## Introduction

Haemophagocytic lymphohistiocytosis (HLH) is a life-threatening condition characterized by uncontrolled immune system activation. Though it has been observed mostly among paediatric populations, it can affect patients of any age [1] HLH could be either primary (inherited) or secondary to a severe infection, malignancy, or rheumatologic condition [2]. Secondary HLH is also referred to as reactive haemophagocytic syndrome (RHS) [3]. Differentiating primary and secondary HLH has become increasingly difficult nowadays as new genetic causes are identified and patients who develop secondary forms of HLH are found to have some genetic aetiology as well [4]. Many elements of the immune system play important roles in the pathogenesis of HLH including macrophages, natural killer (NK) cells, cytotoxic lymphocytes and toll-like receptors (TLR). Macrophage activation syndrome (MAS) is a subset of HLH that occurs in patients with juvenile idiopathic arthritis (JIA) and other rheumatologic diseases [1]. The onset of MAS is usually acute. Patients develop sudden onset of non-remitting high-grade fever associated with hepatosplenomegaly, lymphadenopathy, cytopenia and liver transaminitis. Triglyceridaemia, elevated lactate dehydrogenase and hyponatraemia are also commonly