

17th WORLD DERMATOLOGY CONGRESS

September 25-26, 2017 Dubai, UAE

Case study on mastocytosis

Noof Al 'Abdulsalam

Al Nahdha Hospital, Sohar Polyclinic, Oman

45yrs old Omani gentleman presented to OPD for the first time with history of generalized dusky erythematous maculopapular lesions of 9 months duration. Skin biopsy done and showed cutaneous mastocytosis then progressed to systemic mastocytosis.

Skin:

- Dusky erythematous maculopapular lesions scattered all over the face, trunk, & both of the extremities except the palms & soles.
- There were no lesions in the buccal mucosa.
- "Darier's sign" was positive.

Systemic Review: Unremarkable

Investigations: CBC with differential, UE, LFT, GLU were all normal Histopathology (skin biopsy): Localized dense mast cells infiltrate in the papillary & upper reticular dermis more marked in the papillary dermis. Appearance consistent with cutaneous mastocytosis with features in keeping with urticaria pigmentosa.

Diagnosis: Urticaria Pigmentosa

Results: Mastocytosis is a disorder of increased mast cell proliferation with both cutaneous and systemic manifestations. Mastocytosis occurs in both children and adults, although children tend to primarily manifest cutaneous lesions. Most cases appear to be spontaneous. Inherited patterns of mastocytosis are unusual. Mastocytosis has been classified into disease variants according to a WHO consensus;

- Cutaneous mastocytosis (CM)
- Urticaria pigmentosa (UP) = maculopapular CM (MPCM)
- Diffuse cutaneous mastocytosis (DCM)
- Mastocytoma of skin
- Indolent systemic mastocytosis (ISM)
- Smoldering SM
- Isolated bone marrow mastocytosis
- Systemic mastocytosis with associated clonal, hematologic non-mast-cell lineage disease (SM-AHNMD)
- Aggressive systemic mastocytosis (ASM)
- Mast cell leukemia (MCL)
- Mast cell sarcoma (MCS)
- Extracutaneous mastocytoma

Children with cutaneous mastocytosis (CM) generally have resolution of symptoms by adulthood. In adults, cutaneous disease is usually accompanied by evidence of mastocytosis in other organ systems.

Biography

Noof Al 'Abdulsalam is recently working under Al Nahdha Hospital, Sohar Polyclinic, Oman. His research experience includes various programs, contributions and participation at different countries for diverse fields of study.

Doctor553@hotmail.com