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| **Supplementary Table 1. Cases reported with C1q deficiency and neuropsychiatric systemic lupus erythematosus** |
| **Age at onset/Sex/Flare** | **Country** | **Clinical features** | **NPSLE manifestation** | **Notes** | **Immunological tests** | **Complement functional tests** | **Neuroimaging** | **Medication** | **Mutation** | **Consequence/ Type C1q deficiency** | Ref. |
| 1/M/ND | Yougoslavia | Malar rash, oral ulcers, photosensitivity, arthritis, LN (MPGN), NPSLE (Seizure) | Seizure | Recurrent infectionsDied at13 | ANA + DNA+SSA +Sm+ RNP+ | C1q=0 | ND | Corticoids, frozen plasma, plasmapheresis and IVIG | g.8626C>T | Arg69X / Complete |  [17] |
| 13/F/2 | Saoudi Arabia | Malar rash, discoid rash, oral ulcers, arthritis, leukopenia, thrombopeina, alopecia | Seizures, mononeuritis multiplex | ND | ANA+ DNA-ENA- | C1q=0 | ND | Corticoids, CYC | ND | ND |  [18] |
| 7/F/7 and 20 | Dutch | Malar rash, oral ulcers, LN, fever, alopecia, lymphadenopathy, myositis  | Seizure, hemiplegia and lethargy. Probably cerebral vasculitis | Diedat 20 | ANA+DNA-RNP+ | C1q<0.1C1r/C1s=0C3/C4: NCH50<1%C1inh= N | Brain scintigraphy: multiple spots with activity mainly right sided, probably due to vasculitis | Corticoids, CYC | ND | ND / Dysfunctional |  [13] |
| 9/F/9 | Japan | Malar rash, discoid rash, photosensitivity, oral ulcers, proteinuria (no biopsy), arthralgia | Seizure |  | ANA+DNA –SSA +Sm+RNP + | NDLMW C1q | Calcification of the basal ganglia and the temporal lobe (CT-scan) | Corticoids | g.5499G>A | Gly34Arg / Dysfunctional |  [16] |
| 6/F/18,24 and 29 | Germany | Malar rash, oral ulcers, photosensitivity, leukopenia, pleuritis, arthritis, glomerulonephritis (Type V) and Libman-Sacks endocarditis, peritonitis | Seizure and psychosis | Renal and heart failure died at29 | ANA+DNA+Sm+ | C1q=28%C1r/C1s=NC2-C4f =NC3/C4= NCH50=0AP50=NC1inh =N | ND | Corticoids, plasmapheresis, chlorambucil, CYC, cyclosporin, IVIG | g.5499G>A | Gly34Arg / Dysfunctional |  [14] |
| 9/F/25 | England | Malar rash, photosensivity, leukopenia, alopecia | Seizure and cognitive dysfunction | Recurrent infectionsDied at 28 | ANA +DNA –SSA +Sm +RNP + | C1q=0CH50<5%AP50= N | Periventricular and basal ganglia calcification, with severe cerebral atrophy | Corticoids, Azathioprine, frozen plasma and plasmapheresis | g.8633delC | Gln71fsX137 / Complete |  [17]  |
| 5/F/ND | Saudi Arabian | Discoid lupus, photosensitivity, lupus nephritis (non-specified), alopecia | CNS involvement with cerebral atrophy, non-specified |  | ANA +DNA -SSA +SSB +Sm + | ND | ND. Cerebral atrophy | Unknown | ND | ND / dysfunctional |  [24] |
| 3/F/3 and 10 | Inuit | Malar rash, discoid rash, photosensitivity, oral ulcers | CNS involvement, non-specified | Died at 10 of Pneumocystis carinii pneumonia | ANA +DNA -Sm +RNP +RF + | C1q< 6%C1s/C1r= NC3/C4= NCH50 =1%AP50=NC1 inh= NMBL= N | ND | Corticoids, methotrexate,  | g.13166G>A | Gly244Arg / Complete |  [23] |
| 3/M/10 | Pakistan | Malar rash, oral ulcers, fever | Cerebral vasculitis (Encephalopathy with global dysphasia, quadra and bulbar paresis, generalized hypertoniaand resting tremor) | Bacterial meningitis at 3 | ANA +DNA - SSA +Sm + | C1q=0C2/C3/C4=N CH50=0AP50=N | Bilateral infarction of his basalganglia suggestive of a small vessel vasculitis | Corticoids, CYC, IVIG | ND | ND |  [21] |
