Systemic mastocytosis and Sjögren's syndrome

Sir: I wish to comment on the case report by Bac and Marwick Koooy1 of a patient with systemic mastocytosis labelled as having Sjögren's syndrome. There is obviously some problem here with semantics and nomenclature. The patient described by the authors had features of systemic mastocytosis together with dry eyes and mouth; the latter manifestations apparently resulted from mast cell infiltration of the secretory glands as shown by the biopsy findings of the minor salivary glands. The authors do not mention any lymphocytic infiltration of the salivary tissues characteristic of Sjögren's syndrome, and I presume there was none.2 Nor was there any serological evidence of Sjögren's syndrome, such as the presence of antinuclear factors and specifically the Ro and La antibodies which are found frequently in this disorder.3 On the basis of the authors' findings, therefore, I do not believe that their patient fulfills the required diagnostic criteria for Sjögren's syndrome as described by Fox et al4 or, indeed, by others.5

In my opinion this patient with mastocytosis developed features of sicca syndrome (and not Sjögren's syndrome) because of lachrymal and salivary gland compromise due to heavy mast cell infiltration. Keratoconjunctivitis and xerostomia are characteristic features of this disease caused by lachrymal and salivary gland infiltration.2

Conversely, however, it has to be borne in mind that an occasional patient with primary Sjögren's syndrome may masquerade and be misdiagnosed as sarcoidosis,6 or indeed another rheumatic problem such as rheumatoid arthritis or lupus.7

It is important that Sjögren's syndrome is diagnosed only when patients meet the necessary diagnostic criteria of objective keratoconjunctivitis sicca and xerostomia in the presence of some serological markers of autoimmune dysfunction such as antinuclear factors or more characteristically the antibodies to extractable nuclear antigens—namely, Ro and La. In many cases a lower lip biopsy for histopathological evidence of the typical lymphocytic infiltration in the accessory salivary tissue is necessary to confirm the diagnosis. Patients with keratoconjunctivitis sicca and xerostomia due to other causes, such as sarcoidosis, should perhaps be more specifically called non-Sjögren sicca syndrome.

A H ISDALE
School of Medicine
Rheumatology and Rehabilitation Research Department
University of Leeds
36 Clarendon Road
Leeds LS2 9NZ
United Kingdom


Authors' reply

We would like to thank Dr Pal for his reaction. We completely agree with his statements and in fact we did send in our article with the title 'Mastocytosis and sicca syndrome'. However, this was changed by the editor to 'Mastocytosis and Sjögren's syndrome'. Obviously, there is still some debate about whether criteria are to be applied to patients diagnosed as having Sjögren's syndrome. If Sjögren's syndrome is regarded as an autoimmune disorder with specific histology and serological abnormalities then we should restrict this term to those patients who meet all criteria. We might then probably consider this also as Sjögren's disease.

It was our intention to describe a patient with the sicca syndrome caused by mast cell infiltration of the secretory glands, which was not reported before. Together with haemosiderosis, sarcoidosis, and amyloidosis mastocytosis should also be considered as a non-Sjögren cause of the sicca syndrome.

D J BAC
M van Marwijk Koooy Poli orthopedie Rheumatoïde
University Hospital Rotterdam
Dijkzigt Hospital
Doleizenplein 40
3015 GD Rotterdam
The Netherlands

Correspondence to: Dr van Marwijk Koooy.
Systemic mastocytosis and Sjögren's syndrome.

B Pal

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