SINUSOIDAL OBSTRUCTION SYNDROME/VENO-OCCCLUSIVE DISEASE (SOS/VOD) is an unpredictable, potentially life-threatening complication of conditioning regimens for allogeneic or autologous hematopoietic stem cell transplantation (HSCT), or of chemotherapy alone. Its complex pathophysiology encompasses endothelial cell activation and damage (Carreras & Díaz-Ricart, 2011). Clinical characteristics of SOS/VOD include painful hepatomegaly, jaundice, fluid retention, rapid weight gain, and ascites (Bearman, 1995; DeLeve, Shulman, & McDonald, 2002; Kumar, DeLeve, Kamath, & Tefferi, 2003). Severe SOS/VOD, developing in about 20%–40% of SOS/VOD cases in patients receiving allogeneic transplantation, is typically characterized by the presence of multiorgan dysfunction (MOD), sometimes called multi-organ failure, involving renal and/or respiratory dysfunction, and may be associated with greater than 85% mortality (Coppell et al., 2010). Some mild cases may require only vigilance and supportive care; others can progress unpredictably, and a comprehensive response that includes pharmacotherapy is indicated, particularly for moderate to severe cases.

The incidence of SOS/VOD post-HSCT was estimated as 13.7% (range = 0%–62%) in a meta-analysis of 135 studies from 1979–2007 involving about 25,000 patients with HSCT (Coppell et al., 2010). Increasing use of reduced-intensity conditioning (RIC) regimens may have reduced SOS/VOD risk during recent years (Carreras et al., 2011); however, SOS/VOD occurs post-RIC, with one institution reporting an 8.8% incidence in patients receiving allogeneic transplantation (Tsirigotis et al., 2014). Regarding SOS/VOD after chemotherapy alone, incidence in one study was 11% (15 of 139 participants) (Kantarjian et al., 2016).

Pathophysiology

HSCT conditioning may trigger a potentially rapid pathophysiologic cascade leading to SOS/VOD. Toxic metabolites of conditioning agents may activate and damage endothelial cells lining hepatic sinusoids (Carreras & Díaz-Ricart, 2011). This activation leads to loss of vascular integrity, transformation of endothelial cells from antithrombotic to prothrombotic (Hunt & Jurd, 1998), and release of inflammatory cytokines (Carreras & Díaz-Ricart, 2011).