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CASE BASED REVIEWS

Emapalumab's role in a severe and treatment-resistant paediatric macrophage activation syndrome

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ABSTRACT

Introduction: Macrophage activation syndrome (MAS) is a life-threatening hyperinflammatory condition. Emapalumab, an IFNγ-directed antibody, is approved for use in the USA but not in Europe.
Case: A 15-year-old girl presented with fever, odynophagia and a transient rash. After 9 days of hospitalization under empirical antibiotics, she developed pancytopenia, hypofibrinogenaemia, elevated ALT, AST, LDH, triglycerides, soluble CD25, serum calprotectin and ferritin (peak 357,976 ng/ml), and hepatosplenomegaly. Infectious and immune workups were negative, and bone biopsy confirmed haemophagocytosis. MAS was diagnosed, which was complicated by acute respiratory distress and supraventricular tachycardia. High-dose corticosteroids, anakinra and ciclosporin were initiated, with transient improvement. Subsequent drug-induced hepatotoxicity and microangiopathy, and infections worsened her condition. Given refractoriness to standard therapy, emapalumab was started under compassionate use, leading to sustained clinical and laboratory remission. She was discharged and remains stable at six-month follow-up, off corticosteroids and on canakinumab maintenance.
Discussion: This case illustrates the challenges of treating severe, refractory MAS. Emapalumab, used for the first time in Portugal, was well tolerated and associated with complete and sustained remission after failure of multiple therapeutic lines.

Keywords: Adolescent rheumatology; Rheumatology; Paediatric/Juvenile rheumatology; Juvenile idiopathic arthritis; Macrophage activation syndrome.

INTRODUCTION

Macrophage activation syndrome (MAS), a form of secondary haemophagocytic lymphohistiocytosis (HLH), is a severe and potentially life-threatening complication of rheumatic diseases[1,2]. It is characterised by excessive activation and proliferation of T lymphocytes and macrophages, leading to cytokine overproduction, systemic hyperinflammation, and haemophagocytosis[1,2]. MAS is most commonly associated with Still's disease (SD), occurring in approximately 15–20% of patients[2]. Although it has an estimated mortality rate of 10%, early recognition and treatment significantly improve survival rates[1,3].

Management of MAS involves immunomodulatory therapy, typically high-dose corticosteroids and anakinra, an interleukin-1 (IL-1) receptor antagonist. Additional therapeutic options include ciclosporin, intravenous immunoglobulin (IVIg), and Janus kinase (JAK) inhibitors[1,2]. In addition to MAS-directed therapy, management must also address contributing factors, such as infections[1].

Emapalumab is an anti-interferon-γ (IFNγ) monoclonal antibody. It has been approved by the United States Food and Drug Administration (FDA) for the treatment of refractory, progressive, or recurrent primary HLH and was recently approved for MAS[4,5]. However, the European Medicines Agency (EMA) has not approve its use[6]. Nevertheless, recent evidence supports that emapalumab is a safe and effective option for corticosteroid-resistant MAS cases[7–13].

This report describes a case of severe and prolonged MAS as the inaugural manifestation of SD, refractory to conventional treatments, which achieved remission with emapalumab. It underscores the diagnostic complexities of MAS and highlights the therapeutic potential of IFNγ inhibition in severe, treatment-resistant disease.

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CASE DESCRIPTION

A 15-year-old girl, with a medical history of obesity (BMI 32kg/m^2), presented to the emergency department with a five-day history of odynophagia, fever (maximum temperature 39.5ºC), asthenia and myalgia. The fever spikes were accompanied by a transient, non-pruritic, macular exanthema that started on the lower limbs and progressed to the abdomen, dorsum, chest, and upper limbs (Figure 1A-D). Additionally, she had arthritis of the third and fourth proximal interphalangeal joints of the right hand. Laboratory tests showed elevated acute phase reactants (leucocytes 16,600/µL, neutrophils 15,106/µL, C-reactive protein (CRP) 24mg/dL, and erythrocyte sedimentation rate (ESR) 92mm/hr).

