Immunological conditions

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Sjögren’s syndrome Sjögren’s syndrome (SjS) is a chronic autoimmune disease with lymphocytic infiltration and autoimmune injury to the salivary and lacrimal glands, leading to dryness of the mouth ology of the disease is evolving and is a complex interaction among genetic elements, environmental factors and abnormal host immunity . It mainly a ff ects women in the fourth decades, who present with enlargement of the salivary glands, especially the parotid. Typically there is a delay in the time taken to diagnosis of the disease. Studies have shown a delay of 2–6 months between the first consultation and diagnosis. SjS can be considered to have four stages: initiation stage, preclinical stage, asymptomatic SjS stage and overt SjS stage. Primary SjS is not associated with any illness or disease, whereas secondary SjS is associated with other autoimmune disorders, including systemic lupus erythematosus, rheumatoid arthritis and scleroderma (Table 54.1). W ith no diagnostic markers, the diagnosis is based on a set of five criteria proposed by the European Alliance of Associ ations for Rheumatology (EULAR). One of them is a biopsy of the sublabial salivary glands , which shows histological fea tures of focal lymphocytic sialadenitis. The criteria required f or diagnosis of SjS involve at least one focus score of more 2 than 50 lymphocytes per 4 /uni00A0 mm of parenchymal tissue. Other tests for early diagnosis are evolving; these include testing for increasing levels of biomarkers such as complement C3 and neutrophil elastase in saliva and tears. In addition, identifica tion of traditional antibodies such as anti-nuclear antibodies (anti-SSA/Ro or anti-SSB/La) and rheumatoid factor can also be used. However, a significant proportion of patients with SjS may be seronegative. Tr eatment depends on disease activity and the organs involved. Tear substitutes can be used and xerostomia is treated by dental and oral surgeons. Randomised controlled trials have not shown any benefit with hy droxychloroquine or disease-modifying antirheumatic drugs (DMARDs). Treat ment with glucocorticoids and/or immunosuppressant drugs should be considered in severe systemic manifestations. /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Scleroderma This is an immunologically mediated disease with complex interactions between the vascular network, inflammatory markers and collagen tissue. It mainly a ff ects adults with a female preponderance. Multiple organs may be a ff ected by Christian Frederick Heerfordt , 1871–1953, Danish ophthalmologist, described this syndrome in 1909. Edward C Stafne , 1894–1981, dental surgeon, The Mayo Clinic, Rochester, MN, USA, described these cysts in 1942. is more commonly a ff ected, it can also involve the heart, lungs, kidneys and gastrointestinal tract. Salivary gland parenchyma to fifth may be replaced by collagen tissue, leading to clinical xero - stomia. Biopsy of a minor salivary gland may be useful for diagnosis. Sarcoidosis This is an inflammatory disorder characterised by multiple non-caseating granulomas in multiple systems. A dry cough, fatigue and shortness of breath are its main symptoms. The chest radiograph typically shows bilateral hilar lymphadenop - athy and reticular opacities in the lungs. Skin, heart, kidney , eyes, joints, exocrine glands and the central nervous system may be involved. The aetiology is unclear but one proposed - mec hanism is where an individual with a susceptible genotype is exposed to one or more potential antigens, resulting in a - sustained inflammatory response. The

disease most commonly occurs in those aged 20–60, in people of African or northern European descent and in those with a family history of the disease. Triggers are thought to include infection (mycobac - teria, propionic bacteria and viruses) or exposure to certain chemicals or dust. The function of an organ can begin to be - a ff ected as granulomas form and enlarge. For the salivary glands, the patient can pr esent with a local - ised tumour-like swelling, usually in the parotid – the so-called sarcoid pseudotumour along with xerostomia. In the absence of other disease, the diagnosis is usually made following surgi - cal excision for a presumed neoplasm. Heerfordt's syndrome is a rare manifestation of sarcoidosis that involves parotid swell - ing, anterior uveitis, facial palsy and fe ver. -

TABLE 54.1 Degenerative disorders of the salivary glands. Primary Sjögren's More severe xerostomia syndrome Widespread exocrine gland dysfunction No connective tissue disorder Secondary Sjögren's Male-to-female ratio 1:10 syndrome Middle age Underlying connective tissue disorder Benign lymphoepithelial Diffuse parotid swelling 20% lesion bilateral 5% develop lymphoma

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