

SUPPLEMENTAL MATERIAL

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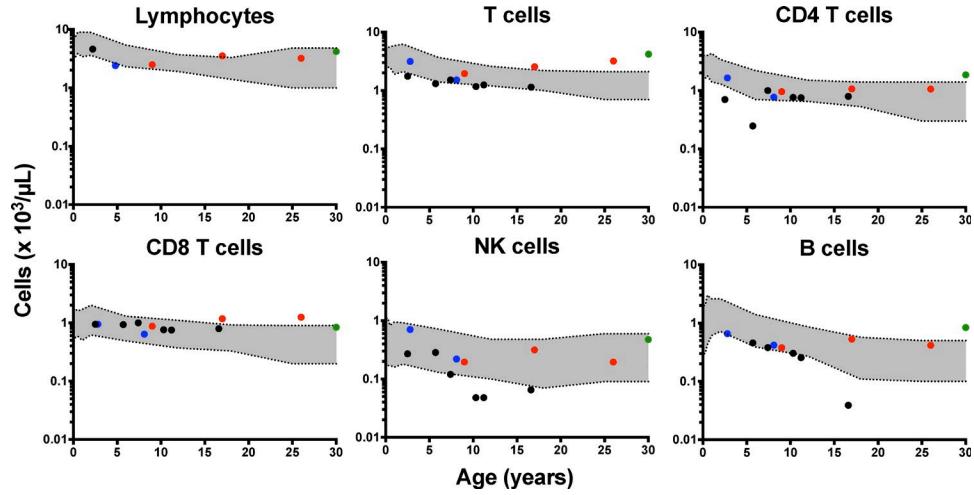


Figure S1. Enumeration of peripheral blood lymphocyte subsets in the patients. Solid circles indicate values graphed according to age of the patients. Red, P1; green, P2; black, P3; red, P4. Each dot represents one data point taken at that particular time. Gray shading indicates the 10–90th percentile for normal ranges according to age (Shearer et al., 2003).

Table S1. Complementary immunological and virological investigations in patients from family 1

Parameter	P1	P2	Reference range
C3 (IU/ml)	112	NA	70–170
C4 (IU/ml)	24	NA	15–55
CH50 (%)	94	NA	90–100
Nitro blue tetrazolium chloride test (%)	100	NA	95–100
Tuberculosis skin test (PPD test)	Negative	NA	5–10 mm
Diphtheria tetanus skin test	5 mm	NA	5–10 mm
Allergen-specific skin prick test	Negative	NA	Negative
			Negative cut off
Anti-HCV antibody (ELISA)	Negative	Negative	<0.8 S/CO
Anti-HCV immunoblot assays (Western blot and RIBA)	Negative	Negative	<1
Anti-HIV-1/2 IgG-Ab (EIA)	Negative	Negative	<0.9 S/CO
Anti-CMV, IgM-Ab, CMIA (AU/ml)	Negative	Negative	<0.85 index
Anti-CMV, IgG-Ab, CMIA (AU/ml)	180.6^a	187.4	<6
Anti-EBV, EBNA IgG-Ab, CMIA	14.56^a	6.33	<0.5 S/CO
Anti-HSV-1, IgG-Ab	4.5^a	8.1	<0.9
Anti-HSV-2, IgG-Ab	Negative	Negative	<0.9
Anti-mycoplasma pneumoniae, IgM-Ab (EIU)	Negative	Negative	<1.1 S/CO
Anti-mycoplasma pneumoniae, IgG-Ab (EIU)	58^a	Negative	<1.1 S/CO
Anti-TBE virus, IgM-Ab	Negative	Negative	<0.5
Anti-TBE virus, IgG-Ab	Negative	Negative	<7
Anti-toxoplasma, IgM-Ab, CMIA (IU/ml)	Negative	Negative	<0.5
Anti-toxoplasma, IgG-Ab, CMIA (IU/ml)	4.6^a	1.8	<1.6
Anti-VZV, IgM-Ab, EIA (mIU/ml)	Negative	Negative	na
Anti-VZV, IgG-Ab, EIA (mIU/ml)	1,100^a	390	<50
Anti-helicobacter pylori, IgG-Ab (IU/ml)	72	NA	na
Anti-helicobacter pylori, IgA-Ab (IU/ml)	4.3	NA	na
			Reference range
IgG1 (mg/dl)	402 ^a	436	280–800
IgG2 (mg/dl)	320 ^a	447	115–570
IgG3 (mg/dl)	17 ^a	27	24–125
IgG4 (mg/dl)	24 ^a	91	5–125
Antibody level before P23 (U/ml)	1 ^a	2	1.2–2
Antibody after 3 wk P23 (U/ml)	16 ^a	17	10–18
Antibody after 1 yr P23 (U/ml)	16 ^a	15	6–20
Isohemagglutinin titer (anti-B) ^b	0	0	>1/8
Triiodothyronine or T3 (RIA, nmol/L)	1.2	NA	1.08–3.14
Thyroxine or T4 (RIA, nmol/L)	55	NA	70–142
Free thyroxine (RIA, nmol/L)	6.02	NA	4.5–14
Thyroid stimulating hormone (IRMA, mIU/L)	0.97	NA	0.36–3.98
Rheumatoid factor (IU/ml)	5	NA	0–8
Direct coombs	Negative	NA	Negative
Indirect coombs	Negative	NA	Negative
Anti-nuclear antibodies (ELISA)	Negative	NA	Negative
Fluorescent antinuclear antibody (titer)	<1/320	NA	<1/80
Anti-double-strand DNA (IU/ml)	4	NA	0–12
Human leukocyte antigen B27	Negative	NA	Negative
Anti-cyclic citrullinated peptides (IU/ml)	1.1	NA	0–5
Parathyroid hormone (ECL, pg/ml)	26	NA	15–65
25-OH vitamin D (ECL, ng/ml)	33	NA	10–53

Abbreviations used: CMIA, chemiluminescent microparticle assays; CO, cut off; ECL, electrochemical luminescence; EIA, enzyme immunoassay; HCV, hepatitis C virus; na, not available; NA, not analyzed; PPD, purified protein derivative; RIA, radioimmunoassay; RIBA, recombinant immunoblot assay; IRMA, immunoradiometric assay; S, signal; TBE, tick-borne encephalitis; VZV, varicella zoster virus.

^aP1 is receiving IVIG therapy.

^bThe blood group of both P1 and P2 was A*.

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