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Original Article

Ferritin/erythrocyte sedimentation rate ratio in macrophage activation syndrome in patients with systemic juvenile idiopathic arthritis: A single center experience

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ABSTRACT

Objective: Systemic juvenile idiopathic arthritis (JIA) is a subgroup of JIA that can involve multiple systems other than the joints. Macrophage activation syndrome (MAS) is a life-threatening condition that can be observed in patients with systemic JIA. The aim of this study was to evaluate the performance of the ferritin/erythrocyte sedimentation rate (ESR) ratio, a diagnostic parameter for MAS, in our cohort.

Materials and Methods: Children with systemic JIA who were followed in our hospital's pediatric rheumatology department and diagnosed with MAS were retrospectively recorded for the presence or absence of MAS. The performance of the ferritin/ESR ratio between these patient groups was analyzed.

Results: The study included 45 patients with systemic JIA without MAS and 11 patients with systemic JIA with MAS. No differences were observed between the two groups in terms of age, gender, and follow-up duration ($p = 0.121$, $p = 0.547$, and $p = 0.495$, respectively). The JADAS-27 score, which includes rash, hepatomegaly, splenomegaly, and disease activity score, was significantly higher in the systemic JIA group with MAS (all $p<0.050$). The ferritin/ESR ratio was significantly higher in systemic JIA patients who developed MAS [247.71 (36.23-886.66) vs. 31.47 (0.39-555.55); $p<0.001$]. A ferritin/ESR ratio of >38.84 had 100% sensitivity and 73.3% specificity for the development of MAS in patients with systemic JIA.

Conclusion: A ferritin/ESR ratio of (>)38.84 had high sensitivity and specificity for predicting the development of MAS in patients with systemic JIA in our cohort.

Keywords: Erythrocyte sedimentation rate, ferritin, macrophage activation syndrome

Introduction

Systemic juvenile idiopathic arthritis (sJIA) is a distinct category of JIA defined by the International League of Associations for Rheumatology (ILAR) criteria, characterized by quotidian fevers, arthritis, and extra-articular manifestations such as lymphadenopathy, serositis, hepatosplenomegaly, and an evanescent rash (1, 2). A severe and life-threatening complication of sJIA is macrophage activation syndrome (MAS), a form of secondary hemophagocytic lymphohistiocytosis (HLH). MAS arises from a dysregulated immune response involving uncontrolled T-lymphocyte and macrophage activation, culminating in a profound cytokine storm. Precipitating factors include infections or modifications to immunomodulatory therapy (3, 4). While the documented prevalence of overt MAS in sJIA is 10–24%, subclinical manifestations may be present in 30–40% of patients (5). The clinical presentation of MAS includes persistent fever, hepatosplenomegaly, lymphadenopathy, hemorrhagic diathesis, and central nervous system (CNS) dysfunction (6-8). Characteristic laboratory findings comprise hyperferritinemia, cytopenias, elevated liver transaminases, hypofibrinogenemia, hypertriglyceridemia, coagulopathy, and a paradoxically decreased erythrocyte sedimentation rate (ESR) (4). Although MAS is most commonly associated with sJIA, it can also complicate other inflammatory conditions

such as juvenile systemic lupus erythematosus and Kawasaki disease (4, 9). While often appearing spontaneously or in response to triggers, a genetic predisposition is implicated in cases of recurrent MAS (4, 6).

Various diagnostic guidelines and methods have been proposed to better differentiate MAS from other diagnoses; these include the HLH-2004 criteria (originally for familial HLH), the 2016 EULAR/ACR/PRINTO classification criteria for MAS in sJIA, the more recent MS score, and the ferritin-ESR ratio (5, 10-12). The MS score, developed by Minoia et al., helps distinguish MAS from an sJIA disease flare, and this score includes assessment of the central nervous system involvement, hemorrhagic manifestations, active arthritis, platelet count, plasma levels of lactate dehydrogenase, fibrinogen and ferritin (11). Concurrently, Eloseily et al. (12) proposed the ferritin/ESR ratio, leveraging the observation that ferritin rises dramatically in MAS while ESR falls due to fibrinogen consumption, creating a high ratio indicative of MAS. Differentiating MAS from active sJIA is clinically challenging due to overlapping features, yet it is critical as early, aggressive treatment is essential for survival.

This study aimed to evaluate the performance of the 2016 MAS classification criteria for the early identification of MAS in a single-center cohort of sJIA patients and to compare its diagnostic utility with other available tool, including the ferritin/ESR ratio.

