Net Quiz

# A brownish-red plaque in an adult

A 50-year-old man was referred to our clinic for further evaluation of a slowly enlarging plaque on his back. His wife gave history of the pimple-sized brownish lesion first being noticed thirty years earlier. Ever since, the lesion has been presenting slowly and continuously enlarging. On examination, the patient, a healthy looking man, had a brownish-red, shiny, well-demarcated, 6.0 mm elevated, rubbery consistency, a 7.8 x 8.2 cm dimension, hairless plaque with peau d'orange surface and 'pasted on' appearance on the left side of his back. The margins of the lesion were surrounded by a brownish zone. The patient did not have any complaint except a slight tingle in the lesion after stroking, pressure, or friction. Inflammatory flare characterized with redness, swelling, and pseudovesiculation was observed in the lesion after stroking [Figure 1A]. The inflammation and pseudovesiculation gradually decreased within a few days. Then, desquamative scales developed over the lesion [Figure 1B].

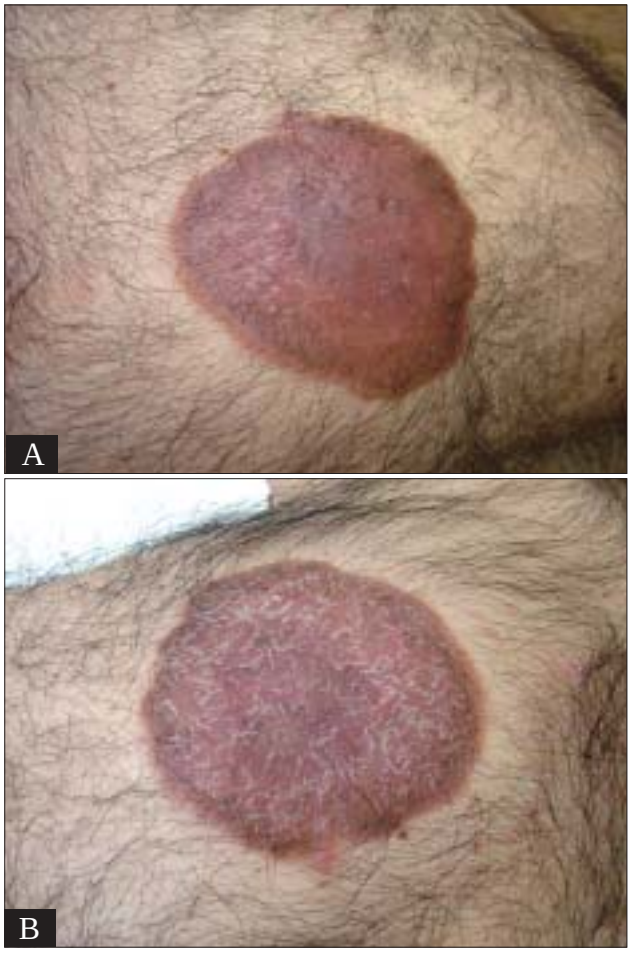


**Figure 1: (A) Indurated large plaque after stroking. Note the edematous erythema and peau d'orange surface due to pseudovesiculation in the lesion, (B) Appearance of the plaque after one month on stroking. Note desquamative scales developing over the lesion**

His systemic examination was unremarkable, and routine laboratory tests were within normal limits. There was no family history of similar skin lesion.

A skin biopsy specimen was obtained [Figure 2].

## WHAT IS THE DIAGNOSIS?

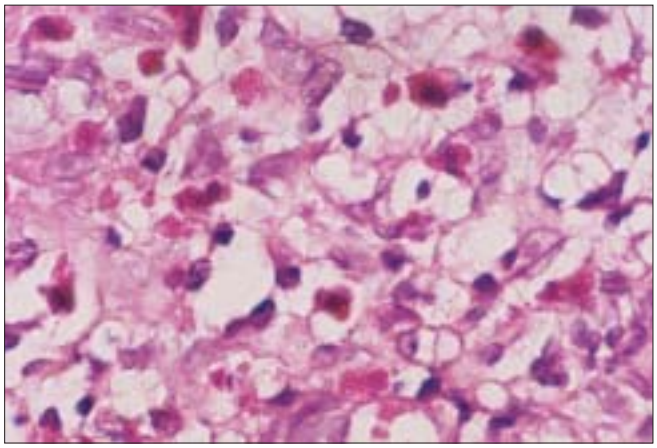

**Figure 2: Note the mast cells containing large granules in their cytoplasm (H&E, x1000)**

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Diagnosis: Solitary mastocytoma

Histopathologic examination of the biopsy specimen stained with hematoxylin and eosin showed a dense mixed cellular infiltrate composed of lots of mast cells that contained giant granules in their cytoplasm within the dermis. Based on histological findings and clinical data, we finally diagnosed this case as solitary mastocytoma.

## DISCUSSION

Mastocytosis is a group of disorders characterized by mast cell proliferation and accumulation within various organs, most commonly the skin.[1] Solitary mastocytoma with onset in adulthood is very rare and only a few cases have been reported in the world literature.[2]

The term mastocytoma has been used to describe nodular infiltrates of mast cells occurring as a single or several isolated lesions.[3] Clinically, it is often presented as a single or several, red to reddish-brown minimally infiltrated nodule(s) or plaque(s). Peripheral hyperpigmentation is more prominent. Increased local concentrations of soluble mast cell growth factors in lesions of cutaneous mastocytosis are believed to stimulate mast cell proliferation, melanocyte proliferation, and melanin pigment production. The induction of melanocytes explains the hyperpigmentation that commonly is associated with cutaneous mast cell lesions.[4]

When a lesion of mastocytosis is stroked, it generally urticates, becoming pruritic, edematous, and erythematous. This change is referred to as the Darier sign, which is explainable based on mast cell degranulation induced by physical stimulation. In this case, inflammation and pseudovesiculation occurred within a few hours after the lesion had been stroked. In the following days, the inflammation and pseudovesiculation gradually decreased, and the surface of the lesion covered with desquamative scales.

The pathogenesis of mastocytosis is largely unclear. Activating mutations of KIT, the receptor of the mast cell growth factor stem cell factor, appear to play a key role in the pathogenesis of sporadic adult onset mastocytosis.[5]

Solitary mastocytoma is usually seen in infancy or childhood. It commonly disappears within a few years, generally before puberty. The lesion, however, may uncommonly persist into adult life and progress to a thick and large plaque. When they are firmly stroked or vigorously rubbed, inflammatory flare and edema develop, but during resolution, desquamative scales may appear over the surface of thick solitary mastocytoma.

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