Table SI. Main symptoms characteristic of Mikulicz's disease and differentiation with Sjögren's syndrome (SS) (based on 3, 13–15 with own modifications)

Features	Mikulicz's disease	Sjögren's syndrome
Clinical features		
First symptoms	Early signs may mimic infections or chronic inflammatory process.	
	The disease may be asymptomatic in the early stage.	Arthralgia, arthritis, ocular dryness or xerostomia.
Dynamics of symptoms development	Slow, over many months.	Relatively quick.
Character and relapsing of lesions	Bilateral, symmetrical and painless, swelling of the lacrimal, parotid, and submandibular glands. The swellings are persistent and maintain for more than 3 months.	The swellings occur repeatedly and disappear without treatment.
Functioning of the involved glands	Saliva and tear secretion normal or slightly reduced. Xerostomia occurs less commonly than in SS, less severe, usually sensitive to treatment.	Xerophthalmia and/or xerostomia occur.
Other associated symptoms	Lymphadenopathy of the head and neck.	Raynaud's phenomenon, arthralgia or arthritis.
Association with other diseases	Frequently (30–40%) occurs with allergic conditions.	SS can occur as a primary disease or co-exist with another autoimmune disease (secondary SS).
Treatment response	Good response to steroid therapy.	Immunosuppressive drugs play a major role in treatment.
Laboratory and serological tests		
Serum IgG4 level	Normal or significantly increased.	Normal or slightly elevated.
	Elevated ratio of serum IgG4 to total IgG.	No changes in serum immunoglobulin levels
	Elevated level of IgE correlating with IgG4.	
Other immunoglobulins	Normo-/hypergammaglobulinaemia.	No changes
	Normo-/hypercomplementaemia.	
Blood morphology	Eosinophilia (one-third of patients with IgG4-RD).	Thrombocytopaenia
Anti-SS-A/SS-B antibodies	Negative	Mostly positive
ANA, ANCA	Negative or insignificant (2)	Positive
Histopathology		
Microscopic image	Dense lymphoplasmacytic infiltrates, storiform (whirl-shaped) fibrosis, obliterative phlebitis.	No detection of IgG4-positive cells.
	IgG-positive cells infiltrate.	
Biopsy localization	Submandibular or labial salivary glands. (Low sensitivity of detection of IgG4+ plasma cells in specimens from labial glands).	Labial or submandibular salivary glands. Parotid salivary glands biopsy for monitor the disease progression.
Imaging		
Salivary gland ultrasonography (SGUS)	Changes mainly in submandibular glands, with normal appearance of parotid glands. (14)	Atrophic changes in submandibular and parotid glands.
	Submandibular glands: The nodal and reticular patterns with normal parenchyma surrounding the affected region, high vascularity.	Submandibular glands: multiple hypoechoic areas, hyperechoic lines and/or spots, and a reticular pattern.
	Parotid glands: normal appearance or similar to SS. Hypoechoic areas in the normal parotid parenchyma without a reduction in the echo intensity level and heterogeneity (15).	Parotid glands: multiple hypoechoic areas and hyperechoic lines and/or spots in the parotid glands. Small dot-like vascularity in Doppler modality.
Other specific methods	FDG-PET/CT (abnormal accumulation of 18F-FDG in the submandibular glands).	MRI ("salt-and-pepper" appearance and "honeycomb" multiple cystic areas in the parotid glands).
	MRI – non-specific lesion pattern.	

ANA: anti-nuclear antibodies; ANCA: anti-neutrophil cytoplasmic antibodies: MRI: magnetic resonance imaging; FDG-PET/CT: fluorodeoxyglucose positron emission tomography/computed tomography.