Materials and Methods

This retrospective study was conducted on 56 children diagnosed with sJIA according to ILAR criteria, who were followed at the Pediatric Rheumatology Clinic of Ankara Bilkent City hospital between August 2019 and September 2025. Demographic and clinical data were extracted from electronic medical records, including demographic details (gender, date of birth, age at sJIA onset, age at MAS onset) and clinical manifestations (fever, hepatosplenomegaly, hemorrhagic manifestations, lymphadenopathy, active arthritis, CNS involvement, serositis). A comprehensive laboratory workup was performed, including ESR, C-reactive protein (CRP), complete blood count, aspartate aminotransferase (AST), alanine aminotransferase (ALT), fibrinogen, D-dimer, ferritin, triglycerides, and immunologic parameters (antinuclear antibodies and rheumatoid factor). Patients were stratified into two groups based on the 2016 classification criteria: “sJIA with MAS” and “sJIA without MAS.” MAS 2016 criteria include fever and ferritin > 684 µg/L plus 2 of 4 criteria: platelets 181×10^9/L, AST > 48 U/L, triglycerides > 156 mg/dl, and fibrinogen ≤360 µmol/L. Ferritin/ESR ratio 21:5 considered as a screening tool for the diagnosis of MAS.

The final version of the Juvenil Arthritis Disease Activity Score (JADAS) included 4 measures; physician global assessment of disease activity 10-cm visual analog scale (VAS) where 0= no activity and 10= maximum activity; parent/patient global assessment of well-being, measured on a 10-cm VAS where 0= very well and 10 = very poor; count of joints with active disease; and erythrocyte sedimentation rate. JADAS-27 includes the following joints: cervical, spine, elbows, wrists, metacarpophalangeal joints (from first to third), proximal interphalangeal joints, hips, knees, and ankles 1.

The ferritin/ESR ratio was calculated using the first available ferritin and coincident ESR values upon hospital admission.

Statistical analysis

Statistical analyses were performed using IBM Statistical Package for the Social Sciences, version 20.0 (SPSS Inc., Armonk, NY, IBM Corp., USA). Descriptive data are presented as requency, percentage, median (interquartile range), or mean and standard deviation. Normality of continuous variables was assessed visually (histograms, probability plots) and analytically (Kolmogorov-Smirnov test). Group comparisons for categorical variables used the Chi-square or Fisher’s exact test, while the Student’s T-test or Mann-Whitney U test was used for continuous variables, as appropriate. The sensitivity and specificity of the 2016 criteria were analyzed using Pearson’s chi-square test. Receiver operating characteristic (ROC) curve analysis was employed to evaluate the diagnostic accuracy of the ferritin/ESR ratio and to determine an optimal cut-off value for our cohort using the Youden index. Area under the curve (AUC) values > 0.8 were considered indicative of good diagnostic performance. Sensitity and specificity values were derived from the ROC curve analysis, while group comparisons were prformed using Pearson’s chi-square test. A p-value < 0.050 was considered statistically significant, with a 95% confidence interval (CI).

Results

A total of 56 children with sJIA were included into this retrospective, cross-sectional study included 11 children with sJIA accompanied by MAS and 45 children with sJIA without MAS. There were no differences in age, follow-up duration, or gender between the two groups (p=0.121, p=0.547, and p = 0.495, respectively). Rash was the most common clinical finding in the MAS group (11/11, 100%, p=0.042), while hepatomegaly and splenomegaly were significantly more common than in the children with sJIA without MAS (p=0.026 and p=0.002, respectively). The disease courses (monocyclic, polycyclic, etc.) of both groups were similar (p=0.639). However, the JADAS-27 score was significantly higher in children with MAS compared to without (35.92±9.15 vs. 30.63±6.61; p=0.032). When the 2016 MAS classification criteria were examined, ferritin vs. triglyceride values were significantly higher in children with MAS (3666.55±1830.13 µg/L vs.1655.68±1112.38 µg/L, p<0.001; 234.36±163.67 mg/dl vs. 142.21±101.87 mg/dl, p=0.022, respectively). Platelet counts were significantly lower in children with MAS (282.7±195.2 $x10^9$/L vs. 484.5±231.9 $x10^9$/L, p=0.010). No significant difference was observed in fibrinogen and AST values (p=0.523 vs. p=0.399, respectively). The ferritin/ESR ratio, which is among the MAS diagnostic criteria, was significantly higher in sJIA patients with MAS than

Table I: Demographic characteristics and laboratory data in children with active SJIA with and without MAS according to the 2016 MAS classification criteria.

