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Multicentric Reticulohistiocytosis Resembling Dermatomyositis: A Report of a Case and Literature Review

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Abstract:

We describe a 36 years old Chinese lady who presented with multiple painless non- pruritc papules and nodules on the hands, elbows, ear, face and feet associated with joint pains involving the knees and the small joints of the hands. She also had photo- distributed rash on the face, chest and back. Her systemic examination was normal. Other than the articular erosions in the interphalangeal joints and erosion on the patella bilaterally, all other investigations were normal or negative. The photo distributed patches and plaque lesion on the front and back of the chest with facial rash and papules on the knuckles of the hands may be mistaken for dermatomyositis.

The histopathological examination revealed dermal infiltrates of multinucleated histiocytic giant cells with an eosinophilic 'ground glass' cytoplasm which confirmed the diagnosis of multicentric reticulohitiocytosis in our case.

Introduction:

Multicentric reticulohistiocytosis (MR) is a rare disorder of skin with systemic granulomatous involvement of no known cause with distinct histopathology. It has been described to involve the skin, mucosa, joints and internal organs but all the involvement may not be found in a single patient [1]. The most prominent clinical features are the cutaneous nodules and distinctive arthritis [2]. MR has few other names and in the literature it has been stated as 'lipoid dermatoarthritis', 'lipoid rheumatism', and 'giant cell reticulohistiocytosis'. The confirmation of diagnosis is based on the histological appearance of an infiltrative histiocytic multinucleated giant

cell with eosinophilic ground-glass cytoplasm [3].We report a case of multicentric reticulohistiocytosis with skin lesions resembling dermatomyositis probably the first reported case in Malaysia.

Case reporting:

A 36 year old Chinese lady was seen in one of our outstation dermatology clinic in June 2007. She complained of multiple skin coloured papules and nodules on the hands, elbows, ear, face and feet for the last 2 years **(Fig 1a- 1d)**. She also complained of photo distributed erythematous patches and plaques lesion on the front and back of the chest and some facial rash.

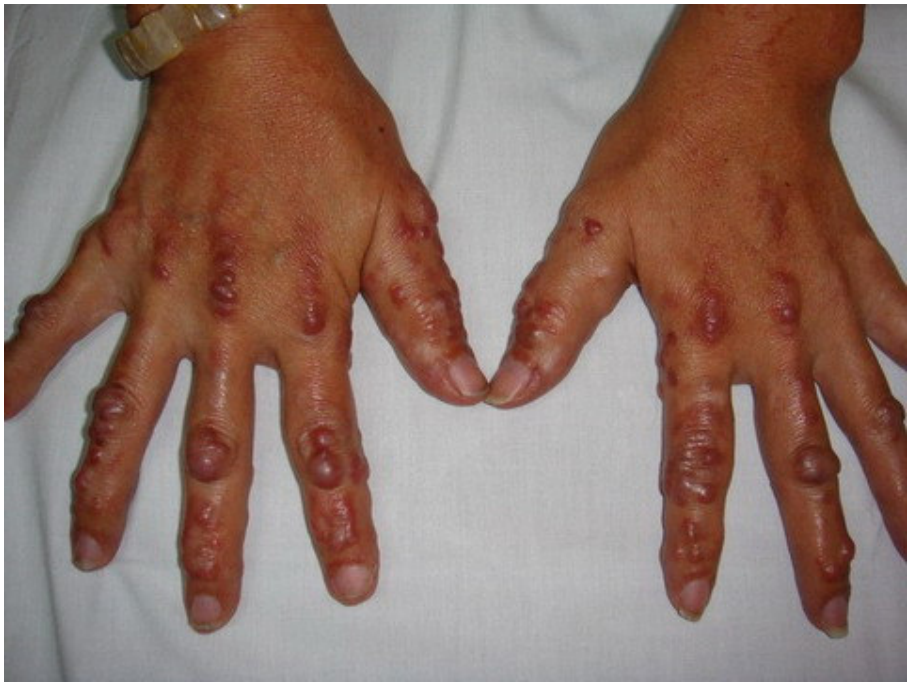

Fig 1a: Papules, nodules on the dorsum of the hands some resembling Gottron's papules.

