peripheral bones affected. Most axial lesions occurred in the T12 and L5 vertebrae. Thirty patients had peripheral bones affected, 19 unilaterally and 11 bilaterally. Fifteen of the unilateral metastases were ipsilateral to the primary tumour (eight right side, seven left side) and four contralateral (three right side, one left side). Of the 11 with bilateral metastases eight were right sided primary tumours and three left sided. Seventeen of the 40 (43%) had presented with symptoms related to their bone lesions. Thus right sided primary tumours are more likely to have either contralateral or bilateral bony metastases, which is supported by the recent report1 and our case.

Fixation, possibly with radiotherapy and cryotherapy, is recommended for a lytic lesion in a weightbearing bone if the life expectancy is greater than three or four months.

Metastases to periarticular foci are not uncommon, especially in the hip, shoulder, and knee, and the synovial reaction is either non-neoplastic or due to extension through the subchondral bone plate to affect the synovial membrane secondarily.2 There is nothing to suggest in the cases presented that there was primary involvement of the synovium, and we would thus suggest that what has been described is not rare but certainly might have been underreported recently.

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Rheumatic problem such as rheumatoid arthritis or lupus.3

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 but 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.

SIR:

Sir: I wish to comment on the case report by Bac and Marwijk Kooij1 of a patient with systemic mastocytosis (not 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 subepithelial 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 of this nature have been previously documented in other conditions where the subepithelial glands are infiltrated by various agents—for example, amyloid, haemosiderin6 and sarcoid granuloma.7 A sicca syndrome associated with idiopathic haemochromatosis has also been reported.8 These heterogeneous groups of ailments are not examples of Sjögren’s syndrome (although sometimes misdiagnosed as such, in the present case) but simply illustrations of glandular impairment clinically manifesting in the sicca syndrome.

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,9 or indeed another

Systemic mastocytosis and Sjögren’s syndrome


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 already 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 subepithelial 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.

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