Recurrent Hemolytic Uremic Syndrome: Case Report

M.	Mietta	Meroni ^a
A.	Angela	Volpi ^a
G.	Graziana	Battini ^a
F.	Ferruccio	Conte ^a
G.	Gianmichele	Ferrario ^a
F.	Ferdinando	Giordano ^a
L.T.	Laura Torri	Tarelli ^b
A.	Adalberto	Tommasi ^a
A.	Adalberto	Sessa ^a

^aServizio di Nefrologia e Dialisi, Ospedale di Vimercate; ^bIstituto di Anatomia Umana Normale, Università degli Studi di Milano, Italia

Dr. M. Meroni, Servizio di Nefrologia e Dialisi, Ospedale di Vimercate, via C. Battisti 23, I-20059 Vimercate (Italy)

Dear Sir,

Hemolytic-uremic syndrome (HUS) is a disease of infants and children, but this condition may be found in adults too. Females are more commonly affected, and they usually show a history of oral contraception or recent delivery [1]. At present, however, a clear distinction between HUS and thrombotic thrombocytopenic pur-pura is quite difficult.

Recurrent attacks of HUS in adults have been seldom recorded, and the factors responsible for the recurrences were not always known [2].

We have observed a 52-year-old female with acute renal failure in course of classic manifestations of HUS: weakness, petechiae, and ecchymoses in the skin, hema-temesis, melena, and hematuria with mild proteinuria. Hemoglobin was 8.8 g/dl, reticulocytes 21%, white blood cells, 12,000/mm³, platelets 105,000/mm³, plasma fibri-nogen 140 mg/dl, fibrin split products 80 µg/ml, blood urea nitrogen 131 mg/dl, and serum creatinine 6.22 mg/dl.

The renal biopsy specimen has been studied by light, immunofluorescent, and electron microscopy and showed pathognomonic lesions of HUS (fig. 1).

The patient was treated with prednisolone pulses, hemodialysis, plasmapheresis and she left the hospital after 42 days. Serum creatinine was 1.36 mg/dl.

In coincidence with abrupt withdrawal of prednisolone, she developed a relapse of HUS with slight renal involvement (serum creatinine 1.79 mg/dl, blood urea nitrogen 32 mg/dl), but with severe involvement of skin, intestinal tract, myocardium, and brain. She died after 25 days in spite of prednisolone, plasmapheretic, and vitamin E treatment. At autopsy the pathologic findings were small microthrombi in brain, myocardium, kidney, thyroid, peritoneum, omentum, jejunum, and ileum.

Fig. 1. Electron micrograph from a biopsy specimen showing thickening of the capillary wall, effacement of foot processes, wrinkling of the basement membrane (asterisks), widening of the subendothelial space, which contains rarefied and granular material of variable electron density

(arrows), and swelling of the endothelial cells; CL = Capillary lumen; MC = mesangial cell; EC = endothelial cell; EpC = epithelial cell. Uranyl acetate and lead citrate. $\times 8,000$.

264

Meroni/Volpi/Battini/Conte/Ferrario/Giordano/TorriTarelli/Tommasi/Sessa

This female, at the age between 42 and 50 years, had four episodes of petechiae in the skin, bloody diarrhea, hematemesis, melena, with gastroscopic and sigmoidoscopic evidence of mucosal hemorrhagic lesions. She had been pregnant four times, though we are not capable of making a temporal correlation between HUS attacks and pregnancies.

We suppose that this patient may have suffered recurrent attacks of HUS with microangiopathic lesions each time more evident at different organs and apparatuses. The possibility of recurrences of HUS as far apart as many years is well known in some patients [2], sometimes with only minor manifestations and very few glomerular lesions [3].

microangiopathic hemolytic anemia (hemolytic-uremic syndrome): clinicopathologic study in 20 adults. *Kidney int.* 75:548 (1979).

Kaplan, B.S.: Hemolytic uremic syndrome with recurrent episodes: an important subset. *Clin. Nephrol.* 5:495 (1977). Levy, M.; Gagnadoux, M.F.; Habib, R.: Pathology of hemolytic uremic syndrome in children; in Remuzzi, Mecca, De Gaetano, Hemostasis, prostaglandins, and renal disease, p. 383 (Raven Press, New York 1980).

References

1 Morel-Maroger, L.; Kanfer, A.; Solez, K.; Sraer, J.D.: Prognostic importance of vascular lesions in acute renal failure with