She was hospitalised and started ceftriaxone 4g/day. Nevertheless, she remained febrile with persistently elevated inflammatory markers. On the 9th day of admission, she developed partial respiratory insufficiency and hypotension. Blood work revealed new-onset cytopenias (haemoglobin 10.7g/dL, platelets 135,000/µl), a decrease in ESR (120 to 68mm/hr) and fibrinogen (846 to 335mg/dL), and elevation of ferritin (57,674ng/mL). Chest radiographs showed diffuse bilateral infiltrates (Figure 2).

She was transferred to the Paediatric Intensive Care Unit (PICU) of a tertiary referral university hospital, where respiratory and vasopressor support were started. She remained in the PICU for 15 days, during which blood work revealed anaemia (lowest 7.4g/dL), leucopenia (lowest 800/µL), neutropenia (lowest 0/µL), thrombocytopenia (lowest 55,000/µL), hypofibrinogenaemia (lowest 75mg/dL) and elevation of aspartate aminotransferase (AST - highest 4,293U/L), alanine aminotransferase (ALT - highest 1,098U/L), lactate dehydrogenase (LDH - highest 12,253U/L), triglycerides (TG - highest 849mg/dL), soluble CD25 (sCD25 - 11,850pg/mL), serum calprotectin (highest >22.76µg/mL), and ferritin (highest 357,976ng/mL).

Aetiologic investigation ruled out an infectious cause, and the immune profile (antinuclear, extractable nuclear, anti-neutrophil cytoplasmic and anti-double stranded DNA antibodies, rheumatoid factor, inflammatory myopathies blot) was negative. Bone marrow aspiration and biopsy, performed as part of the aetiologic work-up to exclude malignancy and infection and to support the diagnosis, showed a moderately hypocellular marrow with hypoplasia of the erythrocytic series, dysmegakaryopoiesis, and numerous macrophages with haemophagocytosis. Myelocultures and *Leishmania* testing were negative. Flow cytometry and immunophenotyping excluded monoclonality and showed no inversion of the CD4/CD8 ratio. An exome-based gene panel for inherited immune dysregulation disorders (576 genes

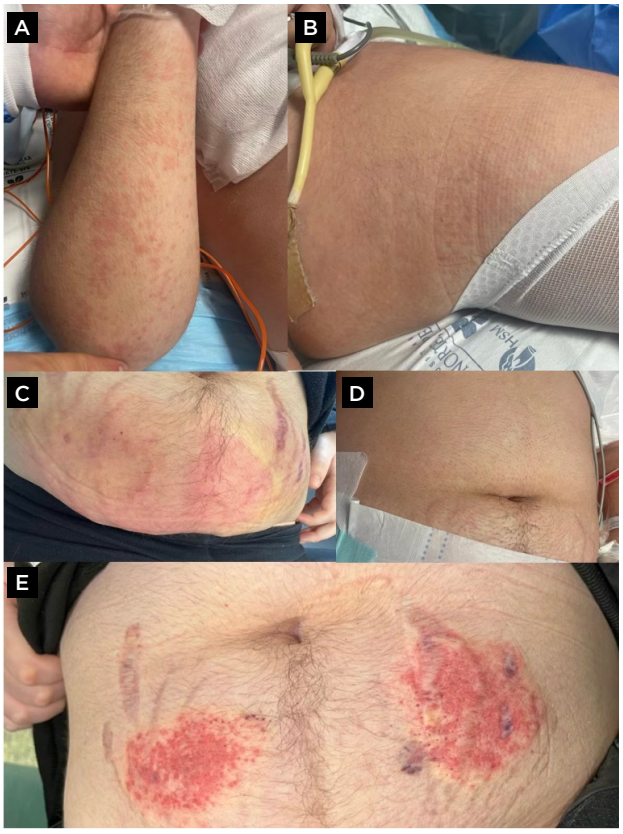

Figure 1. Exanthematous macular rash of the forearm (A), proximal lower limb (B) and abdomen (C,D). Local cutaneous reaction at the site of administration of subcutaneous anakinra (E).

