

An unusual case of neonatal mastocytoma: A case report

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CASE REPORT

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ABSTRACT

Mastocytosis is a group of disorders characterized by the accumulation of mast cells in various tissues. In this article we report an interesting case of cutaneous mastocytoma in a 3-month-old infant. We describe two episodes characterized by an exacerbation of the skin lesion with the typical Darier's sign. Both of them occurred after breastfeeding. In both episodes, the mother had eaten shrimps before the breastfeeding. This peculiar case, difficult to understand regarding its pathogenesis, was successfully resolved just with dietary prescription given to the breastfeeding mother.

Key Words

Mastocytoma, breastfeeding, shrimp, allergen, histamine

Implications for Practice:

1. What is known about this subject?

Mastocytosis is a group of disorders characterized by the

accumulation of mast cells in various tissues.

2. What new information is offered in this report?

Here we report a case of mastocytoma in a 3-month-old patient. We found this case really interesting and original. Actually, we did not find in literature similar cases, in which the described clinical reaction was determined from the mother to the baby, by breastfeeding.

3. What are the implications for research, policy, or practice?

We describe two episodes characterized by an exacerbation of the skin lesion with the typical Darier's sign. Both of them occurred after breastfeeding. In both episodes, the mother had eaten shrimps before the breastfeeding. Here we explain the possible pathogenesis of this peculiar case.

Background

Mastocytosis is a group of disorders characterized by hyperplasia of mast cells and by symptoms caused by the release of mediators, such as histamine and tryptase, following the degranulation of mast cells. Degranulation and release of mediators can be induced by a physical stimulation or specific triggering factors.

The most frequent site of organ involvement is the skin. Less commonly, we can find the accumulation of mast cells in other parts of the body, such as lymph nodes, gastrointestinal tract or bones.¹

The typical cutaneous lesion of mastocytosis was described for the first time by Nettleship et al. in 1869 as an atypical form of urticaria.²

In 2016 a consensus report of the European Competence Network on Mastocytosis, the American Academy of Allergy, Asthma & Immunology, and the European Academy of Allergology and Clinical Immunology revised the classification and the diagnostic criteria of cutaneous

manifestations in patients with mastocytosis.³ The Authors divided cutaneous mastocytosis into two large groups: adulthood-onset and childhood-onset mastocytosis. Each of the two groups were subdivided in 3 subgroups: MPCM (maculopapular cutaneous mastocytosis) previously called urticaria pigmentosa, DCM (diffuse cutaneous mastocytosis) and cutaneous mastocytoma, previously named solitary mastocytoma. However in 2015 Méni et al reviewing 1747 cases of paediatric mastocytosis published in International Literature between 1950 and 2014 divided their cases according to the clinical presentation: urticaria pigmentosa or UP, mastocytoma, diffuse cutaneous mastocytosis or DCM and telangiectasia macularis eruptiva perstans or TMEP.⁴ Regarding the prevalence UP was present in 74.8 per cent of the cases, mastocytoma in 19.5 per cent, DCM in 5.2 per cent and TMEP in 0.3 per cent.

Typically, cutaneous mastocytosis is characterized by positive Darier's sign, which consists in the formation of a wheal after scratching the skin lesions. Darier's sign was present in more than 90 per cent of cases presented by Méni et al. In 34.5 per cent of patients bullous lesions were described.

Mastocytoma can be present at birth or appear during the first months of life. It is generally a solitary lesion, even if it is possible to find multiple forms, and it is characterized by a macular, plaque or nodular aspect. The most common localization of mastocytoma in children is in the extremities, with the exception of the palmoplantar region. Serum tryptase levels are generally normal and no systemic involvement is found. Mastocytomas usually do not persist into adulthood. (3) Here we report a case of mastocytoma in a 3-month-old-patient with a particular cutaneous reaction after mother breast feeding.

Case details

We present the case of a 3-month-old male infant that referred to our Department for the presence of an erythematous nodular lesion on his left leg. At first, the cutaneous lesion was considered to be a skin reaction located in the same site where the birth bracelet was placed. However, the lesion persisted and sometimes presented erythematous reaction. Particularly in one occasion a blister formed on the skin lesion while a flushing localized on face and neck was observed (Figure 1A and 1B). The mother referred that this episode appeared during breast feeding, after she had eaten a meal with many shrimps. This reaction resolved after topical administration of a corticosteroid cream and after this episode the lesion appeared as a no inflammatory reddish- brown nodule,

sized 4mm, on his left leg. The child did not have any other physical or psychomotor disturbances. No significant family or personal history data were reported. When we stroked the lesion, it became red, more elevated and a blister developed (positive Darier sign). Considering clinical data, a diagnosis of cutaneous mastocytoma was made.

Routine laboratory examination was in normal range. Serum baseline tryptase performed was normal.

A dietary prescription was given to the mother but after 3 months she ate again shrimps and second episode appeared; however, in this case there was only exacerbation of the skin lesion with bullous formation, without clinical manifestation on his face (Figure 1C).

