

## CASE PRESENTATION

NS is a 45-year-old previously healthy, Jehovah's Witness female who developed confusion and difficulty expressing herself following a GI flu-like illness. Symptoms resolved spontaneously and reoccurred a week later with lack of coordination in her right hand and fine motor skills. Patient was admitted to a community hospital and underwent an MRI of the head, which showed multiple bilateral cortical small infarcts. Initial lab: Hgb 12.7, WBC 10.7 Platelets 15,000 and LDH 725. Patient received one dose each of: Rituxin, Citotoxan, Vincristine; and 5 doses of IgG. She also received Epoetin Alfa 20,000 units in 4 doses. Ten days later her hemoglobin dropped to 6.3 and the patient developed hemianopsia and was transferred to our hospital due to patient's refusal of blood transfusion, the need for albumin-pheresis and a higher level of care.

Second MRI of the head showed no changes from previous exam. Lab on arrival at #2 Hospital: Hgb 6.3 WBC 2 Platelets 87,000 and LDH 499. Patient was started on Methylprednisolone 125mg q 6h, Epoetin Alfa 40,000 units weekly and received Rituxin, Vincristine, 2 doses of IgG, 10-pks of cryoprecipitate and 3 albumin-pheresis and Depot leuprolide to stop menstrual periods. Ten days later, the patient developed chest pain and was transferred to the cardiac unit. The Troponin results were 14.6 and the Ck-MB 45.1. Hgb 7.5 WBC 5.8 Platelets 11,000 LDH 1,423. The patient then suffered an Acute Myocardial Infarction secondary to small arteriolar occlusion, which was managed medically. Resulting platelets levels dropped to 9,000, She was subsequently started on Cyclosporine 200mg BID and received one more dose of Rituxin and Vincristine, 40 pks of cryoprecipitate and 7 albumin-pheresis treatments were performed. Seven days later the patient complained of calf tenderness, mild erythema in her leg and developed a fever of 102.8F. The lower extremity venous duplex scan was negative and her lab values were: Hgb 5.5 WBC 0.9 Platelets 13,000 and LDH 427. Patient was placed in reverse isolation, with the fever attributed to neutropenia and cellulites. Patient was started on Vancomycin and Imipenem-cilastatin, the next day, resulting in WBC drop to 0.3 with a Hgb 4.7 and platelets 9. Blood cultures were positive for E. Coli. Patient received Filgrastim 480mcg daily for 7 days; another dose each of Rituxin and Vincristine and was started on Epoetin Alfa 20,000 units daily, Ferric Gluconate 125mg daily for 8 days and Cyclosporine was discontinued. Patient was tested for ADAMTS-13 inhibitor and activities with results were: inhibitor 0.4 and ADAMTS-13 activity 8% which were consistent with the diagnosis of Thrombotic Thrombocytopenic Purpura. One-week later, patient began to improve, resulting in Epoetin Alfa 20,000 units being adjusted to 3 times per week. Vancomycin and Imipenem-cilastatin were discontinued and substituted with Moxifloxacin. Patient was restarted on albuminpheresis 3 times per week. Two weeks later, patient developed a fever and the blood cultures came back positive for E. Coli. Previous IV antibiotics were restarted and the patient noticeably improved.

Albumin-pheresis was adjusted to once per week and the patient continued to improve. ADAMTS-13 test was repeated and the inhibitor was <0.4 and the activity 14%. After 54 days in the hospital and a total of 21 albumin-pheresis procedures, patient was discharged home. She continued with albumin-pheresis weekly for 3 more weeks, and her LDH at discharge was 481. A week later, following discharge, her labs were: WBC 9.7, Hgb 11.1 and platelets were 260. Two weeks later, patient was admitted to the hospital due to a community-acquired pneumonia that did not affect her TTP. Patient has been followed as an outpatient and has no reoccurrence of her TTP. Her last lab work showed: ADAMTS-13 inhibitor <0.4 and activity 80%, WBC 6.4, Hgb 13.4, Platelets 249 and LDH 490.

## CASE PRESENTATION

NS is a 45-year-old previously healthy, female who developed confusion and difficulty expressing herself following a GI flu-like illness. The initial flu symptoms were resolved spontaneously, however reoccurred one week later, along with lacking coordination in her hand accompanied with reduced fine motor skills reduction.

