Anaphylaxis Accompanying Mastocytoma in Childhood: A Case Report

Mahsa Rekabi1, Vahab Rekabi2, Seyedeh Atefeh Hashemi Moghaddam1, Maryam Heydarazad Zadeh1 and Sepideh Darougar3*

1Pediatric Respiratory Diseases Research Center, National Research Institute of Tuberculosis and Lung Diseases (NRITLD), Shahid Beheshti University of Medical Sciences, Tehran, Iran
2Fellowship of Molecular Pathology and Cytogenetics, Tehran, Iran
3Department of Pediatrics, Faculty of Medicine, Tehran Medical Sciences, Islamic Azad University, Tehran, Iran

*Corresponding author(s)
Sepideh Darougar, Amir-al-Momenin Hospital, Shirmohammadi Street, Nazi-Abad Avenue, Tehran, Iran
Tel: +98-912-288-1975
Fax: +98-215-534-6301
Email: sepidehdarougar@yahoo.com
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Keywords
- Mastocytoma
- Anaphylaxis
- Wheat allergy
- Cow’s milk allergy
- Mast cell

Abstract

Background: Mastocytosis is a heterogenous disorder characterized by accumulation of mast cells in different tissues. Mastocytosis may be limited to skin or it may involve bone marrow, liver, spleen and lymphatic tissue as systemic mastocytosis. Patients may also present with anaphylaxis. Here we report a six-year-old boy with atopic dermatitis, wheat anaphylaxis, and solitary mastocytoma.

Case report: The dermatologic manifestations of atopic dermatitis appeared early in his first days of life and were all over his face with a dark red infiltrate, edema, small vesicles and severe itching. At the age of 3 months, the patient was fed with an ordinary milk formula for the first time, which immediately resulted in severe vomiting, generalized urticaria and angioedema of the lips and eyes, followed by lethargy and atony. The result of RAST test indicated allergy to wheat and cow’s milk. At the age of two years, his mother noticed a rubber-like skin lesion on his left arm with positive Darier sign. Laboratory tests were all within normal limits except IgE and serum tryptase which were 1425 IU/mL and 31 ng/ml, respectively.

Conclusion: To conclude, Anaphylaxis may accompany cutaneous forms of mastocytosis even mastocytoma. Delayed diagnosis due to the lack of awareness may lead to recurrent life-threatening events, particularly in infancy when anaphylactic reactions are easily misdiagnosed as infants are not capable of expressing their symptoms.

Introduction

Mastocytosis is a heterogenous disorder characterized by accumulation of mast cells in different tissues [1]. Mastocytosis may be limited to skin or it may involve bone marrow, liver, spleen and lymphatic tissue as systemic mastocytosis. Patients may also present with anaphylaxis [2]. The skin is the most frequently affected organ [3]. There are three major types of cutaneous mastocytosis namely a) diffuse cutaneous mastocytosis, b) urticaria pigmentosa, and c) solitary mastocytoma. Pediatric mastocytosis is often confined to the skin [4] with polymorphic maculopapular lesions located on the trunk, extremities, scalp or neck as the most frequent presentation [5]. The etiology of the disease is attributed to a mutation involving KIT D816V. A slight predominance in boys has been noted in
children in different studies [6]. Here we report a child with atopic dermatitis, wheat anaphylaxis, and solitary mastocytoma.

**Case Presentation**

The patient is a six-year old boy, born at 36 weeks of gestation, weighing 2970 grams. He was delivered by cesarean section. The dermatologic manifestations of atopic dermatitis appeared early in his first days of life and were all over his face with a dark red infiltrate, edema, small vesicles and severe itching, which were refractory first but later they were self-subsided. At the age of 3 months, the patient was fed with an ordinary milk formula for the first time, which immediately resulted in severe vomiting, generalized urticaria and angioedema of the lips and eyes, followed by lethargy and atony. A Radioallergosorbent test (RAST) test was performed then, the result of which indicated allergy to wheat and cow’s milk. He was therefore referred to an allergist, followed by a six-food elimination diet for his mother as he was being breast-fed by her. The skin lesions improved significantly after strict food avoidance, which was also started for the baby himself when he reached the age of six months, as the conventional time for receiving supplemental diet. Despite the relative improvement of the skin lesions, he did not show favorable weight gain. Neocate formula was initiated for him at the age of 12 months and continued until he became 20 months old. Because of poor weight gain and chronic constipation, he was switched to Elecare formula. At the age of two years, his mother noticed a rubber-like skin lesion on his left arm, which was waxing and waning over time (Figure 1).

Clinical examination revealed a solitary 2×2 cm, non-tender elevated lesion on his left arm. Upon scratching the lesion with a tongue blade, the skin became edematous and itchy, which was indicative of positive Darier sign. The patient was referred to a couple of dermatologists for skin biopsy in order to confirm the diagnosis. However, no one agreed to biopsy the lesion as the dermatologists believed that the diagnosis was clinical and straightforward due to the appearance of the lesion and positive Darier sign. At the age of two years, laboratory tests were all within normal limits except IgE and serum tryptase, which were 1425 IU/mL and 31 ng/ml, respectively.