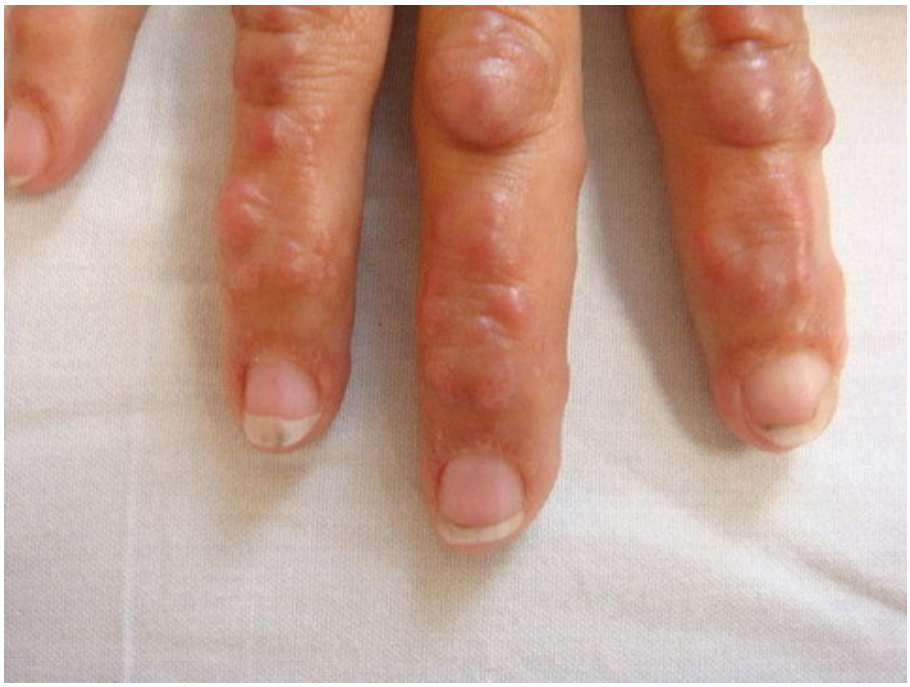

Fig 1b: Coral beading around the finger nails.

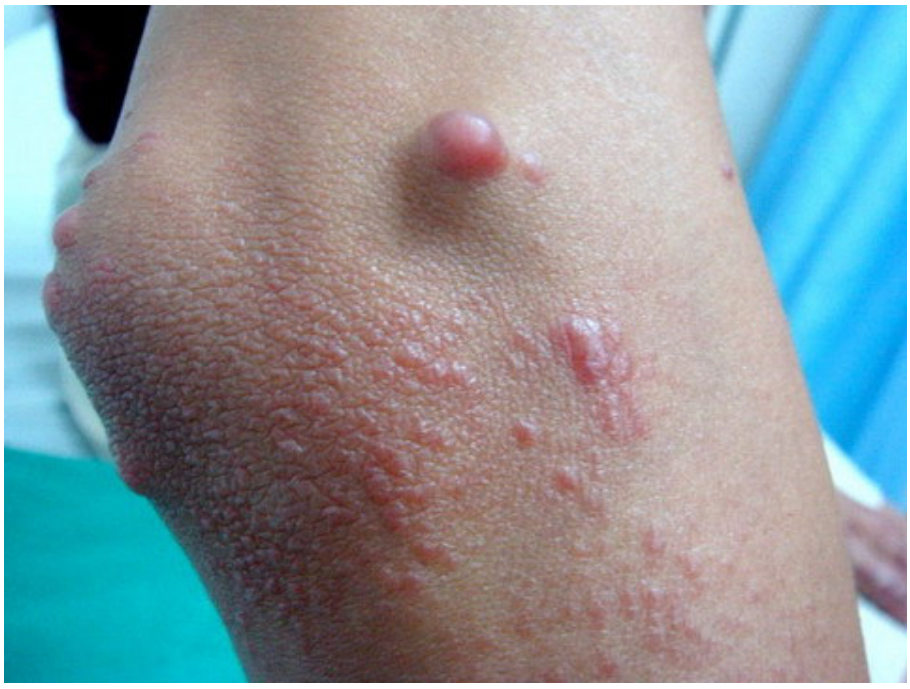

Fig 1c: Cobblestone appearance papules on the elbow.

