

## QUIZ SECTION

### Papules, Arthralgia, Dry Mouth and Dry Eyes: A Quiz

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A 59-year-old Korean woman presented with asymptomatic multiple papules on the superior aspect of her ears, the posterior neck and proximal nail-folds of her right thumb and right 3<sup>rd</sup> and 4<sup>th</sup> fingers for one month (Fig. 1). One month earlier, she visited her GP with a 2-month history of symmetrical polyarthralgia involving the wrists, fingers, shoulders and knees. She was treated with prednisolone, gabapentin and non-steroidal anti-inflammatory drugs for one week with no improvement. She also developed symptoms of dry mouth and dry eyes. She was transferred to the Department

of Rheumatology for further evaluation. The results of laboratory examination, systemic lupus erythematosus (SLE) profile studies, radiological examination, ultrasound-guided arthrocentesis, Schirmer's test, salivary gland scan and lip biopsy were not compatible with rheumatoid arthritis or Sjögren's syndrome.

The patient was transferred to our clinic for the evaluation and we performed a skin biopsy of a yellow-red papule on the right ear (Fig. 1C).

*What is your diagnosis? See next page for answer.*

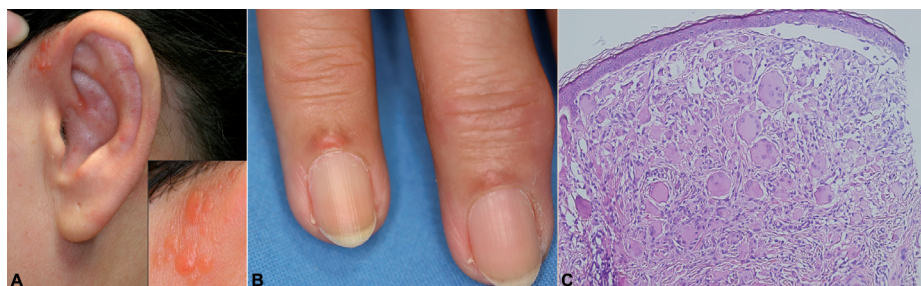


Fig. 1. Asymptomatic, multiple, 2–3-mm diameter, yellow-to-red papules on: (A) the superior aspect of the ears, and posterior neck (inset), (B) proximal nail folds of the right 3<sup>rd</sup> and 4<sup>th</sup>. (C) The biopsy specimen revealed a diffuse infiltration of numerous large, mononucleated or multinucleated histiocytes in the dermis (haematoxylin and eosin  $\times 100$ ).

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## ANSWERS TO QUIZ

**Papules, Arthralgia, Dry Mouth and Dry Eyes: Comment**

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**Diagnosis: Multicentric reticulohistiocytosis**

Multicentric reticulohistiocytosis (MRH) is a well-documented disease of unknown aetiology, which belongs to the group of non-Langerhans' cell histiocytosis. It is characterized by cutaneous or mucosal papular lesions with severe polyarthritides and arthralgias. Although the commonly affected sites are the skin and joints, involvement of the mucosa, muscles, cardiopulmonary system, eyes, gastrointestinal system, thyroid and salivary glands have been reported (1). In addition, a number of conditions, such as rheumatological disorders, diabetes, thyroid disease, tuberculosis and malignancies, have been reported to be associated with MRH (2–6). Cutaneous lesions of MRH usually present as diffusely scattered, asymptomatic, firm, smooth, yellow-to-red papules or nodules on the face, head, elbows and hands. However, the initial clinical findings of MRH can be diffuse symmetrical polyarthritides with arthralgia, and can therefore be misdiagnosed as rheumatoid arthritis (7). In addition, some extraordinary symptoms of MRH, such as xerostomia and dry eyes, may cause confusion with Sjögren's syndrome, as shown in our case. Therefore, careful history-taking, physical examination and histopathological evaluation of the cutaneous lesions are required for a definite diagnosis of MRH.

Treatment of MRH is not established, but reports of remission with immune-suppressants have been published. Tumour necrosis factor- $\alpha$  inhibitors, including etanercept, infliximab and leflunomide, have shown good results in recalcitrant cases of MRH (8–10). Our patient has been well managed with a combination therapy of prednisolone (starting with 8 mg/day, and gradually tapering to 2.5 mg/day), COX-2 inhibitor (400 mg/day), hydroxychloroquine (400 mg/day) and methotrexate (7.5 mg, once a week). The

patient's arthralgia disappeared 2 months after treatment, but there was only a slight improvement in the cutaneous lesions. Extensive investigation did not reveal any underlying malignancy.

## REFERENCES

1. Rapini RP. Multicentric reticulohistiocytosis. *Clin Dermatol* 1993; 11: 107–111.
2. Orkin M, Goltz RW, Good RA, Michael A, Fisher I. A study of multicentric reticulohistiocytosis. *Arch Dermatol* 1964; 89: 640–654.
3. Catterall MD. Multicentric reticulohistiocytosis: a review of eight cases. *Clin Exp Dermatol* 1980; 5: 267–279.
4. Codriansky KA, Runger TM, Bhawan J, Kantarci A, Kissin EY. Multicentric reticulohistiocytosis: a systemic osteoclastic disease? *Arthritis Rheum* 2008; 59: 444–448.
5. Ben Abdelghani K, Mahmoud I, Chatelus E, Sordet C, Gottenberg JE, Sibilia J. Multicentric reticulohistiocytosis: an autoimmune systemic disease? Case report of an association with erosive rheumatoid arthritis and systemic Sjogren syndrome. *Joint Bone Spine* 2010; 77: 274–276.
6. El-Haddad B, Hammoud D, Shaver T, Shahouri S. Malignancy-associated multicentric reticulohistiocytosis. *Rheumatol Int* 2011; 31: 1235–1238.
7. Baghestani S, Khosravi F, Dehghani Zahedani M, Mahboobi AA. Multicentric reticulohistiocytosis presenting with papulonodular skin eruption and polyarthritides. *Eur J Dermatol* 2005; 15: 196–200.
8. Lovelace K, Loyd A, Adelson D, Crowson N, Taylor JR, Cornelison R. Etanercept and the treatment of multicentric reticulohistiocytosis. *Arch Dermatol* 2005; 141: 1167–1168.
9. Kalajian AH, Callen JP. Multicentric reticulohistiocytosis successfully treated with infliximab: an illustrative case and evaluation of cytokine expression supporting anti-tumour necrosis factor therapy. *Arch Dermatol* 2008; 144: 1360–1366.
10. Lonsdale-Eccles AA, Haworth AE, McCrae FC, Young-Min SA. Successful treatment of multicentric reticulohistiocytosis with leflunomide. *Br J Dermatol* 2009; 161: 470–472.