatitis presents as eczema (acute, subacute or chronic), usually without papules or nodules.²

MICROSCOPIC FINDINGS

Histopathological examination revealed interruption of the epidermis, replaced by fibrinopurulent exudate. The dermis showed perivascular inflammatory infiltrate consisting of eosinophils, lymphocytes and histiocytes, compatible with prurigo (Fig. 2).

During hospitalization, the patient was treated with systemic antihistamines, moisturizers for recovery of skin barrier function, topical antibiotic for secondary infection

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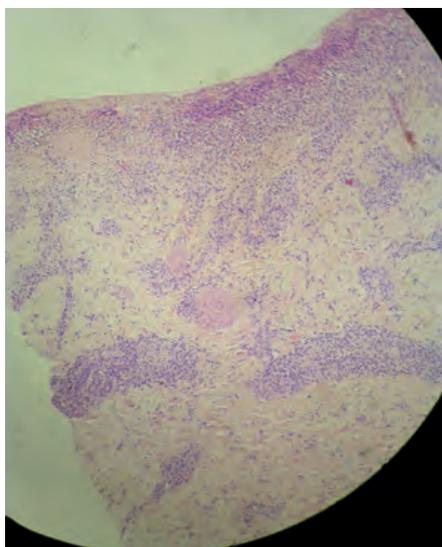


Fig 2 - Fibrinopurulent exudate and perivascular inflammatory infiltrate consisting of eosinophils, lymphocytes and histiocytes, compatible with prurigo. (H&E, 100x).

of the lesions and clinical support. There was improvement of pruritus and he was discharged with a recovery nutrition plan to be followed. With a prescription as outpatient which included moisturizers and repellent, there was significant improvement of the clinical picture.

DISCUSSION

Ferdinand von Hebra described the term prurigo referring to papules arising as a result of scratching.³ This definition could include conditions such as papular urticaria, dermatographism, among others. However, most of the original cases of prurigo described by Hebra referred to wheals arising probably after scratching in atopic children living in precarious social conditions and exposed to insect bites, especially fleas.^{3,4} Such lesions were initiated as tiny normochromic papules, more palpable than visible, which soon were modified by excoriation and eczema, becoming larger and erythematous.⁵ For this dermatosis, in 1892, *Besnier* gave the name "Hebra's prurigo".⁴

Usually, the clinical picture begins early in life with pruritic papules associated with erosion by excoriation. With chronicity, lichenification occurs and there may be a secondary infection with lymphadenopathy. Lesions predominate in the extensor areas of extremities, face, buttocks and abdomen, symmetrically. It is believed to have a multifactorial predisposition.⁶

Hebra's prurigo is considered a chronic and recurrent inflammatory skin disease, with cutaneous hyperreactivity to environmental stimuli (insect bites) possibly

enhanced by skin barrier dysfunction and is often associated with atopy. Skin hyperreactivity has intrinsic (genetic) and extrinsic multifactorial basis and is closely associated with inadequate hygiene and poor nutrition.⁶

A perivascular infiltrate in the central region of the wheal consisting of lymphocytes and eosinophils is observed both in new and old lesions but differences between new and old lesions are noted in histopathology. Recent lesions show hypergranulosis and normal stratum corneum, vasodilation with oedema and vascular endothelial cells protrusion in the lumen as well as increased numbers of eosinophils in the infiltrate, while the old lesions show with thickened collagen and decreased fibrocytes in the dermis. In old lesions there is hypogranulosis, acanthosis, papillomatosis and irregular hyperpigmentation of basal layer.⁷

Regarding immunology, *Almeida*⁴ showed that the serum of patients with Hebra's prurigo had increased immunoglobulins, especially IgE fraction (81% of patients), but also IgG (77% of them), which strengthens the possibility that Hebra's prurigo could be an atopic manifestation.

Therefore, based on the literature, this is a typical case of Hebra's prurigo, with clinical and histopathological features and within the profile of the socioeconomic and environmental predisposing events. Currently, there are only few reports in the literature, which is questionable in view of the worldwide prevalence of predisposing factors. So, knowledge of Hebra's prurigo is important for diagnosis and for the proper approach of cases.

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