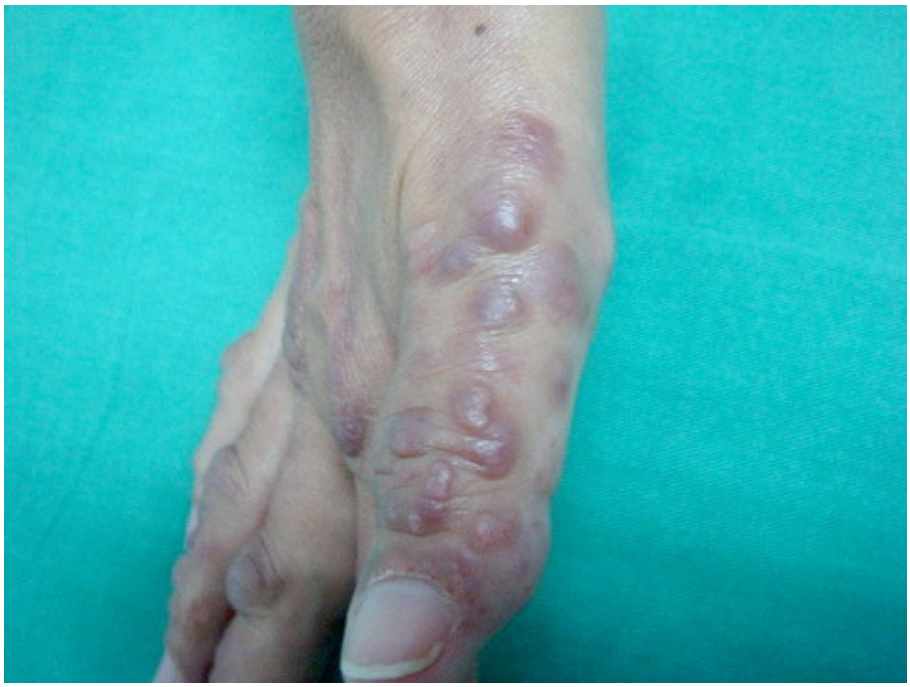

Fig 1d: Nodules of various sizes on the dorsum of the foot.

She also complained of multiple joint pains bilaterally involving the knees and the small joints of the hands associated with morning stiffness for the last 6 months. She had no proximal muscle weakness. She gave history of photosensitivity but did not complain of any systemic problem. On examining the skin there were multiple firm papules and nodules of different sizes ranging from few mm to 1.5 cm on the dorsum of the hand and fingers, legs, feet, toes, both extensor aspects of the elbows, ear and face. The papules and the nodules were painless and non pruritic. The papules on the elbows clustered together to form a cobblestone appearance. All routine blood and urine investigations were normal. Her specific investigations for Rheumatoid factor, Antinuclear antibodies, lipid profile, muscle enzymes, serum protein electrophoresis, faecal occult blood, thyroid function tests, screening for hepatitis B and C, HIV, VDRL, Mantoux test were found to be normal or negative. Her cancer markers; alpha fetoprotein, CEA, Ca-125 were normal too. Chest x-ray and abdominal ultrasound did not show any abnormal findings. Her X-rays of the hands and knees showed articular erosions in the interphalangeal joints and erosion on the patella bilaterally **(fig 2)**.

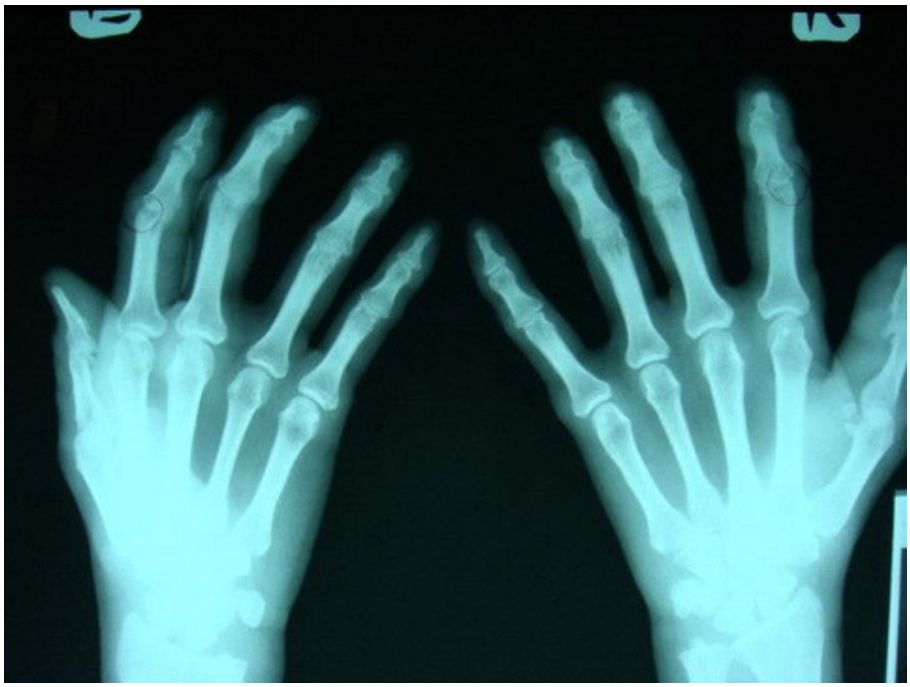

Fig 2: Erosions on the peripheral interphalangeal joints bilaterally.