The patient was admitted to a community hospital, where an MRI of the head was performed, showing multiple bilateral cortical small infarcts. Initial labs: Hgb 12.7, WBC 10.7 Platelets 15,000 and LDH 725. Treatment initiated included one dose each of the following: Rituxin; Citotoxan; Vincristine; along with 5 doses of IgG, and 4 doses of 20,000 units of Epoetin Alfa per dose. Ten days later the patient was transferred to X Hospital following the identification of reduced hemoglobin of 6.3, and the onset of hemianopsia. The transfer decision was supported in part by the patient's refusal of blood transfusion, and the need for albumin-pheresis.

Labs on arrival at X Hospital (day 17): Hgb 6.3; WBC 2; Platelets 87,000; LDH 499. A second MRI was performed with no change from the previous exam. Medications: Methylprednisolone 125mg q 6h; Epoetin Alfa 40,000 units weekly; Rituxin, Vincristine; 2 doses of IgG; 10-pks of cryoprecipitate; 3 albumin-pheresis; depot leuprolide to end menstrual periods.

Ten days later (day 27), the patient developed chest pain and was relocated to the hospital cardiac unit. Labs: Troponin of 14.6; Ck-MB 45.1; Hgb 7.5; WBC 5.8; Platelets 11,000; and LDH 1,423.

It was discovered that the patient had suffered an acute myocardial infarction secondary to small arteriolar occlusion, and which was subsequently managed medically with the following treatment regimen: Cyclosporine 200mg BID was started as a result of platelets drop to 9,000; one additional dose each of Rituxin and Vincristine at the same levels; 40 pks of cryoprecipitate. Seven albumin-pheresis treatments were performed.

Seven days later (day 34) the patient complained of calf tenderness, mild leg erythema, with temperature of 102.8F. A lower extremity venous duplex scan was performed was negative. The elevated temperature was attributed to neutropenia and cellulites. Lab values: Hgb 5.5, WBC 0.9, platelets 13,000 and LDH 427. Treatment initiated: patient placed in reverse isolation, Vancomycin and Imipenem-cilastatin started on second day (day 29). Resulting Lab Values: WBC 0.3; Hgb 4.7; platelets 9; positive blood cultures for E. Coli; ADAMTS-13 inhibitor and activity: inhibitor 0.4; ADAMTS-13 activity 8%. Patient results were consistent with the diagnosis of Thrombotic Thrombocytopenic Purpura. Subsequent treatment initiated: Filgrastim 480mcg daily for 7 days; doses each of: Rituxin; Vincristine; Epoetin Alfa 20,000 units daily; Ferric Gluconate 125mg daily for 8 days. Cyclosporine was discontinued.

Seven days later (day 38), patient started to improve. Treatment regimen: Epoetin Alfa 20,000 units changed to 3 times per week; Vancomycin and Imipenem-cilastatin discontinued and substituted with Moxifloxacin; restarted on albumin-pheresis 3 times per week.

Symptoms two weeks later (day 52): patient developed a fever 101.2; blood cultures positive for E. Coli. Treatment regimen: IV antibiotics restarted. Patient response to treatment was noticeably improved. Treatment adjustment following improvement: Albumin-pheresis once a week, which resulted in continued improvement. Labs following improvement: ADAMTS-13 test, revealing inhibitor <0.4 and the activity 14%.

The patient was discharged home from the hospital on day 60, following 21 albumin-pheresis procedures. Discharge labs; LDH 481. Treatment following discharge: continuation of albumin-pheresis weekly for 3 weeks. Labs one week following discharge: WBC 9.7; Hgb 11.1; platelets 260.

Two weeks later (day 74), patient readmitted to the hospital due to a community-acquired pneumonia that did not affect her TTP; and following treatment was released in 12 days (day 80).

The final outcome of treatment for albumin pheresis: No reoccurrence of TTP 14 days after final release. Final labs performed after hospital release: ADAMTS-13 inhibitor <0.4, activity 80%; WBC 6.4; Hgb 13.4; Platelets 249; LDH 490.