	MAS (n=11)	Non-MAS (n=45)	p
Clinical findings			
Age (months)*	61.8±55.72	90.46±55.18	0.121
Duration of follow up days*	1292.36±1014.70	1307.96±1583.29	0.495
Gender (f/m)	6/5	20/25	0.547
Fever†	11 (100)	45 (100)	1.000
Rash	11 (100)	32 (71.1)	0.042
Hepatomegaly†	8 (72.7)	16 (35.5)	0.026
Splenomegaly†	9 (81.8)	14 (31.1)	0.002
Arthritis†	9 (81.8)	36 (80)	0.892
Lymphadenopathy†	8 (72.7)	19 (42.2)	0.070
Oligoarticular presentation†	4 (36.3)	22 (48.8)	0.455
Poligoarticular presentation†	5 (45.4)	14 (21.1)	0.368
Serositis†	4 (36.3)	6 (13.3)	0.074
Pleural effusion†	3 (27.2)	3 (6.6)	0.048
Pericardial effusion†	2 (18.2)	3 (6.6)	0.230
Peritoneal effusion†	0 (0)	2 (4.4)	0.476
Uveitis†	0 (0)	2 (4.4)	0.476
Disease course†			
Monocyclic	6 (54.5)	21 (46.7)	0.639
Polycyclic	0 (0)	8 (17.8)	
Polyarticular	5 (45.5)	16 (35.5)	
JADAS-27 score*	35.92±9.15	30.63±6.61	0.032
Laboratory findings			
Ferritin µg/L*	3666.55±1830.13	1655.68±1112.38	<0.001
Platelets*	282727.27±195210.70	484488.9±231894.63	0.010
Fibrinogen g/L*	4.32±2.44	5.99±3.12	0.523
Triglyserides mmol/L*	234.36±163.67	142.21±101.87	0.022
AST U/L*	60.73±28.44	46.49±25.66	0.399
LDH U/L*	777.55±489.04	425.22±264.58	0.002
WBC x10^9/L*	20160.91±14560.97	14445.33±6327.90	0.050
Neutrophils x10^9/L*	14678.18±13615.86	10509.33±6202.63	0.312
Lymphocytes x10^9/L*	2929.09±2131.57	2503.56±1311.09	0.402
ESR mm/hr *	72.45±30.42	66.36±33.50	0.584
CRP mg/L*	86.47±65.96	93.35±66.76	0.760
Hb gr/dL*	9.89±2.37	10.43±1.38	0.106
Ferritin/ESR ratio‡	247.71 (36.23-886.66)	31.47 (0.39-555.55)	<0.001

***:** *mean±SD*,**†:** *n(%)*, **‡:** *median (min-max)*, **ALT:** *Alanine aminotransferase*, **LDH:** *lactate dehydrogenase*, **WBC:** *white blood cells*, **ESR:** *erythrocyte sedimentation rate*, **CRP:** *C-reactive protein*, **Hb:** *hemoglobin.*

in those without MAS [247.71 (36.23-886.66) vs. 31.47 (0.39-555.55), p<0.001] (Table I).

As shown in Table II, when the 2016 MAS classification criteria were analyzed individually within our cohort, ferritin >684 µg/L had a sensitivity of 100% and a specificity of 66.7%, with a negative predictive value (NPV) of 100%. When examined for PLT ≤181 × 109/L, it had a sensitivity of 45.5% and a specificity of 91.1%, with a negative predictive value (NPV) of 87.2%. Fibrinogen ≤10.6 g/L had a sensitivity of 100% and a specificity of 2.2%, with a negative predictive value (NPV) of 100%.

In the ROC analysis performed for the ferritin/ESR ratio in our cohort for the prediction of MAS defined by the 2016 MAS classification criteria, the ferritin/ESR ratio > 38.84 (area under curve [AUC]: 0.917, [95% CI (0.84-0.99)], p<0.001) with a 100% sensitivity and 73.3% specificity in MAS (Figure 1). Additionally, as shown in Table III, a ferritin/ESR ratio cut-off point of >38.84 had a positive predictive value of 50% and a negative predictive value of 97.2%.

