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Early Diagnostic Tools of Macrophage Activation Syndrome in Children with Systemic Juvenile Idiopathic Arthritis

Gargi R. Nair[1], Anupama .D[2], Harshamoorthi .C .S[3]

Vazhappally Koickal, Gandhi Junction, Kottarakkara, India
Ph. No-919497756676

Abstract: ***Macrophage activation syndrome (MAS) is a rare complication of childhood with rheumatic disease. It is potentially acute disorder. MAS is one of the leading cause of morbidity and mortality in patients with SJIA. It is characterized by persistent high fever, pancytopenia, purpura, encephalopathy, DIC.***

Keywords: macrophage activation syndrome, haemophagocytic lymphohistocytosis, systemic juvenile idiopathic arthritis, hyperferritinemia

1. Introduction

Macrophage activation syndrome (MAS) is a form of secondary haemophagocytic lymphohistocytosis that complicates rheumatologic disorders. It is one of the most potentially life threatening disorder in children with SJIA and SLE and to a lesser extent in adults; adult onset still disease. MAS is characterised by the dysregulation of the immune response leads to continuous T cell activation and macrophages finally results in multi organ impairment.

- A study conducted by Tellen. D. Bennett reported that organ system dysfunction is common in children with rheumatic disease associated by MAS.
- Rakesh Kumar et al outlined that MAS is an unusual and under recognised complication of Kawasaki disease.
- Mao mizuta et al are also found that overproduction of cytokines (TNF alpha, IL-18, TNF alpha) might be closely related to the development of MAS

Table 1: Clinical and laboratory features of MAS

System	Clinical features	Laboratory features
General	Fever	Fall in esr
Haematological	Coagaulopathy petechiae purpura	Extreme hyperferritinemia, leucopoenia, anaemia, thrombocytopenia
CNS	Seizures Encephalopathy Coma	Csf pleiocytosis
Gastrointestinal	Rectal bleeding Liver dysfunction	Hypoalbuminemia Elevated triglycerides Mildly elevated bilirubin
Renal respiratory	ARD Pulmonary infiltrates	Abnormal renal function

2. Materials and Methods

Retrospectively reviewed 391 cases that have clinical and laboratory evidence of MAS associated with SJIA. A patient having SJIA is suspected to be having MAS if the given diagnostic criterion is following

Ferritin >684 ng/ml and any 2 of the following
Platelet count $\leq$181x 10^9/litre
Aspartate aminotransferase>48 units/litre
Triglycerides>156 mg/dl
Fibrinogen $\leq$ 360 mg/dl

As per the data obtained the laboratory findings available are ferritin, platelet count, aspartate aminotransferase, triglycerides, fibrinogen, WBC count, Hb, D-dimer, ESR, C reactive protein.

3. Result & Discussion

There are so many factors suggestive of MAS. The ferritin level in blood is considered as a highly sensitive and specific tool for MAS. Out of 391 patients choosen 334 were having hyperferritenimia (684ng/ml). Platelet count shows a marked decrease in 98% of the cases. Aspartate aminotransferase and triglycerides levels seems to be elevated in most of the cases. A fibrinogen and ESR level shows a considerable reduction in 80% of cases, where as CRP levels remains elevated. All the above diagnostic tests except CRP Were having a p value <0.0001, shows the statistically significant correlation

Laboratory features	No. of patients with available data	P value
Ferritin	334	<0.0001
Platelet count	385	<0.0001
Aspartate aminotransferase	377	<0.0001
Triglycerides	272	<0.0001
Fibrinogen	314	<0.0001
D dimer	142	<0.0001
Esr	335	<0.0001
LDH	329	<0.0001
C reactive protein	367	0.0501

4. Conclusion

Early diagnosis of MAS plays an important in management & care of SJIA. A single confirmatory test for the diagnosis of MAS is not yet available. But it is a group of parameters contributes to the final diagnosis of the above. They are

Ferritin, Platelet count, Fibrinogen, Aspartate aminotransferase, triglycerides. Among these parameters ferritin play a key role. Hyperferritnemia is associated with 98% of MAS associated SJIA. Apart from the above described parameters, D-dimer and LDH Levels were seems to be elevated in some of the cases.

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