

PAEDIATRIC DERMATOLOGY

## **PEDIATRIC INDOLENT SYSTEMIC MASTOCYTOSIS: ROLE OF THE UVB-NB PHOTOTHERAPY IN THE TREATMENT OF MACULO-PAPULAR CUTANEOUS LESIONS.**

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**Background:** Mastocytosis is a rare disease characterized by an abnormal accumulation of mast cells in single or multiple organs with different clinical presentations and grades of severity based on systemic involvement. The clinical presentation is widely heterogeneous, ranging from skin-limited disease cutaneous mastocytosis (CM) to more aggressive systemic variants with extracutaneous involvement (SM). Although nearly two-thirds of patients with mastocytosis are children and the initial lesions usually appear in the first 6 months of life, the observation of systemic involvement in children is rare.

**Observation:** We report a 13 y.o. girl, affected by indolent systemic mastocytosis (ISM) with maculo-papular cutaneous involvement that had been present since the sixth month of life. Dermatological examination revealed a pruritic light-brown patches and multiple brown-to-red macules and papules with Darier's sign positivity. Laboratory tests of complete blood cell count, liver and kidney function were in the normal range. Highly elevated levels of serum tryptase (>20 ng/mL) were evident. A diagnosis of ISM was suspected and verified by the presence of multifocal, dense aggregates of MCs in the skin, bone marrow and confirmed by expression of CD2, CD25, and CD117 in cells. Given the rarity of the juvenile-onset ISM there are no established protocols for the treatment, but the therapeutic approaches recommended are similar to those applied to adults. The patient was intermittently treated with topical corticosteroids and anti-histamines with poor results. The use of phototherapy UVB-NB has allowed the achievement of clinical remission and improvement of pruritus.

**Key message:** The pathogenetic complexity and biological heterogeneity of mastocytosis variants make the therapy of this diseases difficult. Even if treatment options and outcomes in young patients are not well defined, this study provides evidence that UVB-NB may be used successfully as a safe and useful second-line therapy treatment for ISM in young patients.