

**On-line Table 1: Diagnostic criteria for IgG4+ Mikulicz disease<sup>11</sup>  
(approved by the Japanese Society for Sjögren's Syndrome, 2008)**

Symmetrical swelling of at least 2 pairs of lachrymal, parotid, or submandibular glands continuing for at least 3 months and Elevated serum IgG4 (>135 mg/dl); or Histopathological features including lymphocyte and IgG4+ plasma cell infiltration (IgG4+ plasma cells/IgG+ plasma cells >50%) with typical tissue fibrosis or sclerosis

Differential diagnosis is necessary to distinguish IgG4+ Mikulicz disease from other distinct disorders, including sarcoidosis, Castleman disease, Wegener granulomatosis, lymphoma, and cancer. The diagnostic criteria for Sjögren syndrome (SS) may also include some patients with IgG4+ Mikulicz disease; however, the clinicopathologic conditions of patients with typical SS and IgG4+ Mikulicz disease are different.

**On-line Table 2: Summary of affected sites, including other organs in 15 patients**

Case/ Age/ Sex	Lacrimal Gland Enlargement	Intraorbital Mass	Cranial Nerve Enlargement	Pituitary Gland Enlargement	Dural Thickening	SMG Enlargement	Other CNS HN Lesions	AIP	Other Abdominal Lesion	Chest Lesion
1/73/M	+ bil		+ (V2; ION)	+ (St, PL)				—		
2/68/M	+ bil							—		Lung
3/53/M	+ bil		+ (V2; ION)					—		
4/73/F	+ bil							+	Bile duct	
5/67/M		+	+ (V1; FN, CS)					—		
6/62/M	+ bil	+		+ (St, PL, AL)				+		Lung
7/72/M	+ bil	+			+	+		—	Kidney	
8/66/M		+	+ (CS)		+			—		MF
9/50/M	+ bil	+						+		
10/59/M			+ (CS)	+ (St, PL, AL)	+			+	Kidney, RPF	Lung
11/59/M			+ (V2; ION)	+ (St, PL)	+			+	RPF	Hilar LNs
12/61/M						+		+	RPF	Lung, hilar LNs
13/45/M	+ bil							—		
14/65/F						+		—	RPF	
15/35/F			+ (V2; ION)	+ (St, PL)			Multiple <sup>a</sup>	—	•	•

+ indicates presence; —, absence of a lesion; bil, bilateral; V1, first division of the trigeminal nerve (Vth nerve); V2, second division of the trigeminal nerve; FN, frontal nerve; CS, cavernous sinus; ION, infraorbital nerve; St, pituitary stalk; PL, posterior lobe; AL, anterior lobe; SMG, submandibular gland; AIP, autoimmune pancreatitis; RPF, retroperitoneal fibrosis; MF, mediastinal fibrosis; LN, = lymph node.

<sup>a</sup> paranasal sinusitis, facial nerve lesion with otitis media, concentric thickening of the vertebral artery, and nerve root lesion.