## PNS-relevant autoantibodies against intracellular neuronal antigens with associated neurological diseases

## And possibly associated tumors

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Autoantibody (Synonym(s))	Antigen (Molecular mass(es)) Localisation (L)	Associated diseases Most frequent syndromes (FS) Other syndromes(OS)	Associated tumors Most frequent tumors(FT) Other tumors (OT)
Anti-Hu (ANNA-1, Anti-neuronal Nuclear auto- Antibody, type 1)	Hu protein, basic RNA Binding protein (38 kDa) L: Cell nuclei of neurons In the central and peripheral nervous system	<b>FS</b> :Encephalitides with Focus in brain stem, cerebellum and limbic system;Myelitis;polyneuropathy (autonomous, sensory,Sensory-motor) <b>OS</b> : PCD, extrapyramidal Motor syndromes, LEMS, Focal epilepsy, POMA, Chronic gastrointestinal Pseudoobstruction, retinopathy	FT: SCLC, neuroblastoma OT: Prostate carcinoma, Bladder carcinoma, Ovarian carcinoma, Breast carcinoma, Pancreas carcinoma, Gastrointestinal malignant tumors
Anti-Ri (ANNA-2,Anti-neuronal Nuclear auto-Antibody, type 2)	NOVA, Ri/NOVA1,RNA binding Proteins (55 kDa and 80 kDa) L: Cell nuclei of neurons In the central nervous system	FS: POMA OS: PCD, rhombencephalitis	FT: SCLC, breast carcinoma OT: Hodgkin's lymphoma, Non-Hodgkin's lymphoma, Fast-growing brain stem tumor, ovarian carcinoma
Anti-Yo (PCA-1, Purkinje cell Antibody 1)	CDR2, CDR62, Yo antigen, signal transduction proteins (34 kDa and 62 kDa) L: Cytoplasm (rough ER, Golgi apparatus, cytoplasm membrane) of Purkinje cells of the cerebellum	FS: PCD OS: POMA	FT: Ovarian, breast, uterine carcinoma OT: Adenocarcinoma of the oesophagus,gall bladder carcinoma, prostate carcinoma, Hodgkin's lymphoma, Non-Hodgkin's lymphoma,thymoma
PCA-2	Purkinje cell protein (280 kDa)	FS: Encephalitis, neuropathy	FT: SCLC
Anti-Tr (PCA-Tr)	DNER (Delta/Notch-like Epidermal Growth Factor- Related Receptor)	FS: PCD	FT: Hodgkin's lymphoma
Anti-SOX1	SOX1 protein	FS: LEMS, PCD	FT: SCLC
Anti-PNMA1 (Ma1)	Ma protein (37 kDa)	<b>FS</b> : Rhombencephalitis (brain stem), limbic encephalitis	FT: Breast carcinoma OT: Various tumors
Anti-PNMA2 (Ma2/Ta)	Ma protein (40 kDa) L: Neuronal nucleoli	FS: Rhombencephalitis, PLE, brain stem Encephalopathy OS: POMA, PCD, focal epi- Lepsy, extrapyramidal motor Syndromes, retinopathy	FT: Germinal carcinoma of the Testis (seminoma) OT: Non-small-cell lung carcinoma, parotid gland carcinoma, breast carcinoma, ovarian carcinoma, colon carcinoma, kidney carcinoma,lymphoma
Anti-GAD	Glutamic acid decarboxylase (65 kDa and 67 kDa)	FS: SPS	FT: SCLC Breast carcinoma, Colon carcinoma
Anti- amphiphysin	Amphiphysin (128 kDa protein) L: Membrane of the Synaptic vesicles	FS: SPS OS: Myasthenic syndrome, Polyneuropathy(auto- Nomous, sensory, sensory- Motor), encephalomyelitis With rigidity, cerebellar	FT: Breast carcinoma, SCLC OT: Thymoma, Hodgkin's lymphoma, Colon carcinoma
Anti-CV-2 (Anti-CRMP5)	CV2/CRMP5 (66 kDa protein) L: Cytoplasm of oligo- dendrocytes	FS: Limbic encephalitis OS: PCD, polyneuropathy (autonomous, sensory, Sensory-motor), retinopathy, Uveitis, myasthenic Syndrome, chronic gastro- Intestinal pseudoobstruction, Chorea, POMA, romb- Encephalitis, focal epilepsy	FT: SCLC, thymoma OT: Uterine sarcoma
Anti-AQP-4 (NMO lgG)	Aquaporin-4 protein	Neuromyelitis optica (NMO), LETM, rec. ON	
Anti-NMDA receptors	Extracellular domain of The NR1 subunit of the receptor	Anti-glutamate receptor (type NMDA) encephalitis	Teratoma (ovary, testis)
Anti-AMPA Receptors (GluR1 and GluR2)	GluR1 and GluR2 Subunits of the receptor (each approx 100 kDa)	Limbic encephalitis	Breast carcinoma, thymoma, lung carcinoma

Anti-mGluR1	Metabotropic glutamate receptor (approx 140kDa)	Cerebellar degeneration	Hodgkin's lymphoma
GABAв receptors	Extracellular domain of The GABAB subunit of the receptor	Limbic encephalitis	SCLC
Anti-LG1	Protein (60 kDa) Associated with voltage- Gated potassium channels (VGKC)	Limbic encephalitis	SCLC, ovarian teratoma, thymoma, various tumors
Anti-CASPR2	Protein associated with Voltage-gated potassium Channels (VGKC)	Limbic encephalitis, neuro- Myotonia, Morvan's syndrome	Thymoma, uterine carcinoma
Anti-recoverin	Recoverin (23 kDa protein)	FS: Retinopathy	FT: SCLC
Anti-titin	Titin	FS: Myasthenia gravis	FT: Thymoma

## Flow chart for diagnosis of PNS