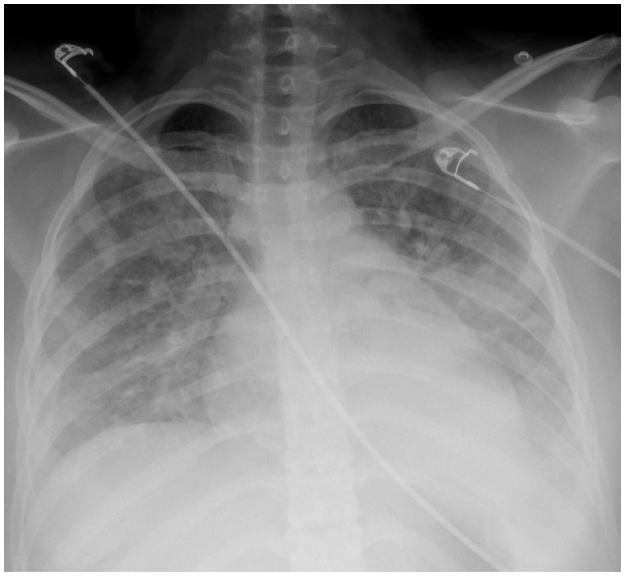

Figure 2. Chest radiograph revealing bilateral patchy opacities suggestive of acute respiratory distress syndrome.

associated with HLH, autoinflammatory syndromes, and immunodeficiencies) did not identify pathogenic variants. An abdominal ultrasound and computed tomography scan revealed hepatomegaly (19cm) and splenomegaly (15cm).

The diagnosis of MAS, most likely secondary to SD, complicated by acute respiratory distress syndrome (ARDS), was established. The diagnosis was established based on clinical and laboratory findings, and immunomodulatory treatment was promptly initiated upon clinical suspicion, without delay for completion of the diagnostic work-up. She was treated with intravenous (iv) methylprednisolone 1g daily for 5 days, followed by iv methylprednisolone 1mg/kg/day (90mg/day), and with iv anakinra 5mg/kg/day (100mg q6h). After three days, anakinra was stopped due to hepatotoxicity (AST peak 4,086 U/L, ALT peak 1,116 U/L), with subsequent improvement in transaminase levels. Ciclosporin 3.75 mg/kg/day (150mg q12h) was then started, and anakinra was eventually reintroduced at a reduced dose of 2.5mg/kg/day (50mg q8h), without a significant transaminase rise. Additionally, she was given broad-spectrum antibiotic therapy.

ARDS was managed with non-invasive ventilation during the first seven days of PICU stay, and vasoplegic shock was managed with vasopressor support for three days. Following MAS-directed therapy, she improved, with resolution of fever, hypoxaemia, and rash. After 15 days in the PICU, she was transferred to the paediatric ward, on iv methylprednisolone 90mg/day, iv anakinra 3.75 mg/kg/day (100mg q8h), and ciclosporin 3.75 mg/kg/day (150mg q12h). However, she developed a urinary tract infection accompanied by recrudescence of fever and rash, and worsening of inflammatory and haemophagocytic parameters. Directed antibiotic therapy was started, two cycles of IVIg 1g/kg/day were administered, and anakinra dose was increased to 5mg/kg/day (100mg q6h), with resolution of the fever and rash. She also developed ciclosporin-associated hypertension, which was managed with amlodipine 10mg/day.

During this period, she had four episodes of epistaxis and reported nausea, abdominal pain, and headache. Blood work revealed worsening thrombocytopenia (18,000/µL) and haemolytic anaemia (5.6g/dL), with an increase in LDH and unmeasurable levels of haptoglobin. Coombs test was positive, a blood smear revealed spherocytes and schizocytes, and ADAMTS13 levels were normal. Considering the possibility of drug-induced microangiopathy, ciclosporin was discontinued, and four daily pulses of iv methylprednisolone 1g were given. Despite significant clinical improvement and haptoglobin normalisation, there was only a partial and transient improvement of the pancytopenia and haemophagocytic markers (haemoglobin 6.6g/dL, leucocytes 1800/µL, neutrophils 1220/µL, platelets 66,000/µL, fibrinogen 95mg/dL, AST 397U/L, ALT 566U/L, LDH 823U/L, TG 244mg/dl, ferritin 97,417pg/ml) and she was dependent on erythrocyte transfusions and filgrastim. Additionally, during the hospital stay, she experienced multiple symptomatic episodes of supraventricular paroxysmal tachycardia, requiring bisoprolol (maximum dose 10mg/day) and flecainide (maximum dose 200mg/day) to maintain sinus rhythm.