Currently, the patient is 11-month-old, he did not suffered for similar episodes while the mother finally excluded shrimps from her diet.

Discussion

The diagnosis of mastocytoma in our patient has been made clinically. We did not perform a cutaneous biopsy with histological examination because of the young age of our patient and considering the fact that the clinical aspect was very typical for mastocytosis, with a positive Darier sign. Furthermore, the parents of the patient were against the performance of a histological examination. However regarding the diagnosis in the literature the major criterion is considered the clinical one with typical skin lesions of mastocytosis associated with Darier's sign³ as observed in our patient. Minor criteria include increased numbers of mast cells in biopsy sections of lesional skin and KIT mutation in lesional skin tissue.

In our opinion, this case is really interesting because we observed two clinical Darier episodes in our patient, with an erythematous and raised blister, following the ingestion of shrimps by the mother. Curiously, the cutaneous reaction in the baby has been provoked by breastfeeding, probably because of the presence of shrimp molecular particles in the breast milk. On the contrary the mother did not have any clinical cutaneous reaction due to the shrimp intake.

As we described before, the first episode was characterized by an exacerbation of the primitive lesion and a flushing circumscribed on the face and neck. On the other hand, in the second episode we observed only blistering on mastocytoma, without skin manifestation in other sites.

In cutaneous mastocytosis, the onset of symptoms resulting from the activation and release of mediators from the mast cells are observed in 25 per cent of paediatric patients.⁴ The most frequent triggers for the release of mediators and also of anaphylaxis are food, insect stings and drugs.

In mastocytosis, many different kinds of food were reported as a possible cause of allergic reactions, such as cow's milk, peanut, peach, egg and shellfish including shrimps.⁵

Food allergy is an allergen-specific IgE mediated type I response and, specifically, in case of shrimps, the allergens responsible for this hypersensitivity reaction have been identified as tropomyosin, arginine kinase, sarcoplasmic calcium-binding protein, myosin light chain, troponin C and triosephosphate isomerase. Among these, the most involved shrimp allergen is the muscle protein tropomyosin.^{6,7} Shrimp allergen can induce mast cell degranulation through an IgE related mechanism as suggested in some studies on mice⁸ even if a no IgE mediated mechanism may not be excluded.

It has been suggested that diets restricted in biogenic amines or histamine-releasing foods may be beneficial for some patients with systemic mastocytosis.⁵ However, many authorities do not mention diet intervention as a common therapeutic measure.⁵ In 2005 Vlieg-Boerstra et al., using Medline (1966–2004), Cinahl (1982–2004) and the Cochrane Library searched for double-blind placebo-controlled food challenge (DBPCFC) studies with biogenic amines and/or histamine-releasing foods in mastocytosis but no report was found. Our case report may be considered like a DBPCFC in vivo study regarding shrimps and newborn.

During the first episode the patient showed a bullous reaction on cutaneous mastocytoma with flushing localized on his face and neck. In our opinion, the circumscribed localization of flushing on his face could be due to a more intense vascularization and to a larger presence of vascular receptors in this area.

During the second episode, again a bullous reaction was registered on cutaneous mastocytoma as an in vivo challenge of absorption of shrimps and skin reaction. However the absence of flushing on the face could be explained by the improvement of cutaneous mastocytoma clinically observed and commonly reported in the literature.^{3,4} Specifically, this clinical improvement could be characterized by a quantitative decrease of mast cells and consequently by a reduced release of histamine. Therefore,

a reduced release of mediators would determine only a local exacerbation without other distant manifestations.

Finally regarding allergic reactions in newborn and breast feeding at our knowledge no similar case has been reported in the Literature. Commonly mother breast feeding is regarded as a protective action against the development of allergy in newborn while proteomic studies of mother milk proteins are rarely reported.⁹

Conclusion

Cutaneous mastocytoma is a benign tumour, usually characterized by a spontaneous regression during the years. Generally, no surgical excision neither histological examination are needed, unless in case of diagnostic doubt.

The most important problem of mastocytoma is the possibility of intense vasoactive reactions or anaphylaxis in case of massive release of histamine induced by triggers, such as food or drugs. To prevent this from happening, it is necessary to avoid the exposition to the known triggers as we observed in our case where no relapse was found after dietary prescription to the breast feeding mother.

In conclusion, we did not find in Literature similar cases, in which the described clinical reaction in the baby was determined by mother shrimps intake and subsequently breastfeeding.

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PEER REVIEW

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CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

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PATIENT CONSENT

The authors, *Pranteda G, Magri F, Pigliacelli F, Muscianese M, D'Arino A, Pranteda G, Di Silvio G, Bottoni U*, declare that:

1. They have obtained written, informed consent for the publication of the details relating to the patient(s) in this report.
2. All possible steps have been taken to safeguard the identity of the patient(s).
3. This submission is compliant with the requirements of local research ethics committees.

Figure 1: (A and B) This episode exacerbated the skin lesion and a flushing localized on face and neck was observed



Figure 1C: Exacerbation of the skin lesion with bullous formation



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