key points

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Primary Sjögren's syndrome is a chronic systemic immune-mediated condition of unknown aetiology characterised by focal lymphocytic infiltration of exocrine (mainly salivary and lacrimal) glands. It affects 0.1-4.6% of the European population and 90% of cases are female. Although it usually presents in the fifth or sixth decade, it can be seen in younger people who tend to present with systemic disease and are less likely to have classical sicca symptoms.

Although patients generally present with sicca symptoms i.e. dryness of the mouth, eyes or vagina, other symptoms may include arthralgia, fatigue, Raynaud's phenomenon, sensorimotor neuropathy or a dry cough. In patients who develop an inflammatory arthritis, this tends to be polyarticular and symmetrical, typically affecting the wrists and small joints of the hands.

While Sjögren's syndrome is an important condition to consider in people presenting with sicca symptoms, a variety of other conditions may contribute to, or cause, mucosal dryness. These include head and neck radiotherapy, sarcoidosis, acute anxiety, IgG4 disease, hepatitis C and HIV. Drugs which may induce sicca symptoms include: opioids, antidepressants, beta-blockers, proton pump inhibitors, antihistamines and muscarinic antagonists. Chronic ocular conditions such as blepharitis and conjunctivitis are a common cause of dry eyes.

On examination the eyes may show dilatation of the conjunctival vessels, corneal lesions, and blepharitis. The mouth may look dry and a wooden tongue depressor may stick to the tongue. Patients are more prone to oral candidiasis and dental caries. There may be bilateral submandibular or, more obviously, parotid gland enlargement. There may be features of other autoimmune disorders such as rheumatoid arthritis, SLE, scleroderma and primary biliary cirrhosis. Patients with suspected primary Sjögren's syndrome should be tested for antinuclear antigen to look for the presence of anti-Ro/La antibodies.

Patients should be referred to a rheumatologist to confirm diagnosis, and this may involve scintigraphy/sialography and/or labial gland biopsy. Treatment comprises symptom control and immunosuppression, and multidisciplinary involvement. Glucocorticoids may be used in systemic flares, or at a low dose to improve sicca symptoms and salivary flow. Hydroxychloroquine may improve joint pain, fatigue, Raynaud's and sicca symptoms, while methotrexate is used in patients with inflammatory arthritis.

Severe systemic disease occurs in about 15% of cases. This may include interstitial lung disease, myositis, idiopathic thrombocytopaenic purpura, autoimmune liver disease, and renal involvement. B-cell lymphoma is a serious complication which affects 5-10% of patients. Pregnancy is associated with an increased risk of complications including preterm delivery, low birthweight, and pulmonary hypertension. Anti-Ro antibodies may cross the placenta and induce congenital heart block and neonatal lupus.