Sarcoidosis or primary Sjögren’s syndrome?

Sir, We were interested to read the case report by Melsom and coworkers. We have been following up a patient who presented a similar difficult diagnosis.

A 59 year old woman was seen in 1983 complaining of a recurrent non-deforming, non-destructive arthritis since 1970. The sicca syndrome was obvious, with a Schirmer’s test of 0, positive rose Bengal staining, and keratoconjunctivitis. Antinuclear antibodies were not detected and latex agglutination was negative. Despite a normal erythrocyte sedimentation rate there were 20x10^9 cells/l (89% lymphocytes) in the synovial fluid of the left knee, and the synovial biopsy specimen was considered non-specific in spite of the presence of few epithelioid cells.

Clinically, the differential diagnosis lay between sarcoidosis and Sjögren’s syndrome. As the labial gland biopsy specimen showed acinar atrophy with fibrosis and a mild infiltration of lymphocytes Sjögren’s syndrome was suggested.

Two years later cervical lymphadenopathy developed, and biopsy showed non-caseating granulomata with multinucleate giant cells. The Mantoux test was negative, the angiotensin converting enzyme raised, and scintigraphy with gallium showed mediastinal and splenic uptake.

This patient presented in a similar fashion to the one reported, but the prolonged course of the articular disease with sicca syndrome before the appearance of the cervical adenopathy is in contrast.

We confirm that sarcoidosis may mimic Sjögren’s syndrome in the absence of features suggestive of sarcoidosis in the lip biopsy specimen.

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References

Sir, We have read the report of Melsom et al of a patient with sarcoidosis and a lip biopsy consistent with Sjögren’s syndrome (SS). Although no granulomata were present in minor salivary gland histological sections, the authors concluded that early involvement of these tissues by sarcoid mimicking SS was the most likely explanation for this finding. We have recently encountered a patient with similar clinical and histological features, in whom, however, we reached different conclusions.

The patient was a 62 year old woman with a 12 year history of recurrent polyarthralgia, anterior uveitis, and skin lesions suggestive of erythema nodosum. During the past three years she had presented a typical sicca syndrome. When she was referred to our clinic physical examination was normal except for the absence of saliva around the base of the lingual frenulum. Schirmer’s test showed decreased lacrimation (5 cm). Lip biopsy disclosed periductal fibrosis, acinar atrophy, and a moderately intense but diffuse lymphoplasmacytic infiltrate, consistent with histological class III (3+) on Tarpley’s classification.

Routine laboratory studies were normal and immunological markers were not detected. Chest radiographs and thoracic computed tomography showed bilateral hilar adenopathies. This posed the differential diagnosis between lymphoma and sarcoidosis in a patient with SS. The presence of uveitis did not necessarily favour sarcoidosis as it has been described in patients with SS. A mediastinoscopy directed biopsy of one of these nodes was thus performed. The pathological study showed multiple non-caseating granulomata as did a bronchoscopy directed bilateral transbronchial biopsy. The Mantoux test was negative. Mycobacterial and fungal microbiological studies were negative. We reviewed the lip biopsy sections thoroughly but still found no evidence of sarcoid infiltration.

Lip biopsy is an established method for histological confirmation of sarcoidosis. Even though the presence of granulomata is indispensable for this purpose, sarcoidosis may in some cases show non-specific findings, such as scattered lymphoplasmacytic infiltrates and multinucleated cells. Both of these may also be encountered in SS. Giotaki et al recently reviewed 60 lip biopsy specimens from 32 patients with sarcoidosis and 28 patients with SS.
Correspondence

They found that biopsy specimens from the patients with sarcoidosis showed lymphoid infiltrates consistent with class I (+) or less on Tarpley’s classification, and that all patients with class II (2+) or higher had SS. As we have not found any other studies on a similar scale that test the discriminatory efficacy of lip biopsy for these two diseases we believe that present evidence favours the hypothesis that our patients had both sarcoidosis and SS. Although Melsom and colleagues did not employ Tarpley’s classification, their description is compatible with at least class II (2+). Hence probably both diseases were also present in their case. A statistically significant association between SS and sarcoidosis is yet to be proved; nevertheless, it would not be surprising if we bear in mind that both diseases are characterised by an intense cellular immune response, predominantly composed of T lymphocytes, at sites of disease.7,8 Sarcoidosis could therefore become a new member in the list of immunological disorders associated with ‘secondary’ SS. Several clinically relevant considerations could be inferred from this association. The presence of adenopathies in a patient with a diagnosis of SS, as occurred in Melsom’s case as well as ours, suggests the possibility not only of an associated lymphoma but also of coexistent sarcoidosis. On the other hand, lip biopsy in patients with sarcoidosis may not only be useful as confirmatory evidence of sarcoid involvement, but also for eliminating concomitant SS.

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References


Sir, Sarcoidosis may present with swelling of salivary glands and symptoms of dry eyes and dry mouth very similar to Sjögren’s syndrome (SS).1 Giotaki et al showed that labial minor salivary gland biopsy can discriminate between sarcoidosis and SS.1 They also pointed out, however, that in sarcoidosis a mononuclear cell infiltrate may precede the development of granulomata in pulmonary tissue. They suggest that this situation may also occur in other tissues and that in salivary glands this would mimic the lymphocytic infiltrate seen in SS. We therefore read with interest the reports of two further patients who had the sicca syndrome and changes of SS on lip biopsy but also histologically proved sarcoidosis. Indeed, in the case described by Ferrer et al the features of polyarthralgia, erythema nodosum, anterior uveitis, and bilateral hilar lymphadenopathy are highly suggestive of sarcoidosis. Although it is possible that sarcoidosis and SS may have coexisted in these three patients either by chance or because of a common immunological disturbance, we believe the explanation is that the early changes of sarcoid in salivary tissue may mimic those of SS and that all three patients described have underlying sarcoidosis only. In this context it is interesting to note that none of these three patients had serology supporting a diagnosis of SS. Further evidence to support this view would be obtained if serial lip biopsies in patients with proved sarcoidosis demonstrated histological progression from lymphocytic infiltration to granuloma formation.

With reference to the point made by Ferrer et al, the lip biopsy of the patient we described had grade 4 changes of SS by the Chisholm and Mason Scale,2 which is equivalent to class III by Tarpley’s classification.3

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References


Micromechanical testing of articular cartilage: recent improvements to test apparatus

Sir, We feel that it will be of interest after the article by O’Connor et al in this journal1 to note recent technical advances made in our laboratory in micromechanical testing of articular cartilage. O’Connor et al measured the
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