Portal vein thrombosis and Budd-Chiari syndrome as onset of Polycythaemia Vera

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Budd-Chiari syndrome may be defined as an heterogeneous group of vascular disorders characterized by obstruction of hepatic venous return to the level of hepatic venules, supra-hepatic veins, inferior vena cava or right atrium. The main cause of this syndrome is represented by myeloproliferative diseases, and, in particular, by polycythaemia vera. The latter may cause multiple splanchnic thrombosis, including portal vein thrombosis, particularly important, especially for its clinical outcomes (ascites, collateral vessels genesis, etc.). Here we reported two cases of a Budd-Chiari syndrome induced by polycythaemia vera characterized by an abnormal clinical onset, both as regards subject's age (29 and 39 years old, respectively) and set of symptoms, signs and laboratory data. After a complete clinical, instrumental and genetic diagnosis, the patients were treated with combined therapy, using acetylsalicylic acid and hydroxyurea. The therapy proved successful and patients are still in follow-up in our Institution. Polycythaemia vera should be suspected in patient affected with portal vein thrombosis and Budd-Chiari syndrome even if its clinical onset might be unusual. A correct and early diagnosis should be kept to start appropriate therapy as soon as possible and to prevent patients from useless diagnostic and therapeutic treatments.