The clinical symptoms of poor weight gain and constipation as well as mucoid secretions in stool despite strict food avoidance persisted until preschool years. During these years, his family realized that any consumption of the wheat, even a little piece of bread, or skin contact with wheat as well as inhalation of the wheat flour could lead to a severe anaphylactic reaction. During these attacks, the child experienced generalized urticaria, angioedema of the lips and eyes, decreased blood pressure, cough, wheezing, shortness of breath, nausea and vomiting. Because of the life-threatening nature of the attacks, the patient underwent oral wheat immunotherapy. After six months of oral immunotherapy, he was allowed to have pasta, which was well tolerated. At the end of the procedure, the child could tolerate bread as well. After wheat oral immunotherapy, he underwent milk immunotherapy. Both of these procedures were finished successfully and the child could tolerate wheat and milk. Meanwhile, the skin lesion on the left arm, diagnosed as mastocytoma, resolved spontaneously at the age of 4 years.

**Discussion**

This report is an illustration of a child with a couple of allergies and mastocytoma. Wheat sensitization with recurrent anaphylaxis and chronic relapsing skin disorder such as atopic dermatitis may be associated with mast cell disorders. It has been shown that mast cells as well as basophils are the primary effector cells in IgE-mediated food allergy. However, the evaluation of tissue mast cell population is difficult.

In this child, anaphylactic reaction after wheat consumption was the life-threatening event, which made the parents seek medical interventions in order to keep the child’s allergy under control. Immunotherapies with the theoretical potential to
induce immunological transition from desensitization to allergy regulation may result in functionally beneficial alterations.

Pediatric mastocytosis is often confused with more common rashes such as atopic dermatitis, urticaria and angioedema, maculo-papular rashes and even bullous exanthema [7]. Solitary mastocytoma accounts for 10–15% of cutaneous mastocytosis and is the second most common type of cutaneous mastocytosis, with the majority of the lesions present during the first year of life [8]. In most children, skin lesions regress spontaneously around puberty [9].

Anaphylaxis is an uncommon manifestation that is generally seen in systemic mastocytosis, yet it may still occur in the cutaneous forms [10]. Higher number of hyperactive mast cells in mastocytosis may predispose the patients to anaphylaxis [11]. Several factors may trigger the release of mast cell mediators in mastocytosis including insect stings, foods, drugs, alcohol, mechanical irritation, emotional or physical stress, sudden temperature changes, and infections. In addition to above-mentioned factors, other potential triggers in children are fever and febrile responses to vaccination and teething [12]. Specific trigger factors for anaphylaxis in children with mastocytosis remain unidentified but extensive blistering skin disease infiltrated with mast cells is suggested as the strongest triggering factor for anaphylaxis in these children [13]. However, the evaluation of tissue mast cell population is difficult.

Wheat is a widely consumed grain currently used by humans worldwide [14], which may cause food allergy by an IgE-mediated reaction [15]. Wheat was the triggering factor leading to anaphylactic reactions after every consumption by either the mother or the child. In 2010, Kunnath, et al. [16] reported a 9-month old male who was treated for atopic dermatitis due to cow’s milk intolerance as he had shown intensely pruritic skin rash from the age of six weeks. He did not tolerate Neocate formula either. A skin biopsy later confirmed the diagnosis of cutaneous mastocytosis. There was a striking similarity between the above-mentioned case with our patient. Retrospectively, we consider this non-pigmented itchy lesions, diagnosed as atopic dermatitis in the first weeks of life, to be cutaneous mastocytosis, which later developed as solitary mastocytoma. If our patient was evaluated with a skin biopsy of the lesions, which was initially presumed atopic dermatitis, the condition could have been diagnosed much earlier. However, atopic dermatitis is a clinical diagnosis with its own characteristic criteria, so skin biopsy is not required in the majority of cases.

**Conclusion**

Anaphylaxis may accompany cutaneous forms of mastocytosis even mastocytoma. Since mast cell disorders should be considered in differential diagnosis of these patients with recurrent anaphylaxis and skin lesions, oral immunotherapy may impose significant risk to the patients with food allergy and different types of mastocytosis. Therefore, performing such procedures demands substantial caution. Delayed diagnosis due to the lack of awareness may lead to recurrent life-threatening events, particularly in infancy when anaphylactic reactions are easily misdiagnosed as infants are not capable of expressing their symptoms.

**Declarations**

**Ethics approval and consent**

This case study has been approved by the ethics committee of National Research Institute of Tuberculosis and Lung Diseases.

**Consent for publication**

Written informed consent was obtained from the patient’s legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

**Competing interests**

The authors declare that they have no competing interests.

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