Meeting Report

Castleman-Kojima Disease (TAFRO Syndrome): A Novel Systemic Inflammatory Disease Characterized by a Constellation of Symptoms, Namely, Thrombocytopenia, Ascites (Anasarca), Microcytic Anemia, Myelofibrosis, Renal Dysfunction, and Organomegaly: A Status Report and Summary of Fukushima (6 June, 2012) and Nagoya Meetings (22 September, 2012)

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Recently, a unique clinicopathologic variant of multicentric Castleman's disease (MCD) has been identified in Japan. This disease is characterized by a constellation of symptoms, as listed in the title, and multiple lymphadenopathy of mild degree with a pathologic diagnosis of atypical CD, often posing diagnostic and therapeutic problems for pathologists and hematologists, respectively. These findings suggest that this disease represents a novel clinical entity belonging to systemic inflammatory disorders with a background of immunological abnormality beyond the ordinal spectrum of MCD. To define this disorder more clearly, Japanese participants presented clinicopathologic data at the Fukushima and Nagoya meetings. Many of the patients presented by the participants were significantly accompanied by a combination of thrombocytopenia, ascites (anasarca), pleural effusions, microcytic anemia, fever, myelofibrosis, renal dysfunction, and organomegaly (TAFRO). Multiple lymphadenopathies were generally of mild degree, less than 1.5 cm in diameter, and consistently featured the histopathology of mixed- or less hyaline vascular-type CD. Autoantibodies were often detected. However, this disease did not fulfill the diagnostic criteria for well-known autoimmune diseases including systemic lupus erythematosus. Castleman-Kojima disease and TAFRO syndrome (the favored clinical term) were proposed for this disease. The patients were sensitive to steroid and anti-interleukin-6 receptor antibody (tocilizumab), but some exhibited a deteriorated clinical course despite the treatment. The
participants proposed a future nationwide survey and a Japanese consortium to facilitate further clinical and therapeutic studies of this novel disease. [J Clin Exp Hematop 53(1): 57-61, 2013]

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TAFRO syndrome is a systemic inflammatory disorder characterized by a constellation of symptoms; thrombocytopenia with reticulin fibrosis of bone marrow, anasarca including pleural effusion and ascites, fever, renal dysfunction, and organomegaly (hepatosplenomegaly and lymphadenopathy). Although several histopathological features of TAFRO syndrome resemble those of mixed type of multicentric Castleman disease (MCD), some cases of TAFRO syndrome don't show any significant lymphadenopathy. In addition, several clinical and laboratory findings of TAFRO syndrome are different from those of MCD. Castleman-Kojima disease (TAFRO Syndrome) is a novel systemic inflammatory disorder characterized by a constellation of symptoms, namely, thrombocytopenia, anasarca, myelofibrosis, renal dysfunction and organomegaly, and multiple lymphadenopathy of mild degree with histopathology of mixed- or hyaline vascular-type Castleman's disease (CD). Molecular testing was negative for B-cell or T-cell monoclonality. The search for HHV8 and EBV was also negative. The histology pattern was considered not diagnostic for malignant lymphoma and related to a lymphadenopathy with features resembling multicentric Castleman's disease. The diagnosis was reviewed at the referral center of Bologna (Prof. Pileri) and confirmed.