Budd-Chiari Syndrome and Myeloproliferative Disorder

Budd-Chiari syndrome (BCS) is a rare clinicopathological condition characterized by occlusion of the major hepatic veins with or without membranous obstruction (MOVC) of the inferior vena cava (IVC) at its hepatic portion. Ascites, hepatomegaly and upper abdominal pain constitute the classical triad of symptoms, and the disease usually progresses to congenital liver cirrhosis and portal hypertension (1). Most of the Japanese cases of BCS are idiopathic with MOVC, and no associated etiology is demonstrable, whereas in Western countries, most patients have underlying thrombogenic conditions, such as myeloproliferative disorders (MPD), paroxysmal nocturnal hemoglobinuria, use of contraceptives, coagulation disorders, and others (1, 2).

It has been generally believed that MOVC in BCS is caused by a congenital vascular malformation. However, the recent pathological study by Kage et al (2) clearly supports the acquired theory which postulates “thrombosis” rather than a “congenital anomaly” as the cause. Prof. Kunio Okuda (3) strongly suggested in his 1982 editorial in Gastroenterology to a paper from South Africa (4) which reiterated the congenital malformation theory and frequent complicating hepatocellular carcinoma, that the pathogenesis of MOVC would be an acquired one for the following reasons: first, this is mainly an adult disease (age, 20–40 years in the majority) and the age of onset is totally incompatible with a congenital disease; second, none of the reported cases of MOVC in Japan had evidence of other congenital abnormalities; and lastly, the types of occlusion in IVC have a wide variety, which is also unusual if it were congenital in etiology. Based on his and other data, Okuda proposed a new classification of BCS, dividing it into two types (5). One is an “idiopathic” and the other a “secondary” type with an underlying etiology. The former often has a mild clinical presentation with a slow disease progression, probably due to an early development of collaterals that entails a relatively benign prognosis. Most of this type has MOVC. In contrast, the “secondary” type BCS often presents acutely with severe clinical manifestations that follow a sudden thrombotic obstruction of the major hepatic veins, and the prognosis is poor. There are fulminant cases. Thus, liver transplantation is frequently indicated in this type of BCS with which MOVC is rarely associated (5).

In this issue of INTERNAL MEDICINE, Usui et al describe a patient with BCS secondary to MPD (6). This case is interesting in several aspects. Although it is a rare cause of BCS in Japan, MPD is the most commonly associated etiologic disor-der in Western countries (1, 7, 8), where it is vitally important to make a diagnosis of MPD at its occult stage for the determination of the most appropriate treatment strategy and successful management after liver transplantation. Chromosomal studies and in vitro culture of bone marrow cells (9, 10) or detection of endogenous burst-forming units-erythroid (BFU-E) (11, 12) are believed to be useful in the diagnosis of latent MPD. In this patient, liver transplantation was performed in Australia because brain death is not legally recognized as a biological death in Japan. The first transplantation was a failure due to recurrence of thrombosis as a result of the hypercoagulable state of the patient, as previously experienced (13, 14), but retransplantation was a success as it was combined with the administration of hydroxyurea and anticoagulants, such as aspirin and warfarin, a well documented countermeasure in Western countries (14–18).

Although BCS without membranous obstruction of IVC secondary to MPD was rare in Japan in the past, it may increase in the future as disease epidemiology in Japan is slowly approaching that of Western countries. Therefore, the possibility of occult MPD should always be kept in mind as a possible cause when one sees a patient with BCS in Japan in the future. In such a case, liver transplantation followed by supportive medical management for MPD will prove remarkably effective.

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References

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