Thrombotic thrombocytopenic purpura: report of seven cases

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From May 1999 to January 2002 we observed 7 patients (4 females and 3 males, median age 55 years, range 31-81 years) with thrombotic thrombocytopenic purpura (TTP). Six patients has been previously undiagnosed and 1 patient was at second relapse. Trigger factors of TTP were identified in 6 patients: ticlopidine treatment (2 patients); an acute cutaneous infection episode immediately before the features of TTP (1 patient); presence of devices: orthodontic (1 patient) and intrauterine contraceptive (1 patient), Mycoplasma urealyticum vaginal infection (1 patient). In all the 7 patients the clinical status was mainly related to the hemolytic anemia, thrombocytopenia and neurological events. One of these patients presented with hemolytic-uremic syndrome with acute renal failure and macrohematuria at onset, another one showed a systemic exanthema post-infection-like. Six out of 7 patients presented with different neurological events: headache, confusion, focal neurological failure. All the 7 patients were promptly treated with plasma-exchange and cryosupernatant plasma infusion. In addition they received prednisone 25-50 mg/day. All the 7 patients achieved a complete remission after plasma-exchange, one relapsed 3 months later and was treated with plasma-exchange again. All the patients are in complete remission with a median follow-up of 36.3 months (range 20-62 months). From these cases we suggest: 1) clinicians should take in mind the suspicion of TTP in every patient with hemolytic, negative direct Coombs test, anemia, thrombocytopenia, high level of lactate dehydrogenase; 2) the treatment of choice is plasma-exchange; 3) the response of treatment is good if therapy is promptly and aggressively administered; 4) the possible role of a trigger factor for removing it and to prevent relapses.