Hypocomplemetemia in a patient with nephritis: cause or consequence?

Valeria Scaglioni, MD.
Rheumatology Unit. Internal Medicine Department
Hospital Italiano de Buenos Aires
Buenos Aires-Argentina
CASE PRESENTATION:

A 38-year-old female was admitted into our hospital because of nephrotic syndrome.

Her past medical history was unremarkable, she was in excellent health until one month before admission.

She started with mild asthenia and frothy urine.

Initial work up:

- Hb: 11.1
- WBC: 3700
- Lymphs: 851
- ANA: 1/1280
- dsDNA: 1/320
- C3: 29.2
- C4: <1

Normoalbuminemia, hyperlipidemia and abnormal urinalysis with red cast cells and proteinuria (4.5 g/24 hs).

Normal LFT, normal renal function, Levels of IgG, IgA and IgM were normal.
On day 1 a renal biopsy was performed, showing lupus nephritis (class IV G-A/C).

She was diagnosed with systemic lupus erythematosus and started on prednisone 60 mg/day.

She was diagnosed as having lupus psychosis and on day 8 received methylprednisolone 1000 mg/day IV for 3 days followed by CYC 1000 mg. After the first dose of methylprednisolone she had a complete recovery of her mental status.

On day 7 presented abnormal behavior, aggression and impaired judgment. An extensive work up showed a normal MRI/MRA, normal cerebral spinal fluid (CSF), normal chest XR and negative ID work up.

On day 12 she presented with seizures and persistent impaired judgment. No fever.

On day 15 she started with fever and severe alteration of mental status requiring ventilatory support.

On day 19 the patient died with multisystem organ failure due to disseminated pneumococcal infection.

The lumbar puncture showed a high pressure (100 cmH2O) with elevated WBC, abundant Gram-positive diplococci were seen on Gram stains of CSF. The brain CT showed cerebral edema, chest CT with an extensive infiltrate on the left lower lobe. CSF and blood cultures were positive for Streptococcus Pneumoniae.
In brief… patient with new onset lupus nephritis fulfilling the 2012 SLICC lupus criteria who presented with very low levels of complement along with an intense deposition of immune complexes in the renal tissue.

The patient had no family history of autoimmune diseases or primary immunodeficiencies.

Is it possible that the patient had an early complement component deficiency (C1, C2, C4, C3)? Did the patient have a SLE with secondary complement deficiencies?
Over the last 4 decades, stringent immunosuppression with glucocorticoids and cytotoxic drugs has dramatically improved survival rates of patients suffering from SLE.

Several intrinsic factors have been hypothesized to explain the high risk for infection in SLE, including immunoglobulin deficiency (in case of nephrotic syndrome), acquired or inherited complement deficiencies, defects in chemotaxis and phagocytosis, impaired clearance of opsonized bacteria by the reticuloendothelial system and functional asplenia.

THANK YOU