| 7/F/7 | Arabian | Discoid rash, arthritis | Seizure, ACS, multiple ischemic lesions | Hyper IgM Syndrome Recurrent infections | ANA +DNA +SSA+SSB+ | C1q=5%CH50=0 | Multiple ischemic lesions involving both white matter and grey matter of hemisphere with left-sided predominance and also basal ganglia | Corticoids, CYC | g.5499G>A | Gly34Arg / Dysfunctional |  [20] |
| 6/F/6 and 15 | Pakistan | Malar rash, oral ulcers, alopecia, Raynaud, fever and arthralgia | Seizure, cerebral vasculitis | Recurrent infections | ANA+DNA-Sm+ | C1q=0 (ELISA)C3/C4= NCH50=0AP50=N | Left frontal lobe infarct secondary to cerebral vasculitis | Corticoids, Azathioprine, frozen plasma | g.5564delG | Gly55fsX83 / Complete |  [15] |
| 32/M/ND | Arabic | Thrombopenia, lymphopenia, AIHA, polymyositis | Seizure, transverse myelitis | Recurrent infectionsDied of bacterial septic shock and multi-organ failure | ANA+RNP+Ribosomal P +ACA + | C1q= NormalC2-C9=NCH50=0 | MRI brain and spine: brain atrophy, thoracic spinal cord atrophy | Corticoids, CYC | Codon 48 *Bchain* | Gly63Ser / dysfunctional |  [6] |
| 1/M/ND | Brasil | Photosensitivity, pericarditis | Psychosis and transverse myelitis | Cutaneous pyogenic infections and septic shock | ANA +Sm +RNP +  | C1q=0AntiC1q=0 | ND | ND | ND | ND |  [22] |
| 1/F/2 and 4 | Maltese | Malar rash, oral ulcers, fever, Raynaud, vasculitic lesions fingers | Seizure, cerebral vasculopathy/vasculitis with several strokes, encephalopathy associated with spasticity | Salmonella infection | ANA +DNA +Ribosomal P + | C1q=0CH50=0 | Bilateral frontal infarcts and basal ganglia calciﬁcation. Acquired moya-moya pattern with bilateral occlusive disease of the terminal segments of the internal carotid arteries and associated basal collaterals. Perfusion studies marked hypo-perfusion of the left hemisphere. | Corticoids, Azathioprine, MMF, CYC, frozen plasma | c.287del | Gly96Alafs / Complete |  [19] |
| 1/M/9 | Iraq | Malar rash, LN (Type II), fever | CNS involvement, non-specified (Lethargy, difficulty to walk) | Recurrent infectionsDead at 9, 4 months after allo-HSCT | ANA +SSA +RNP + | ND | MRI: Contrast enhancement in the left putamen in T1-weighted sequences. Enhanced signal in the right basal ganglia and capsula interna. | Rituximab, frozen plasma, plasma exchangeAllo-HSCT | ND | Gln208X / Complete | [5] |
| 1/M/24 | Dutch | Malar rash, photosensitivity, oral ulcers, arthritis, LN (Type V) | Cognitive dysfunction, CVD | Recurrent infections | ANA +DNA -SSA+Sm +RNP + | C1q=low 20%CH50=0AP50=NC3/C4=NMBL=NC3c and C5b9=N | MRI: Multifocal diffuse grey matter hyperintensities located in the fronto-temporal right lobe and high-intensity area on T2 in multiple regions of the right frontal and parietal lobes with high-intensities on the diffusion weighted imaging study. CT-angiography: no signs of cerebral vasculitis. | Corticoids, CYC | g.5499G>A | Gly34Arg / Dysfunctional | Present case |
| ACS: acute confusional state; CYC: cyclophosphamide; IVIG: intravenous immunoglobulin therapy; LN: lupus nephritis; MMF: mycophenolate mofetil; ND: non described; NPSLE: neuropsychiatric systemic lupus erythematosus |