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A Case of Pediatric Urticaria Pigmentosa

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Case

A three-month-old boy was admitted to the dermatology clinic with the complaint of red-brown spots that started to appear on his body two months ago. In his dermatological examination, brown, some slightly erythematous macules were detected on the scalp, trunk, and extremities (**Figure 1a, b**). Darier's sign was positive (**Figure 1c**). In the histopathological examination of the biopsy, mild acanthosis and an increase in the basal layer melanin pigment content were observed in the epidermis, and diffuse mast cell infiltration was accompanied by eosinophils in the papillary dermis. In the immunophenotypic examination of mast cells, positive with tryptase and cluster of differentiation were detected. Blood tryptase level, hemogram, and liver function tests (LFT) were normal. Hepatosplenomegaly and lymphadenopathy were not observed. The patient was diagnosed with urticaria pigmentosa (UP).

Mastocytosis (M) describes a group of rare diseases seen in the excessive proliferation of mast cells in the skin and/or systemic organs (bone marrow, liver, spleen, lymph nodes). It is divided into two main groups systemic and cutaneous. There are three types of cutaneous M: UP (maculopapular cutaneous M), diffuse cutaneous M, and solitary mastocytoma.[1] UP is a common subtype that is seen mostly in children without systemic organ involvement. It presents numerous brownish macules and papules. Bullae can be seen over the lesions. It is commonly located on the trunk, neck, scalp, and distal extremities. These lesions usually occur within the first 6 months or may appear until adulthood. After rubbing the surface of lesions with a blunt object, an itchy urticarial plaque is observed, which is called the Darier's sign.[1,2]

Diagnosis is made by histopathological examination of typical skin lesions. Intense mast cell infiltration in the dermis and increased melanin in the basal layer are the most important findings.[2] Serum tryptase level, hemogram, and LFT should be normal, lymphadenomegaly and hepatosplenomegaly should not be detected for diagnosis of cutaneous M.[1] Systemic involvement is observed in very few children. It is recommended to repeat them every 10-12 months. Bone marrow biopsy is not routinely recommended in pediatric patients.[2]

Releasing of mast cell mediators causes pruritus, urticaria, flushing, abdominal pain, bone pain, diarrhea, and cardiovascular symptoms. Rarely, anaphylaxis and death have been reported. Exercise, exposure to hot and cold, emotional stress, local trauma to lesions, consumption of alcohol, spicy food, and some drugs (narcotics, salicylates, nonsteroidal anti-inflammatory drugs, vancomycin, polymyxin B, contrast agents, etc.) may trigger systemic symptoms of M.[1,2]

The differential diagnosis includes urticaria, nodular scabies, xanthogranuloma, pseudolymphoma, and spitz nevus.[1-3] There is no curative treatment for UP. Skin lesions usually regress spontaneously before puberty. Eliminate triggering factors and symptomatic treatments are recommended.

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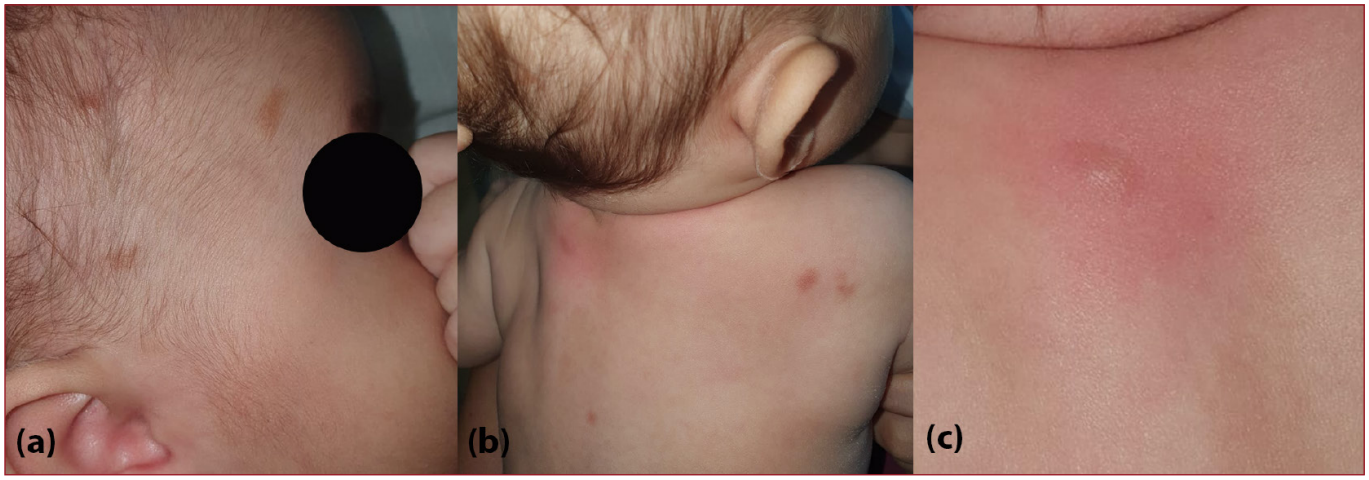

Figure 1. (a) Brown macules and papules located on the head, (b) Erythematous brown macules located on the back, (c) Darier's sign positive lesion on the back

Antihistamines, oral cromolyn sodium, steroids, and topical calcineurin inhibitors are usually used.[1]

Pediatricians see this type of rash frequently. Patients are referred to dermatology outpatient clinics in the late period or with false prediagnosis. Including M in the differential diagnosis of brown maculopapular lesions is vital for patients with this diagnosis.

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