

APPENDIX 1 - Sample characterization.

Pt	Sex	Profile	Neurological syndrome	CSF cell count Cell/mm ³	CSF protein g/L	CTC	Other IS	Systemic involvement	PET hypermetabolic regions	Serum IgG4 onset/after CCT mg/dL	CSF IgG4 mg/dL	Type of IgG4-RD diagnosis
1	F	chronic	Meningoencephalitis (cognitive deterioration, seizures, dizziness, gait instability)	14 (MN)	1,06	IV, PO	CLO AZA	Lung micronodule	-	145/91.2	12,4	Possible
2	M	subacute	Meningoencephalitis/Hypertrophic pachimeningitis (headache, fever, gait instability)	195 (MN)	1,05	IV, PO	AZA	Pericardial effusion); mediastinal fat densification	-	197/36.2	1,23	Possible
3	F	subacute	Orbital pseudotumor (retroorbital pain, oculodystyria, CN III and VI palsy)	1 (MN)	0,32	PO	CTC	Parotid and lachrymal gland nodules; mediastinal fat densification	-	196/136	0,392	Definitive
4	F	acute	Cranial neuropathy (III)	0	2,92	IV, PO	AZA CTC	Thyroid nodules	Subcutaneous nodules	191/-	5,8	Possible
5	F	subacute	Orbital pseudotumor (retroorbital pain, oculodystyria, CN III palsy); Meningoencephalitis/Hypertrophic pachimeningitis	15 (MN)	0,32	IV	RTX	Parotid gland enlargement	Parotid	194/60	0,55	Possible
6	F	subacute	Recurrent meningoencephalitis (headache, fever)	180 (MN)	0,87	-	AZA	Thyroid nodule; Pancreatic nodule	Thyroid	170/-	5,3	Possible
7	F	subacute	Cavernous sinus syndrome (retroorbital pain, CN III and VI palsy, facial hypoesthesia)	-	-	IV	AZA	-	-	264/72.3	-	Possible
8	M	chronic	Peripheral neuropathy (sensitive ganglionopathy)	2 (MN)	0,28	IV	RTX	Lung micronodule and bronchiectasis; obstruction of unilateral parotid gland; pancreas cyst.	Hilar-mediastinal lymph nodes	67,8/37.6	0,291	Probable
9	F	subacute	Radiculopathy, Adie's pupil	4 (MN)	0,66	PO	-	Lung micronodule, liver nodules, sclerotic lesion of iliac wing.	Lung micronodule, hilar-mediastinal lymph nodes, iliac wing	110/60.8	-	Possible
10	M	acute	Multiple cranial neuropathy (vertigo, diplopia, dysphonia, CFP)	13 (MN)	1,07	-	-	-	.	110/-	0,861	Possible

11	M	acute	LETM (cervical-dorsal pain, paraparesis)	88 (MN)	0,82	IV, PO	CTC	Lung micronodule and emphysema	Hilar-mediastinal lymph nodes	231/-	-	Possible
12	F	subacute	LETM (dorso-lumbar pain and paraparesis)	230 (MN)	39	IV, PO	RTX MTX	Hepatomegaly	-	177/-	14,7	Possible
13	F	acute	Meningoencephalitis (headache, fever)	630 (MN)	3,21	IV, PO	AZA CTC	Mediastinal fat densification)	Tonsils, spleen, nasopharynx	102/96.8	-	Possible
14	M	-	Pachimeningitis (imaging finding)	1 (MN)	0,44	-	-	Emphysema; parotid gland enlargement; thyroid nodules	-	212/-	0,51	Possible
15	M	acute	Right hemiparesis and hemihyposnesia	2 (MN)	0,28	IV	-	Interstitial lung disease	Thyroid, hilar-mediastinal lymph nodes	179/-	-	Possible

AZA, azathioprine; CLO, cyclophosphamide; CN, cranial nerves; CTC, corticosteroids; CSF Cerebrospinal fluid; LETM longitudinal extensive transverse myelitis; MTX, methotrexate; RTX, rituximab; PET – positron emission tomography