

Table 3. *Unusual sites of IgG4-RD with their clinical presentation and the differential diagnosis.*

Organs	Clinical presentation	Differential diagnosis
Lymph nodes [2,3,7,19,42,43]	Localized or generalized, non-tender lymphadenopathy. Their size ranges from 1 to 3 cm and occasionally causes compression of vital structures.	Lymphoproliferative, infectious, and other immunologic diseases
Lungs [2,3,7,19,30,44-46]	Incidental findings on radiographs (~75%) or non-specific symptoms (cough, sputum production, dyspnea, chest pain, and hemoptysis). Imaging reveals tumefactive lesions, central airway disease, or pneumonitis. Pulmonary fibrosis is a potential complication with traction bronchiectasis and honeycombing. Reactive hilar and mediastinal lymphadenopathy is common.	Lung cancer, vasculitis, sarcoidosis, and other causes of interstitial lung disease
Serosal cavities [2,3,7,30,44,46-52]	Pleural disease appears as effusion, increased vascularity, thickening, nodular lesions, plaques, and adhesions within the pleural cavity. Acute pericarditis, pericardial effusion, constrictive pericarditis, and ascites are other manifestations. A case involving ascites, massive right pleural effusion, pericardial effusion, and elevated tumor markers (cancer antigen 125 and α -fetoprotein) may suggest pleuroperitoneal communication in the context of Meigs or pseudo-Meigs syndrome.	Acute coronary syndrome, infectious diseases, malignancies, rheumatic diseases, and cardiac myxoma
Skin [2,53]	Nodules, plaques, masses, macules, papules, or gangrene mainly distributed in the upper body, especially the head and neck area. Usually accompanied by internal organ involvement.	Lymphoproliferative disorders, psoriasis, cutaneous lupus, and systemic sclerosis
Central nervous system [3,7,19,30,54]	Pachymeningitis, cerebral inflammation, and cavernous sinus thrombosis cause cranial nerve palsy, radiculopathy, headache, sensorimotor abnormalities, visual deficits, sensorineural hearing loss, or dementia. The term “hypertrophic pachymeningitis” describes mass-like lesions in the meninges. Pituitary pathology, such as edema or tumor-like lesions, leads to headache, and visual field defects like bitemporal hemianopsia.	Infectious diseases, malignancies, vasculitis, and infiltrative disorders, e.g. sarcoidosis, histiocytosis.
Peripheral nerves [2,19,54]	Rapid or subacute development of sensorimotor neuropathy or the formation of perineural masses.	Vasculitis, demyelination
Prostate [2,3,19,55]	Urgency, incomplete voiding, and hesitancy.	Benign prostate hyperplasia, prostatitis, prostate cancer
Breast [56-58]	Painless and hard breast mass.	Breast cancer