

Supplementary Table 1. Summary of the Patient in Our Study and Individuals with *TRAF3* LoF Variants Reported in Previous Studies

Patient	Rae et al. ⁵						Urban et al. ⁶						This patient	
	A1	B1	B2	B3	B4	C1	C2	D1	E1	F1	F2	G1		
<i>TRAF3</i> variant	p.Y425*	p.S356Pfs6*				p.Q114*		p.R163*	p.Y425*	p.R163*		p.Q407*	p.P487Lfs8*	
Effect on protein	Stop-gain	Stop-gain				Stop-gain		Stop-gain	Stop-gain	Stop-gain		Stop-gain	Stop-gain	
Age, Sex	40, M	10, F	15, M	47, F	71, F	15, F	50, M	15, M	20, M	43, F	67, F	36, M	6, F	
Age of onset	Childhood	Childhood	Adolescence	Childhood	n.a.	Childhood	Adolescence	Childhood	Childhood	Childhood	Childhood	Childhood	Childhood	
Origin	Unknown	Dutch	Dutch	Dutch	Dutch	Dutch	Dutch	Unknown	Mexican	Spanish	Spanish	Argentinian	Japanese	
Infections	RSPI, ROM	RSPI, ROM, lymphadenitis with abscess, pulmonary NTM	Recurrent sinusitis, ROM	RSPI, lymphadenitis, peritonitis	RSPI	RSPI, lymphadenitis	RSPI	ROM, lymphadenitis	RSPI, ROM, pulmonary NTM	RSPI	RSPI	RSPI	ROM, lymphadenitis	
Lymphoproliferation	SMG, PLA	SMG, PLA	-	SMG	-	PLA	-	Tonsils, adenoids, peripheral LA	SMG, cervical, inguinal, PLA	SMG	SMG, bilateral cervical, retropectoral, and axillary LA	SMG, paratracheal and mediastinal LA	-	
Allergy/Atopy	-	-	-	-	-	-	-	AD, allergic sensitization	AD, allergic sensitization, food and drug allergy	AD	-	Pyoderma gangrenosum	-	AD
Autoimmunity	Sicca syndrome	-	-	Sjogren's syndrome	Sjogren's syndrome, vasculitis	Celiac disease	-	Hashimoto's thyroiditis, sJIA	Vasculitis, APS	-	-	-	Sjogren's syndrome, osteomyelitis, arthritis	
Pulmonary Involvement	BE, lymphoid hyperplasia	BE	-	BE	BE	BE, non-allergic asthma	-	-	BE, follicular bronchitis, lung fibrosis	BE	Interstitial lung disease	BE	Asthma	
Gastrointestinal Involvement (endoscopic and pathological findings)	Lymphoid hyperplasia with patchy ulceration	-	-	-	-	-	-	-	Chronic esophagitis, pan-gastropathy, chronic nodular duodenitis	Chronic atrophic gastritis, nodular lymphoid hyperplasia, intestinal metaplasia	Nodular lymphoid hyperplasia, ulcerative ileitis, granuloma	Nodular lymphoid hyperplasia, ileitis, active chronic pancolitis	Multiple small ulcerations on the edematous mucosa in stomach, small intestine, and colon, cryptitis and crypt abscess	
Immunoglobulin	IgG: low IgA: low IgM: low	IgG: high IgA: normal IgM: normal	IgG: high IgA: normal IgM: high	IgG: high IgA: normal IgM: high	IgG: normal IgA: normal IgM: normal	IgG: high IgA: low IgM: high	IgG: high IgA: high IgM: normal	IgG: high IgA: high IgM: high	IgG: high IgA: normal IgM: ormal	IgG: low IgA: low IgM: low	IgG: low IgA: low IgM: low	IgG: low IgA: low IgM: low	IgG: high IgA: high IgM: normal	

M, male; F, female; RSPI, recurrent sinopulmonary infections; ROM, recurrent otitis media; NTM, nontuberculous mycobacteria; SMG, splenomegaly; PLA, pulmonary lymphadenopathies; LA, lymphadenopathy; AD, atopic dermatitis; sJIA, systemic juvenile idiopathic arthritis; APS, antiphospholipid syndrome; BE, bronchiectasis; Ig, Immunoglobulin.