Due to refractoriness to high-dose corticosteroids and anakinra, ruxolitinib 5mg q12h was started, but it had to be discontinued due to CMV reactivation. Subsequently, compassionate use of emapalumab was requested from the national medicines authority. Emapalumab was started on the 45th day of hospitalisation. The first 6mg/kg (450mg) dose was followed by nine 3mg/kg (250mg) administrations, each spaced three days apart. She showed progressive clinical improvement until becoming asymptomatic, with concurrent improvement in inflammatory and haemophagocytic markers (haemoglobin 10.1g/dL, leucocytes 13,100/µL, platelets 91,000/µL, fibrinogen 234mg/dL, CRP 0.05mg/dL, AST 31U/L, ALT 162U/L, LDH 649U/L, TG 121mg/dL, ferritin 872pg/mL). Corticosteroid tapering was started, and she was discharged on the 82nd day of hospitalisation, receiving oral prednisolone 80mg/day (40mg q12h) and subcutaneous anakinra 2.5mg/kg/day (100mg q12h).

One month after discharge, anakinra was switched to canakinumab due to severe cutaneous reaction at the sites of administration (Figure 1E). Six months later, receiving iv canakinumab 8mg/kg q4w and off corticosteroids, she remains asymptomatic and with no laboratory evidence of inflammation or haemophagocytosis.

Of note, during hospitalisation and follow-up, the patient received multiple prophylactic therapies: acyclovir 11 mg/kg/day (300mg q8h), which was later switched to valganciclovir (900mg q24h); sulfamethoxazol+trimetoprim (960mg 3x/week) and fluconazol (100mg q24h). Despite this, she experienced several infectious complications, including one urinary tract infection, one central venous catheter infection with bacteraemia, one viral respiratory infection, a pilonidal sinus infection, two oropharyngeal candidiasis episodes, and a CMV reactivation.

DISCUSSION

This case underscores the importance of a high index of clinical suspicion in the setting of MAS. Diagnosis can be challenging owing to limited medical awareness of this condition and its non-specific clinical features.

TABLE I. Case series, reports and trials on the use of emapalumab in macrophage activation syndrome.

Authors	Type of manuscript (N patients)	Age / Sex	Disease context	Other therapies	Emapalumab regimen	Follow-up time	Response	Adverse events
Grom et al. (7)	Two open-label, single-arm trials (39)	1-64y 31F 8M	MAS in patients with previous diagnosis of SD.	**Previous** - High-dose intravenous corticosteroids; - 31 patients: anakinra; - 24 patients: calcineurin inhibitor; - 4 patients: intravenous immunoglobulin.	**Dosing** - 1st administration: 6mg/kg; - following administrations: 3mg/kg. **Timing** - first 16 days: every 3 days; - 16th day - 28thday: twice a week. Time of treatment: ≥28 days. Total administrations: not specified.	2 months.	At week 8 (composite endpoint): - 21 patients: complete response; - 11 patients: partial response; - 2 patients: dead. 33 patients achieved complete response at any time. Corticosteroid dose reduction.	Viral and bacterial infections, most mild-moderate. 6 SAEs reported in 4 patients, not specified. 14 infusion reactions (none serious).
De Benedetti et al. (8)	Open-label, single-arm trial (14)	2-25y 10F 4M	MAS in patients with previous diagnosis of SD.	**Previous** - High-dose intravenous corticosteroids; - 7 patients: anakinra; - 8 patients: ciclosporin; - 3 patients: intravenous immunoglobulin. **Concomitant** - High-dose intravenous / oral corticosteroids; - 5 patients: anakinra; - 6 patients: ciclosporin.	**Dosing** - 1st administration: 6mg/kg; - following administrations: 3mg/kg. **Timing** - every 2-3 days. Time of treatment: 7-39 days. Total administrations: 3-17.	12 months.	At a median of 25 days: - 13 patients: complete clinical and laboratorial response; - 1 patient: complete clinical and partial laboratorial response. Corticosteroid dose reduction.	Viral infections and viral positive tests; namely CMV. 2 infusion reactions (pruritic rash).
Gabr et al. (9)	Case report (1)	22y, F	MAS as inaugural manifestation of SD.	**Previous** - High-dose intravenous / oral corticosteroids; - Subcutaneous anakinra, 100mg/day. **Concomitant** - High-dose intravenous / oral corticosteroids.	**Dosing** - 1st administration: 1mg/kg; - 2nd - 9th administrations: 3mg/kg. **Timing** - 1st - 4th: every 3 days; - 5th - 9th: every week. Time of treatment: 59 days. Total administrations: 9.	5 months.	Clinical improvement; residual rash of the hands Laboratory abnormalities resolution. Corticosteroid dose reduction.	No infusion reactions. Adverse events not reported.
Faggioli et al. (10)	Case report (1)	41y, F	MAS in a patient with previous diagnosis of DM and CD.	**Previous** - High-dose corticosteroids; - Subcutaneous anakinra, 1200 mg/day; - Intravenous immunoglobulin, 1g/kg/day, 2 days; - Eculizumab, 900mg/week, 4 weeks. **Concomitant** - High-dose corticosteroids.	**Dosing** - 1st administration: 6mg/kg; - following administrations: 3mg/kg. **Timing** - first 15 days: every 3 days; - 15th day - 28th day: twice a week. Time of treatment: 28 days. Total administrations: not specified.	9 months.	Clinical and laboratorial remission. Corticosteroid dose reduction.	Not reported.

