Urticaria pigmentosa. A case report

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SUMMARY

We present the case of 22-month-old boy with Urticaria pigmentosa. Clinical feature was presented as disseminated yellow-brownish patches, on the skin of entire body, including skin of the scalp, 1x2 cm in diameter. No subjective symptoms were present. First skin lesion occurred in the second month of life. Darier's sign was positive. Serum triptase levels were elevated. Diagnosis of mastocytosis was confirmed by pathohistology.

KEY WORDS

mastocytosis, urticaria pigmentosa, mastocytes

Introduction

Urticaria pigmentosa is the prototype of mast cell disease. This name is given to a type of cutaneous mastocytosis, in which there are yellow-tan to reddish-brown patches on the skin due to abnormal collections of mast cells. (1, 2). It is the most common skin manifestation of mastocytosis in both children and adults (3).

Urticaria pigmentosa was first described by Nettleship and Tay in 1869 (4) as a rare form of urticaria. Unna in 1887 (5) demonstrated the relationship between urticaria pigmentosa and the mast cells. Mastocytosis is defined as an overproliferation and accumulation of tissue mast cells (6, 7, 8, 9, 10). In cutaneous mastocytosis, release of histamine from their granules, leads to urticarial edema of the lesions, which results in secondary hiperpigmentation due to melanocytic activity at the dermoepidermal junction (11).

Mast cells were first described by Erhlich (12, 13). Mast cells are normally widely distributed in the skin. They contain granules that contain histamine and other mediators of inflamation. When the mast cell is irritated, these chemicals are released into the surrounding skin, causing leaking of the blood vessels, resulting in localized itching, swelling and redness (2).

Urticaria pigmentosa most often affects infants. In more than 50%, the onset is before 2 years of age (14). Usually the first skin lesions appear at a few months of age. They persist and gradually increase in number for several months or years. They can appear on any part of the body including the scalp, face, trunk and limbs. There is no sex preference (2, 3, 11, 14).

Clinical feature consist of reddish or brownish patches on the skin, with dissemination of variable density. Trunk

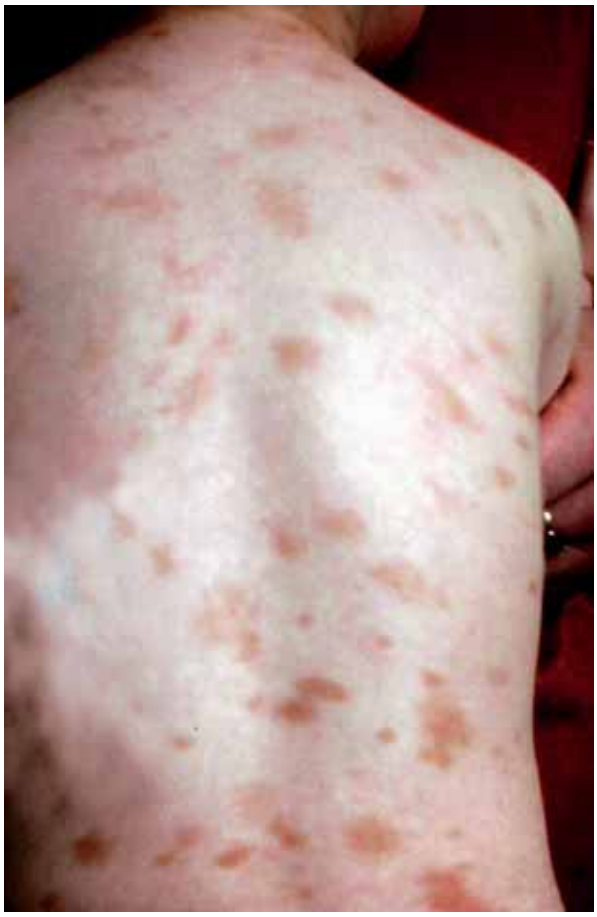

Figure 1. Disseminated reddish-brown patches on the skin of the back side of the body (with the site of the biopsy on the left)

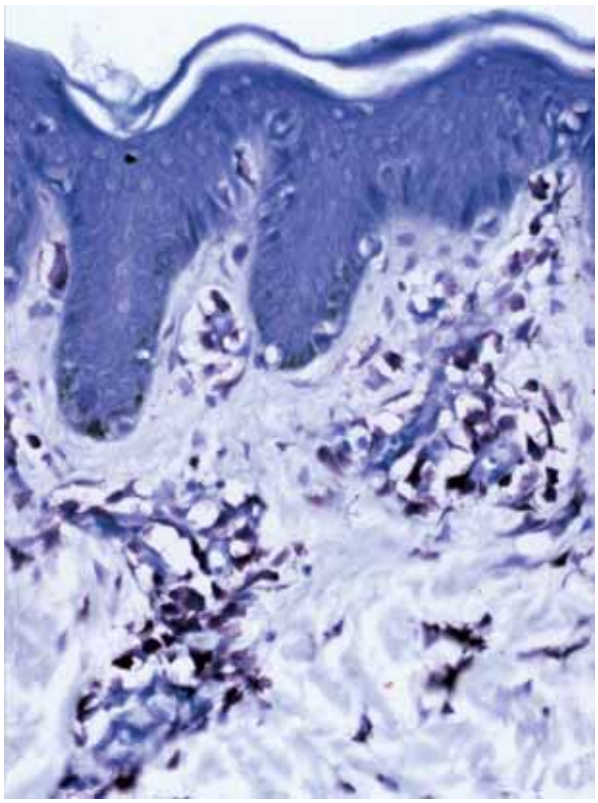

Figure 2. Pathohistology: Unna's staining revealing mast cells

is the most common site of the lesions but they can also present on the other parts of the body. Lesions are variable in size, round or oval, while nodular or tumor-like elevations are uncommon. Usually variable amount of pruritus may be present 2, 3, 11, 14).

