A Case of Urticaria Pigmentosa

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Abstract

We present a case of the most common form of cutaneous mastocytosis in children, Urticaria Pigmentosa (UP).

Introduction

Mastocytosis is a group of rare disorders involving excess proliferation and accumulation of mast cells. It is divided into two entities: cutaneous mastocytosis involving the skin, and systemic mastocytosis, which affects multiple organs (1). We present a case of the most common form of cutaneous mastocytosis in children, Urticaria Pigmentosa.

Case Presentation

A 3-month-old male presented to dermatology clinic for a rash that started a few weeks after birth. Mom stated the brown spots appeared on the arms initially, gradually involving the abdomen, chest, back, legs, and face. ID ruled out rash secondary to congenital infection with negative serology for Rubella, CMV, RPR. On review of systems, the patient had intermittent episodes of constipation, which his pediatrician attributed to an umbilical hernia. No diarrhea, wheezing, or signs of respiratory distress. The patient was reaching developmental milestones and growing appropriately per the pediatrician. On physical examination, the patient appeared comfortable. There were multiple dark brown-reddish macules and patches on the face, arms, legs, chest, abdomen, and back (Figure 1). Darier sign was positive. 2 mm punch biopsy revealed a proliferation of mast cells with +CD117 marker, consistent with Urticaria Pigmentosa. On follow up, we learned that the patient was scheduled to have surgery for umbilical hernia repair.

Discussion

UP is the most common form of cutaneous mastocytosis in childhood. It presents in the first few weeks of life as 5 to 15 mm pink to brown colored, urticular, macules, papules, vesicles, or nodules. Lesions are more commonly found on the trunk and generally spare the central face, palms, and soles. The lesions exhibit the classic Darier’s sign, which is urtication upon local rubbing of a lesion. Most children with UP will have a limited course with spontaneous resolution by adolescence. However, 10-15% of cases may persist into adulthood (1). Diagnosis of UP can be confirmed with biopsy. On histology, there is a dense dermal collection of uniformly spaced mast cells. Eosinophils are also commonly found within the dermis (Figure 2). Staining with Leder stain will show red colored mast cells, while with the Giemsa stain mast cell granules will stain a metachromatic purple color. Toluidine blue, tryptase and CD117 stains (c-kit tyrosine kinase) may also be used (2) (Figure 3). Further workup may be considered if the patient exhibits systemic symptoms (2). Treatment is largely focused on symptom relief and prevention of mast-cell degranulation by avoiding triggers. Exacerbating factors include exercise, heat, friction of skin, and ingestion of hot spicy foods. Systemic agents that can cause mast-cell degranulation include alcohol, narcotics, NSAIDs, polymyxin B, and anticholinergic medications. There are several systemic anesthetic drugs that can actually precipitate an anaphylactic reaction in mastocytosis. Lidocaine, however, is safe when used as a local anesthetic. Safe systemic anesthetics include fentanyl, sufentanil, remifentanil, midazolam, propofol, ketamine, desflurane, sevoflurane, cisatracurium, pancuronium and vecuronium bromide. Antihistamines, both first and second generation, are a mainstay of treatment. Topical, intraleosional and oral steroids can be used depending on disease severity. Psoralen with UVA (PUVA) along with other light therapies have shown benefits as well (3,4).

Conclusion

We present a case of Urticaria Pigmentosa in a patient who was scheduled for a surgical procedure requiring systemic anesthetics. It was imperative to advise and counsel our patient about the risks associated with anesthetics i.e, an anaphylactic reaction (4). Thus, this case exemplifies the importance of counseling patients with mastocytosis about medication and lifestyle-related triggers to prevent adverse reactions.

References

5. James, William D., MD; Berger, Timothy G., MD; Elston, Dirk M., MD. Andrews' Diseases of the Skin, Published January 1, 2016. © 2016.