

Table 2. *The immune components of IgG4-related disease.*

Pattern	Pancreatic-Biliary- Liver Disease	Retroperitoneum and Aorta	Head and Neck	Mikulicz’s and Systemic Disease
Typical manifestations	Type 1 Autoimmune Pancreatitis (Type 1 AIP) Sclerosing Cholangitis	Retroperitoneal Fibrosis Large Vessel Disease	Enlargement of salivary and/or lacrimal ducts; eye involvement.	Symmetrical enlargement of lacrimal/salivary glands with involvement of the thorax/abdomen
Dominant sex	Male	Male	Female	Male
Average Age (years)	63	58	55	63
Serum IgG4	Elevated	Normal or elevated	Elevated	Markedly elevated
Inflammatory markers (CRP/ESR)	Normal	Elevated	Normal	Normal
Mimickers	Pancreatic cancer Autoimmune Pancreatitis type II (AIP type II) Primary Sclerosing Cholangitis Cholangiocarcinoma	Lymphoma Erdheim-Chester disease Takayasu arteritis Giant cell Arteritis	Connective Tissue Diseases Giant Cell Arteritis Lymphoma	Connective Tissue Disease Giant cell Arteritis Lymphoma
Fibrosis	Absent	Present	Absent	Present