Two skin biopsies; one taken from a papular lesion on the hand and another from a plaque lesion of the back showed similar pathology. The HPE revealed dermal infiltrates of multinucleated histiocytic giant cells with an eosinophilic 'ground glass' cytoplasm. Scattered lymphocytes were seen throughout the dermis. Immuno-histochemical study was positive for CD68 and negative for S100. These above findings confirm the diagnosis of multicentric reticulohistiocytosis **(fig 3)**.

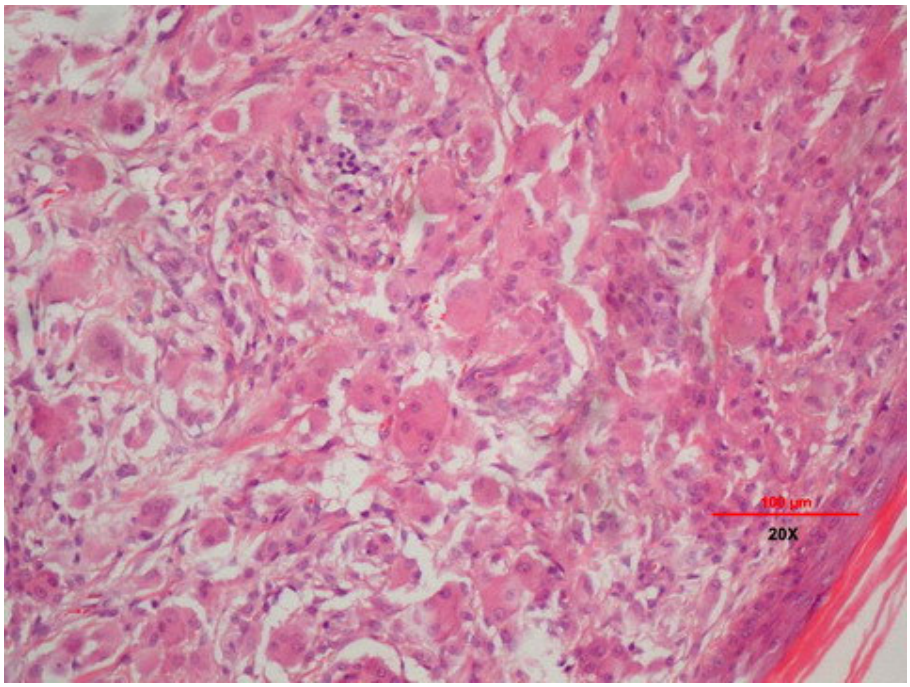

Fig 3: Multinucleated giant cells with an eosinophilic granular 'ground glass' cytoplasm.

We treated her with oral prednisolone and methotrexate initially. Prednisolone was slowly tapered down and stopped over a 4 months period and methotrexate was increased gradually from 5 mg to 15mg weekly and maintained on that dose. She improved significantly during this period .We noticed that most of the skin lesions were regressed and she complained of less joint pain at the end of one year treatment **(Fig 4- 6)**. Although initially we noticed some deformity of the distal interphalgeal joints of both the index fingers but during the follow-up visits we observed that the joints were stable with no pain. At the end of the two years in June 09, she maintained that improvement with no new skin lesions and no worsening of joint pains and she is still maintained on 15 mg of methotrexate weekly.

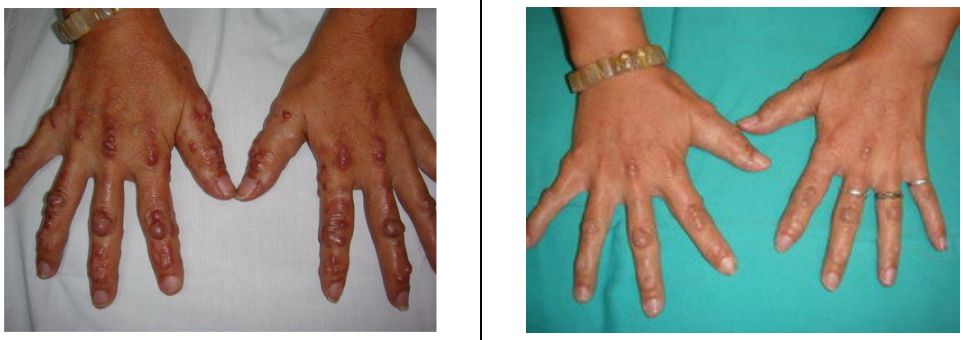

Fig 4: Shows the regression of the papules and nodules.

