At an international consensus conference in 2011, multifocal chronic fibrosing inflammatory processes, which are associated with elevated IgG4 serum levels and/or tissue infiltration with IgG4 positive plasma cells, were recognized as a distinct disease entity called IgG4-related disease (IgG4-RD). As IgG4-RD responds well to steroid treatment but imitates a tumor in many organs, particularly in the pancreas, a biopsy for confirmation of the diagnosis is often warranted. The histological criteria for IgG4-RD as defined in 2011 are based on the following main features: 1) dense lymphoplasmacytic infiltrate, 2) storiform fibrosis and 3) obliteratorive phlebitis. The diagnosis is further supported by immunohistochemical demonstration of an increased infiltration of IgG4-positive plasma cells and an elevated IgG4/IgG ratio. The morphological criteria of IgG4-RD are in most cases detectable in biopsies and can significantly contribute to the diagnosis of this disease, in concert with clinical, serological (elevated serum IgG4 level) and radiological features.