Discussion

The absence of a gold standard for diagnosing sJIA-associated MAS underscores the importance of validated diagnostic tools. The 2016 MAS classification criteria and the ferritin/ESR ratio were both derived from large multinational cohorts. This study represents a single-center, external validation of two different approaches for the early detection of MAS in sJIA.

Kostik et al. (14), found comparable rates of lymphadenopathy between MAS and non-MAS patients. In contrast, a larger study by Kelly et al. (15) reported significantly higher frequencies of hepatosplenomegaly, lymphadenopathy, CNS

Table II: Sensitivity and specificity of components of 2016 MAS classification criteria

	Cut off	MAS*	Non-MAS*	p†	Sensitivity	Specificity	PPV	NPV
Ferritin µg/L	> 684	11 (100)	15 (33.3)	<0.001	100	66.7	42.3	100
PLT×10⁹/L	≤ 181	5 (83.3)	4 (8.9)	0.452	45.5	91.1	55.6	87.2
Fibrinogen g/L	≤ 10. 6	11 (100)	44 (97.8)	0.618	100	2.2	20.0	100
Triglycerides mg/dl	> 156	5 (83.3)	15 (33.3)	0.003	45.5	66.7	25	83.3
AST U/L	> 48	5 (83.3)	10 (22.2)	0.119	45.5	77.8	33.3	85.4

: n (%), †: Pearson chi-square test, **PLT: platelets, **AST:** alanine aminotransferase, **PPV:** Positive predictive value, **NPV:** Negative predictive value*

Table III: The performances of Ferritin/ ESR ratio for diagnosing macrophage activation syndrome (MAS) in systemic juvenile idiopathic arthritis (sJIA) patients

	Cut off	Sensitivity	Specificity	PPV	NPV
Ferritin/ESR	≥21.5	100	68.9	44	100
	>38.84	100	73.3	50	97.2

***ESR:** Erythrocyte sedimentation rate, **PPV:** Positive predictive value, **NPV:** Negative predictive value*

dysfunction, and hemorrhage in MAS patients). In our study, rash, hepatomegaly, and splenomegaly were observed more frequently in children with sJIA who developed MAS, while lymphadenopathy was observed at similar rates in children with sJIA who developed MAS, unlike previous studies.

The hallmark laboratory features of MAS are a sharp decline in platelet count and ESR, coupled with elevated ferritin, triglycerides, and lactate dehydrogenase (LDH) (15,16). Hoeg et al. (17) who found significantly higher levels of ferritin, triglycerides, ALT, and LDH in patients with MAS compared to the group without MAS. In addition, the levels of platelets and fibrinogen were significantly lower in MAS patients. Kostik et al. (14) found similar differences in patients with active sJIA with and without MAS. Zeng et al. (18) collected clinical and laboratory data from thirteen sJIA patients with MAS and found elevated levels of triglycerides, ALT, and LDH. In our cohort, children who developed MAS had significantly higher ferritin, triglyceride, and LDH levels, while platelet levels were significantly lower. No significant changes were observed in AST and fibrinogen levels, which are components of the 2016 MAS criteria. According to similar studies, the differing results may depend on factors such as early disease stage, sampling timing, and small sample size.

Although a ferritin level >684 µg/L is a mandatory criterion in the 2016 guidelines, its specificity has been questioned. Hoeg et al. (17) revealed that 56% of non-MAS patients had ferritin levels above this threshold. In our cohort, ferritin levels >684 µg/L were present in 100% of children with MAS, compared with one-third of children with sJIA who did not develop MAS. This had a sensitivity of 66.7%, a specificity of 100%, and a negative predictive value of 100%.

