Systemic mastocytosis and Sjögren's syndrome

Sir: I wish to comment on the case report by Bac and Marwijk Kooy1 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 fulfils 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 features of Sjögren's syndrome.5–7 It is characteristic of this disease that serological abnormalities are frequently found in patients who later develop this syndrome.8 Indeed, some authors9 recommend that any patient with the sicca syndrome should be investigated for Sjögren's syndrome. In the case reported by Bac and Marwijk Kooy, however, it was not investigated. Therefore, a diagnosis of systemic mastocytosis should be considered first.

I do not believe that this patient has Sjögren's syndrome. In any case, the patients with systemic mastocytosis have a systemic disease which cannot be treated with conventional immunosuppressives. I believe that a systemic mastocytosis patient with xerostomia should be treated with mast cell stabilisers, such as cromolyn sodium.5

The authors should refer to a specialist in Sjögren's syndrome before attributing her symptoms to this disease. I believe it is important to consider a systemic mastocytosis patient with the sicca syndrome, because of the possible side-effects of immunosuppressives.

I wish to thank Dr W. J. M. van Marwijk Kooy for permission to comment on his case report.

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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 for 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 which 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 haemorrhagia, salicorralia, and amyloliodosis mastocytosis should also be considered as a non-Sjögren cause of the sicca syndrome.

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doi: 10.1136/ard.51.10.1183-a

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