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TABLE I. continuation.

Authors	Type of manuscript (N patients)	Age / Sex	Disease context	Other therapies	Emapalumab regimen	Follow-up time	Response	Adverse events
Slaney et al. (11)	Case report (1)	2y, M	MAS (6th episode) in a patient with previous diagnosis of SD. Trigger: adenovirus infection.	**Previous** - High-dose intravenous / oral corticosteroids; - Intravenous anakinra, 25 mg/kg/day; - Intravenous ciclosporin / oral tacrolimus. **Concomitant** - High-dose intravenous / oral corticosteroids; - Intravenous anakinra, 46mg/kg/day; - Etoposide, 150 mg/m^2 weekly, 8 weeks - Oral tacrolimus. **Others:** intravenous cidofovir.	**Dosing** - 1st administration: 3mg/kg; - 2nd administration: 6mg/kg; - following administrations: 10mg/kg. **Timing** - twice a week. Time of treatment: 16 months. Total administrations: not specified.	24 months.	Clinical and laboratorial remission. Anakinra and etoposide suspension. Corticosteroid suspension.	Possible opportunistic respiratory infections.
Chellapandian et al. (12)	Case report (1)	4y, F	MAS (recurrent episodes) in a patient with previous diagnosis of SD.	**Previous (for SD/MAS)** - High-dose intravenous / oral corticosteroids; - Tocilizumab; - Anakinra, 4mg/kg/day; - Methotrexate, 10mg/week; - Canakinumab, 4mg/kg every 4 weeks. **Concomitant** - High-dose intravenous corticosteroids.	Dosing - 1st administration: 6mg/kg; - following 6 administrations: 3mg/kg. **Timing** - twice a week. Time of treatment: 4 weeks. Total administrations: 7.	21 months.	Clinical and laboratorial remission. Disease control before allogeneic hematopoietic stem cell transplantation (with reduced-intensity conditioning).	Not reported.
Macaraeg et al. (13)	Case report (1)	20y, M	MAS (2 episodes) in a patient with previous diagnosis of SD. Trigger: -1" episode: EBV infection; - 2nd episode: trimethoprim/ sulfamethoxazole	**Previous (for SO/MAS)** - High-<lose intravenous corticosteroids; - Ganakinumab, every 4 weeks. **Concomitant** - High-<lose intravenous / oral corticosteroids; - Ruxolitinib (added on the 2Mepisode).	Dosing - not specified. **Timing** - twice a week. Time of treatment: not specified.. Total administrations: 6.	1 month.	Clinical and laboratorial improvement. 2M MAS episode after 4 emapalumab administrations; assumed trigger: trimethoprim/ sulfamethoxazole; treated with ruxolitinib.	Not reported.