Over the next few years the urticaria pigmentosa becomes less irritable and eventually the patches fade away. By the teenage years, most patches will resolve spontaneously (2, 3, 14).

Rubbing of a cutaneous mastocytosis lesion within a few minutes results in the formation of a wheal or even a vesicle. This characteristic response is known as Darier's sign and is considered clinically diagnostic (15). The diagnosis of urticaria pigmentosa made clinically, it should be confirmed by histopathology (3, 16).

Although in 90% of patients there are no evidence of systemic involvement (14), sometimes urticaria pigmentosa may be associated by ulcers, malabsorption, bone abnormalities, hepatomegaly, splenomegaly, lymphadenopathy, peripheral blood abnormalities, elevated levels of plasma or urinary histamine or histamine metabolites, prostaglandin D_2 and other metabolites in urine or plasma (3).

Case report

22-month-old boy was admitted to the Clinic of pediatry. On the skin of the entire body, including skin of the scalp disseminated yellow-brownish patches were present, 1x2 cm in diameter. No subjective symptoms were present.

First skin lesion occurred in the second month of life on the skin of the neck and upper extremities, as reddish infiltrates. Therapy with topical corticosteroids prescribed by the general practitioner showed no effect. Lesions have spread to the skin of the entire body including the scalp (Fig. 1). In consultation with dermatologist diagnosis of urticaria pigmentosa was made. Darier's sign was positive.

Skin biopsy of a lesion from the trunk was taken. Specimens were stained by haematoxylin-eosin (HE), and Unna's method for mast cells (17). Pathohistological examination confirmed diagnosis of mastocytosis. HE staining showed massive lymphocytic infiltrate in dermis (Fig. 2), while Unna's staining confirmed that great majority of cells in infiltrate were mastocytes.

Personal history: Second child from second pregnancy. Delivery was on term. Breast feeded 14 months. No other health problems reported by parents.

The family history relevated that the grandfather (fathers' side) allegedly had some similar skin disease in childhood.

Physical examinations including cardiology, pulmonology, neurology, ophthalmology, ECG examinations and echosonography of abdomen were without disturbances.

Laboratory analyses: Serum tryptase levels were elevated, 21 ng/mL. Normal values are up to 11.5 ng/mL (18). Other laboratory analyses (blood cell counts and biochemistry parameters) of blood and urine were within normal limits.

Patient was followed during one year. There were no clinical changes and no new lesions appeared. Since there is not involvement of other systems, except skin, it can be expected that lesions may regresse during next years.

Discussion

Cutaneous mastocytosis is a condition characterized by mast cell hyperplasia in the skin. Urticaria pigmentosa is the most frrequent manifestation of cutaneous mastocytosis. The lesions of urticaria pigmentosa appear as small yellow-tan to reddish-brown macules or slightly raised papules disseminated on the skin of the body. The palms, soles, face and scalp may be free of lesions (3). Usually the first skin lesions appear at a few months age. Mild trauma, scratching or rubbing of the lesion may led to urtication and erythema around the lesion which is known a Darier's sign. The condition is accompanied by variable degree of pruritus (3). Sytemic involvement is rare in urticaria pigmentosa (14).

In our patient, disease occured in the second month of life. Clinical feature is typical for urticaria pigmentosa, including reddish-brown macules and slightly raised plaques on the skin of the entire body. Lesions were present also on the skin of the neck and scalp which is not so usual. Darier's sign was positive.

Blood cell count was within normal values, wich is in accordance with cutaneos mastocytosis, while in systemic mastocytosis, it may reveal anemia, thrombocytopenia, thrombocytosis, leukocytosis, and eosinophilia (3, 14 19).

If a diagnosis of mastocytosis is uncertain, tests for elevated mast cell mediators and degradation products may help establish the diagnosis. Serum tryptase levels are elevated in patients with mastocytosis. Our patient also had elevated serum triptase levels. Tryptase levels may be more useful than histamine levels, because histamine can be elevated in hypereosinophilic states (19). Urinary *N*-methylhistamine (NMH) and *N*-methylimidazoleacetic acid levels may be more specific and sensitive than urinary histamine levels. NMH levels correlate directly with the extent of skin lesions. Urinary prostaglandin D_2 metabolite levels, even during asymptomatic periods, may exceed the normal level 1.5-150 times (19).

In conclusion, according to the clinical feature, positive Darier's sign, elevated serum triptase level and pathohistological examination, diagnosis of Urticaria pigmentosa was confirmed. Since this is a form of cutaneous mastocytosis, without systemical involvement, prognosis is good and it can be expected that lesion will resolve during next few years.

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