Hemolysis, Elevated Liver Enzymes and Low Platelet (HELLP) Syndrome Associated with Systemic Lupus Erythematosus

Key words: pleuritis, anti-phospholipid antibody, pre-eclampsia, aspirin

A 29-year-old woman with no medical history had been conservatively treated at another hospital the 24th week of her gestation. The treatment consisted of restricted sodium intake for hypertension and proteinuria ascribable to pre-eclampsia. In the 29th week of her pregnancy she was admitted to the hospital because of sudden-onset nausea and abdominal pain. Laboratory data demonstrated elevated levels of aspartate aminotransferase (56 U/l, normal 12-37 U/l) and lactate dehydrogenase (690 U/l, normal 114-220 U/l) as well as anemia (hemoglobin 9.6 g/dl, normal 11.1-15.1 g/dl) and severe thrombocytopenia (3.0x10^9/mm^3, normal 13.2-36.8x10^9/mm^3). Anti-nuclear, anti-DNA and anti-cardiolipin IgG antibodies were positive, and anti-platelet antibody, negative. Blood smear specimens showed burr cells and schistocytes suggestive of hemolysis, and urinalysis indicated proteinuria. To relieve these symptoms cesarean section was performed two days after admission. She delivered a baby weighing 1,860 g without any signs of asphyxia at birth. Soon after the delivery she showed improvement in her symptoms and thrombocytopenia without the aid of corticosteroid therapy, and she was discharged from hospital.

Although liver enzymes and platelets examined at the outpatient clinic had returned to normal levels within one month after the delivery, anti-nuclear, anti-DNA and anti-cardiolipin IgG antibodies remained positive.

Since the delivery the present patient sometimes and transiently experienced erythematous eruptions in the bilateral cheeks as well as photosensitivity. At age 36 she was admitted to our hospital because severe pain had developed in the left lower chest with no precipitating cause. Physical examination and laboratory data demonstrated no abnormal findings except those for positive inflammatory reactions, including erythrocyte sedimentation rate (ESR, 59 mm/h, normal 3-11 mm/h) and CRP (1.82 mg/dl, normal level less than 0.1 mg/dl), while anti-nuclear and anti-DNA antibodies were >640 and 157 U/ml, respectively. Both anti-β2 glycoprotein I and anti-cardiolipin IgG antibodies were slightly positive. Chest X-ray examination and computed tomography (CT) demonstrated bilateral pleural effusion with predominant involvement of the left side (Fig. 1A), leading to a diagnosis of pleuritis as a cause of her chest pain. She was conservatively treated with antibiotics, and the chest pain quickly improved along with normalization of ESR and CRP. Pleural effusion was no longer seen on CT ten days after admission (Fig. 1B), and she was discharged and has been taking aspirin orally at a dose of 81 mg/day. Examinations at the outpatient clinic have demonstrated that she has since been in good general condition.

When this patient was admitted to the first hospital, hematological studies showed the presence of bicytopenia, while proteinuria and immunological abnormalities such as posi-

---

Figure 1. Chest computed tomography (CT) obtained at admission to our hospital demonstrates bilateral pleural effusion (arrowheads) predominantly on the left side (A). Pleural effusion is no longer seen on CT obtained around the time of discharge (B).
tive autoantibodies were also detected. These findings led to the diagnosis of systemic lupus erythematosus (SLE) in accordance with the established classification criteria (1). When acute thrombocytopenia accompanied by liver dysfunction is seen in SLE patients during pregnancy, four disorders should generally be considered as possible causes: hemophagocytic syndrome, thrombotic thrombocytopenic purpura, HELLP (hemolysis, elevated liver enzymes and low platelet) syndrome and exacerbation of SLE itself (2-4). Severe abdominal pain and nausea developed in the present patient without renal dysfunction or psychoneurological symptoms, laboratory data demonstrated hemolytic anemia with no decrease in complements, and improvement was evident soon after the delivery without the aid of corticosteroid therapy. These findings suggested that her clinical features were most compatible with those of HELLP syndrome, although SLE may also have played a role in the development of pleuritis. The precise etiology could not be identified, however, because of her quick recovery without the aid of corticosteroid therapy.

HELP syndrome is considered to be a severe form of pre-eclampsia ascribed to disseminated intravascular coagulation associated with microangiopathic hemolysis (5, 6). There are several risk factors for pre-eclampsia, and the HELLP syndrome of our patient was probably related to the presence of anti-phospholipid antibodies in addition to her being primigravida (5, 7). It is well known that anti-phospholipid antibodies are frequently associated with SLE as seen in this patient. When SLE patients are pregnant, especially those with positive anti-phospholipid antibodies, careful observation is necessary to detect development of pre-eclampsia, including HELLP syndrome, as early as possible. Low-dose aspirin is thought to reduce the frequency and severity of pre-eclampsia because of its inhibitory effects on thromboxan synthesis by platelets (2, 3, 8), and it is now administered to high-risk women as a prophylactic agent as it is in the case of primary anti-phospholipid antibody syndrome (3). Low-dose aspirin was also prescribed for the present patient in an effort to avoid unfavorable events induced by anti-phospholipid antibodies.

Acknowledgement: The authors are grateful to Dr. Penny J. Ballem, British Columbia’s Women’s Hospital & Health Centre, Vancouver, for providing us with clinical information regarding this patient.

Masayuki MATSUDA, Shigeaki MITSUHASHI, Megumi WATARAI, Kanji YAMAMOTO, Takao HASHIMOTO and Shu-ichi IKEDA

The Third Department of Medicine, Shinshu University School of Medicine, Matsumoto
Received for publication April 7, 2003; Accepted for publication August 7, 2003
Reprint requests should be addressed to Dr. Masayuki Matsuda, the Third Department of Medicine, Shinshu University School of Medicine, 3-1-1 Asahi, Matsumoto 390-8621

References