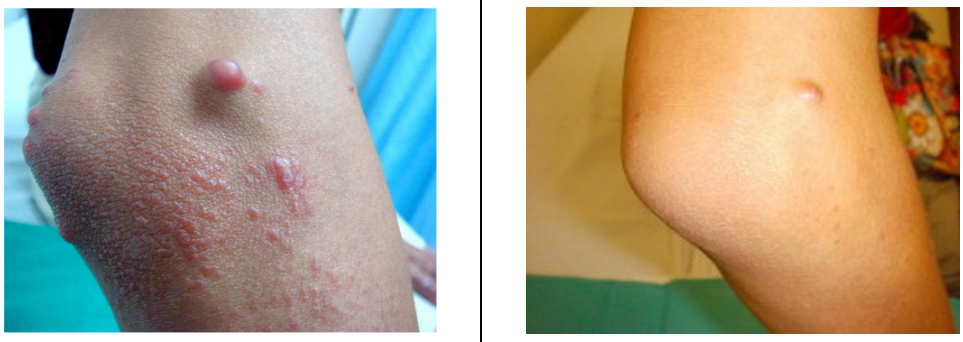

Fig 5: Shows no more cobble stone appearance on the elbow.

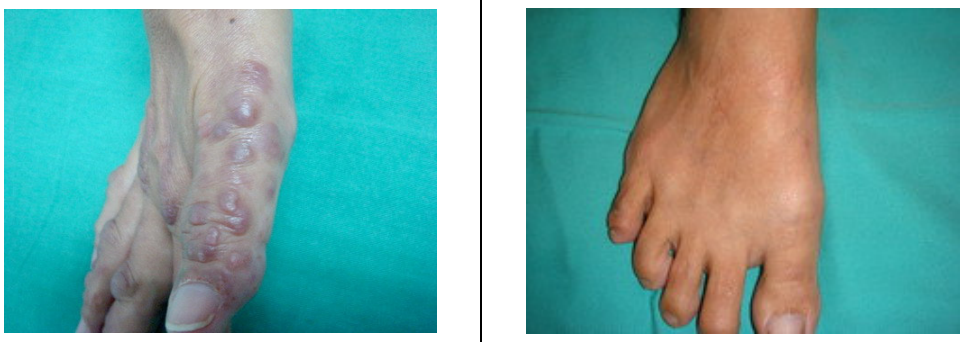

Fig 6: Completely cleared papules and nodules on the dorsum of the foot.

Discussion:

Multicentric reticulohistiocytosis is a rare disorder with systemic involvement for which no cause has been identified. This was first described by Goltz and Laymon [3] in 1954 and so far only less than 200 cases have been reported. Malignancy has been associated with MR [4]. There is a controversy regarding its paraneoplastic nature but in about 20% of cases it may be associated with malignancy [5]. Our patient discussed here represent with most of the cutaneous symptoms of MR and is most likely to be the first reported case in Malaysia.

In addition to other clinical manifestations, our case presented with erythematous papules on the knuckles of the hands **(fig 1a)**, a patch and like erythema on the face, plaque like eyrthema on the chest and back, which can be mistaken for dermatomyositis but in our case the presence of joint erosion without proximal muscle weakness and normal values of muscle enzymes were not in favour of dermatomyositis. Moreover, skin biopsies from two areas did not show any features of dermatomyositis but concluded the diagnosis of multicentric reticulohistiocytosis.

Polyarthritis has been described as the commonest first presenting symptom of MR [1,6]. We also noted similar presentation in our patient. Patient may present only with symptoms of joint pain in about 40% of cases, only skin lesions in 30%, and joint and skin symptom in about 30% of cases [5]. There may be other areas of involvement including the heart, lungs, thyroid, and bone marrow [5,7]. Papules may form around the nail producing a characteristic "coral beads" appearance as in our case **(fig 1b)**. In about one half to one third of cases mucosal involvement have been reported which may include the nasal and buccal mucosa, tongue, lips larynx and trachea [1,5]. Despite extensive skin involvement, our patient did not have any mucosal involvement. Balachandran et al [8] and Mittal et al [6] in their reports also did not mention any involvement of the mucosa.

There has been variable success in treating MR with corticosteroids, hydroxychloroquine, methotrexate, alkylating agents such as chlorambucil, cyclophosphamide, and azathioprine [9-12]. The disease activity has been reported to resolve after approximately 8 years [13]. Our patient responded significantly with combination of oral prednisolone and methotrexate and at the end of one year she had less joint pain and clearance of skin lesions. Prednisolone was tapered off at four months time. As MR can mimic dermatomyosistis, as in our case, it is important to differentiate between the two as early treatment can prevent deformity and disability in MR.

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