Solitary Mastocytoma: An Inusual Presentation in Children

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Abstract

Mastocytosis is abnormal proliferation of tissue mast cells, solitary mastocytoma being the most common. Evolution tends to spontaneous healing, constituting a small percentage of solitary mastocytoma refer surgical removal.

Keywords: Solitary mastocytoma; Expectant management; Treatment resolute

Introduction

Mastocytosis [1-4] is a group of diseases characterized by abnormal proliferation of mast cells in different tissues, which may be located in a localized skin as Solitary Mastocytoma (MS); or diffuse and diffuse cutaneous mastocytosis, telangiectasia eruptive macularis perstans and urticarial pigmentosum; and or systemic injury. The most common forms of urticarial pigmentosum mastocytosis are followed by solitary mastocytoma. The aim of this paper is to present a case of solitary mastocytoma and resolve the doubts raised by when should perform surgical resection? When? Maintain an expectant attitude when investigating systemic involvement?

Case Presentation

Child three-month old who presents erythema and edema on the right side of the face, arm and hand it presents injury back of his right hand since birth that has intensified coinciding with widespread redness. It is an erythematous yellowish-brownish hue of 25 x18 mm in diameter with a central indurated 10 x 15 mm (Figure 1). He was quoted in consultation dermatology for evaluation of possible mastocytoma, and there the diagnosis was confirmed, recommending conservative treatment.

Discussion

Solitary mastocytoma [1-8] represents approximately 10% of cases of cutaneous mastocytosis. It is described as one or more oval lesions with a smooth or “orange peel” surface, usually located on limbs or trunk. In the differential diagnosis of mastocytosis, Darier sign (Figure 2) is essential, which is pathognomonic positive in 90% of cases, producing one more or less intense urticarial reaction, with rubbing surface that is essential.

What age range we can find the MS and what your evolution?

The MS appears in two peaks of age, the first before 6 months of age and the second in adulthood. Normally the MS in childhood resolves spontaneously during the first years of life [1-7].

When investigating systemic involvement?

10% of patients with mastocytosis [1,2] has a different location on the skin, not the involvement of it being necessary to find an abnormal proliferation of mast cells in bone lesions, hepatosplenomegaly, gastrointestinal tract, hematopoietic system and lymph nodes among other organs and systems. This is systemic mastocytosis. If we find skin lesions it is not indicated further testing to find mastocytosis in other locations, unless there are systemic manifestations.

When should perform surgical resection? When maintaining a waiting attitude?

Being an auto resolutive process the best recommendations is a conservative and symptomatic treatment, using H1 and H2 antihistamines to reduce itching, blistering and symptoms in general. Corticosteroid therapy is used in over 2 years. We can also use mast cell stabilizers [5]. There are triggers of mast cell degranulation, resulting in urticaria reaction and may occur even anaphylaxis and shock. Some of these factors are sudden changes in temperature, trauma or friction, exercise, heat, stress, certain foods such as chocolate or nuts, drugs like aspirin and NSAIDs. In a small
percentage of patients the plate has an activity significantly higher than what requires surgical removal of the same [6].

References