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IMMUNE-RELATED PANCYTOPENIA : A RARE CASE REPORT IN SURABAYA, INDONESIA

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ABSTRACT : Immune related pancytopenia (IRP) is a peripheral pancytopenia caused by bone marrow failure due to an autoimmune process. A 41 year old female presented with gum bleeding 4 days prior to admission. She had anemic conjunctiva and no organomegaly. Laboratory results were as follows Hb: 7.9 g/dL, WBC: 3,200/ìL, PLT: 6,000/ìL, IPF: 4.7% and RET: 0.44%. Bone marrow aspiration was dominated by clusters of erythroblastic islands and activated macrophages. Confirmatory examination with bone marrow mononuclear cells coombs test (BMMNC Coombs Test) showed positive results. The patient received dexamethasone and trasnfusions with both packed red cells (PRC) and platelet concentrate (PC) apheresis. Mechanism of pancytopenia in patients is an autoimmune process caused by an autoantibody towards hematopoeitic cells in the bone marrow. B lymphocytes over produces this autoantibody. Ig G autoantibody also activates macrophages that will phagocyte hematopoeitic cells, whereas the Ig M autoantibody activates the complement system that causes the lysis of hematopoietic cells. The diagnosis of IRP in this patient was based on peripheral pancytopenia with an increase of Erythroblastic Island clusters and activated macrophages in the bone marrow. The BMMNC Coombs Test confirmed the diagnosis.

Key words : IRP, Erythroblastic Islands, BMMNC Coombs test.

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INTRODUCTION

In early 1998, there were patients presenting with pancytopenia that did not suit the diagnostic criteria for either hematological nor non-hematological diseases. Some patients responded well towards corticosteroids and high dose intravenous immunoglobulin therapy (He *et al*, 2001). The cause of pancytopenia in some patients were due to an autoimmune process that was caused by an autoantibody towards the membrane of hematopoietic cells of the bone marrow. autoantibodies were detected on the membrane of bone marrow hematopoietic cells by the bone marrow mononuclear cells (BMMNC) Coombs test, a modification of the peripheral blood coombs test, using BMMNC Coombs test to detect the binding of autoantibodies to bone marrow hematopoietic cells (BMHCs) (Bing *et al*, 2012; Xiao *et al*, 2020). At first, some of these patients were diagnosed as atypical aplastic anemia (AA), early stage AA, proliferative AA or chronic AA. In the 1980's, some patients were diagnosed as myelodysplastic syndrome (MDS) due to a bad response towards AA therapy. Due to the indication of the involvement of the body's immune system in the disease pathogenesis, these abnormalities were then called immune related pancytopenia (IRP), marked by cytopenia and a positive BMMNC Coombs test (Fu *et al*, 2014).

IRP cases are rarely reported and there is still limited data in China. Wang *et al* (2014) reported 81 cases of IRP in China and Liu *et al* (2012) also reported 20 cases in China. Patients characteristics were cytopenia or pancytopenia, normal or high reticulocyte count (RET), hypoplastic bone marrow or hyperplastic bone marrow with an increase of erythroblastic islands, and erythroid hyperplasia (Chasis, 2006; Manwani and Bieker, 2008;

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