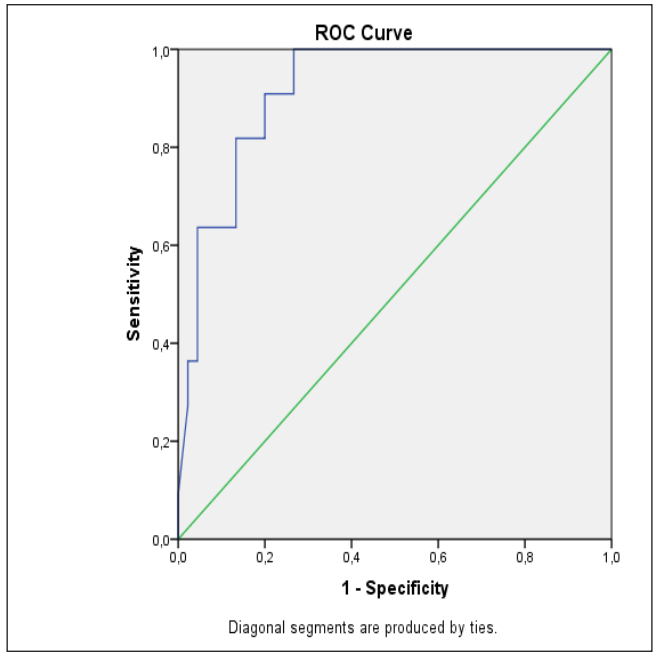

Figure 1: Receiver operating characteristics (ROC) plots illustrating the area under the curve (AUC) model for the prediction of MAS defined by the 2016 MAS classification criteria.

Individual laboratory parameters showed high specificity for MAS. A platelet count ≤181×10⁹/L and AST >48 U/L demonstrated specificities of 91.1% and 77.8%, respectively, identifying them as potentially useful markers for distinguishing MAS from an sJIA flare in hyperferritinemic patients. Since AST, ALT, and triglycerides in general are normal in patients with sJIA, a simple increase above the upper limits of these parameters in combination with other criteria and clinical signs may be adequate to indicate the development of MAS (4). This aligns with Ravelli et al.'s (19) earlier finding that thrombocytopenia (≤ 262×10⁹/L) is a highly sensitive (100%) and specific (92%) indicator of MAS. For the patients in our study, the 2016 MAS classification criteria of PLT ≤ 181×10⁹/L had a specificity of 91.1% and an NPV of 87.2%. Another criterion, AST > 48 U/L, had a specificity of 77.8% and an NPV of 85.4% in children who developed MAS.

The optimal hemophagocytic syndrome diagnostic score (HScore) cut-off value was proposed as 190.5, revealing a sensitivity of 96.7% and specificity of 98.4% (20). MS-score is a new tool for differentiating MAS from disease activation

in sJIA patients. This score also has weighted values for each parameter, and patients with a score above -2.1 points were defined as having MAS (11).

The ferritin/ESR ratio demonstrated the highest sensitivity but lower specificity among the MAS diagnostic tools evaluated. Hoeg et al. (17) showed that the best cut-off value of the ferritin/ESR ratio was ≥19.4. This is compatible with the cut-off value of ≥21.5 in the original paper by Eloseily et al. (12). In our study, a ferritin/ESR ratio cut-off point of ≥21.5, as documented by Eloseily et al. (12) had 100% sensitivity, 68.9% specificity, and 100% NPV. In our cohort, a ferritin/ESR ratio cut-off point of >38.84, which we found after ROC analysis, had similar 197.2% sensitivity, 73.3% specificity, and 100% NPV (area under curve [AUC]: 0.917, [95% CI (0.84-0.99)], p<0.001).

Limitations

This study has several limitations. Its single-center, retrospective design and small sample size limit statistical power and generalizability. The absence of some laboratory data, particularly from patients transferred from other institutions, may have introduced bias. Furthermore, using the 2016 classification criteria as the reference standard may have missed subclinical MAS forms, potentially leading to an underestimation of the tools' diagnostic capabilities. One of the most important limitations of our study is that the necessary validation analyses for these criteria, such as the ferritin/ESR ratio, could not be performed because the HLH criteria and MS score criteria components, which are among the other diagnostic criteria, could not be obtained retrospectively.

Conclusion

The ferritin/ESR ratio is a valuable, readily available tool to differentiate MAS from active sJIA. In our cohort, a cut-off value of ≥38.84 provided an optimal balance of sensitivity and specificity. To refine and validate these diagnostic scoring systems, large-scale, prospective, multi-center studies with comprehensive data collection are essential which may facilitate earlier recognition and treatment of MAS in children with sJIA.

Ethics committee approval

This study was conducted in accordance with the Helsinki Declaration Principles. The study was approved by Ankara Bilkent City Hospital (08.10.2025, reference number: 25-1771).

Contribution of the authors

Conceptualization: ŞE, BÇA; Methodology: ŞE, BÇA, EE, Formal analysis and investigation: EÖ, MIE, SNY, DÖ, YUE, ŞET; Writing - original draft preparation: ŞE, ,CK, BÇA; Writing - review and editing: ŞE, BÇA; Supervision: BÇA

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Conflict of interest

The authors declare that there is no conflict of interest.

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