

57-Year Old SLE Patient With Evolving Symptomatology Dr. Shawn Macalester, DO

A 57-year-old African American female with SLE was under the care of Dr. Macalester and started complaining of **worsening symptoms**. The patient had a medical history of fatigue and idiopathic thrombocytopenia, which had been successfully treated using intravenous immune globulin (IVIG).

The patient also had a medical history of:

- Musculoskeletal pain in the large muscle groups, as well as pain in her palms and distal interphalangeal joints
- Multiple miscarriages
- Pleural and pericardial effusions
- Thoracentesis revealed increased white blood count, 26% neutrophils but gram stain and culture were negative
- An axillary lymph node biopsy, which was complicated by a staph infection, showed reactive changes
- Bone marrow biopsy showed hypercellularity consistent with an atypical myeloproliferative disorder, which was treated using prednisone; platelet count came up to 148,000
- Volume loss in both lung bases, prednisone dose was increased to 20 mg
- No history of blood clots

Previous labs revealed border-line positivity for IgG anticardiolipin and moderate positive for IgM anticardiolipin. Additional labs also revealed, the patient had ANA positivity at 1:1280 and anti-dsDNA positivity. Lab results were also positive for: anti-RNP, anti-SSA, anti-SCL-70 and anti-SMA. However, the patient had **consistently normal levels of soluble complement at every visit**.

The patient had been treated with a combination of 200 mg hydroxychloroquine (HCQ) and tapered doses of prednisone. She was later placed on azathioprine, with remarkable clinical improvement. Quinacrine was briefly added for treatment of a skin rash.



Since being treated by Dr. Macalester, the patient had been stable for over 4 years. To assess serological evidence of disease activity, Dr. Macalester ordered the AVISE SLE Monitor test and the AVISE HCQ test to help assess adherence to 200 mg of HCQ.

AVISE SLE Monitor Results

Assays	Results	Interpretation
+ EC4d- Erythrocyte-Bound C4d	44 Net MFI	POSITIVE
Complement C3	100 mg/dL	Normal
! Complement C4	9 mg/dL	ABNORMAL- LOW
Anti-dsDNA IgG	10 IU/mL	Negative
Anti-C1q IgG	4 Units	Negative
+ PC4d- Platelet-bound C4d	321 Net MFI	POSITIVE
Hydroxychloroquine	<30 ng/mL	Underexposed

Testing revealed EC4d and PC4d positivity as well as under exposure to HCQ. As a result, Dr. Macalester increased HCQ to 400 mg and reinforced the importance of adherence. The patient's history of multiple miscarriages, positive anti-SCL-70, EC4d and PC4d suggested the patient may have SLE and APS overlap. This alerted Dr. Macalester to monitor the patient more closely for risk of thrombosis.