CD - Crohn's disease; CMV - cytomegalovirus;O M - dermatomyos" ilis;E BV - Epste,n-8arrv irus; F - female; M - male; MAS - macrophagea ct,vations yndrome; SAEs - serious adverse events; SO - Still's disease.

Careful evaluation of clinical and laboratory patterns, interpreted collectively and longitudinally, is particularly important. MAS, particularly when secondary to SD, can easily be misdiagnosed as an infectious disease, as illustrated in this case. Clinical worsening despite antibiotic therapy, the lack of isolation of an infectious agent, a paradoxical decrease in ESR and fibrinogen despite clinical deterioration, and ferritin elevation were key to the diagnosis. Measurement of ferritin upon clinical suspicion is critical, as it is a sensitive marker of disease. It is fundamental for early diagnosis, which is a key determinant of treatment response and prognosis[1,2]. New, more specific biomarkers, such as IL-18, CXCL9, sCD25, and serum calprotectin are increasingly available in daily practice and, consequently, clinically relevant[1,2].

The diagnostic delay in this case was partly due to MAS being the initial presenting manifestation of SD. While MAS typically develops in patients with an established diagnosis of SD, physicians must keep this diagnosis in mind even in the absence of a known underlying rheumatic diagnosis[3].

The management of MAS requires a multifaceted approach. While the diagnostic workup is underway, it is essential to concurrently investigate potential underlying conditions—such as rheumatic or oncologic diseases—identify contributing factors and possible disease flare triggers, initiate supportive care, and begin immunomodulatory therapy. In this case, this comprehensive diagnostic, investigative, supportive, and therapeutic strategy was implemented concurrently while the patient was admitted to the PICU.

Differentiating between a MAS flare and an infectious complication can be challenging, given their potentially overlapping clinical and laboratory presentations. In the event of an apparent clinical and/or laboratory worsening of MAS characteristics, one must assess the evolution of the remaining MAS-associated abnormalities as well as markers suggestive of other possible explanations. In this patient, the proactive approach to investigation facilitated the early detection and management of complications, such as MAS-associated ARDS, MAS-associated supraventricular paroxysmal tachycardia, infections, and ciclosporin-induced microangiopathy. A multidisciplinary approach was essential for the identification and management of the various disease manifestations and drug-associated complications.

Previous reports, including the first trials in MAS, have documented the role of emapalumab in its treatment. (Table I)[7-13] To date, 58 cases of MAS treated with emapalumab have been described in the literature, including adults and children, mostly with a previous diagnosis of SD. All patients were refractory to high-dose corticosteroid therapy. Multiple drug regimens have been described, with the most common being an initial 6mg/kg administration followed by 3-day spaced, or twice weekly 3mg/kg perfusions. The longest treatment time was 16 months, with a follow-up period extending up to 24 months. The majority of patients presented a complete clinical and laboratory sustained response, while some had a partial laboratory response. One patient had a recrudescence of MAS after the fourth emapalumab infusion and was treated with add-on ruxolitinib[13]. Multiple reports show good efficacy and tolerability of emapalumab in addition to other immunomodulatory therapies, such as anakinra and ciclosporin[7,8,11,13]. The most common adverse events reported were mild to moderate viral infections, namely CMV. Mild infusion-related reactions were also described[7-13]. In this case, compassionate use of emapalumab was followed by disease remission, after a prolonged period of uncontrolled activity. This treatment was well tolerated and enabled corticosteroid discontinuation and anakinra dose reduction.

CONCLUSION

This case, describing a severe episode of MAS as the inaugural manifestation of a presumed underlying SD, highlights the diagnostic and therapeutic challenges of MAS.

Diagnosis was delayed due to initial misdiagnosis as infection, underscoring the need for a high index of suspicion and early testing for sensitive markers such as ferritin.

This case also supports the therapeutic potential of IFNγ inhibition in severe, refractory MAS. The disease was refractory to high-dose corticosteroids, ciclosporin, and IL-1 blockade, with only partial and transient benefit. Emapalumab, an IFNγ inhibitor not yet approved in Europe and administered for the first time in Portugal, was associated with rapid and sustained remission after more than two months of uncontrolled disease.

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