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**Manuscript NO:** 55279

**Manuscript Type:** CASE REPORT

**Macrophage activation syndrome as a complication of dermatomyositis: A case report**

Zhu DX *et al.* MAS as a complication of dermatomyositis

Ding-Xian Zhu, Jian-Jun Qiao, Hong Fang

### Abstract

#### BACKGROUND

Macrophage activation syndrome (MAS) can be a fatal complication of rheumatic disorders, which occur most commonly in patients with systemic juvenile idiopathic arthritis or systemic lupus erythematosus; it has rarely been reported in patients with dermatomyositis. Here, we describe a fatal case of MAS that developed in an adult patient with dermatomyositis.

#### CASE SUMMARY

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Oct 16, 2019 · **Macrophage activation syndrome** is a rare entity, rarely reported during **primary Sjögren's syndrome**. Its **spontaneous evolution** is invariably fatal. There is no consensus on **therapeutic treatment**.

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Jul 30, 2018 · The present **case** describes **macrophage activation syndrome (MAS)** associated with **dermatomyositis (DM)** and **cytomegalovirus (CMV)** viremia. **MAS** is a rare condition of **profound systemic** but ineffective **immune activation** affecting multiple organs with high mortality.

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**Keywords:** **Macrophage activation syndrome** , **Sjögren's syndrome** , Africa south of the Sahara , **Case report** Background **Macrophage activation syndrome (MAS)** is a phenomenon characterized by cytopenia, organ dysfunction, and coagulationopathy associated with an inappropriate **activation** of macrophages [1]. It remains a rare pathology, with a guarded

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**Cited by:** 1 **Author:** Allison Virginia Lange, Salahuddin Kazi...

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### [\[PDF\] Fatal case of macrophage activation syndrome \(MAS\) in a ...](#)

<https://casereports.bmj.com/content/casereports/2018/bcr-2018-225231.full.pdf>

We describe a **case** of an adult with **dermatomyositis (DM)** who presents with a **rash**, high fevers, tachycardia and hypotension, initially concerning for an **infectious aetiology** or a **DM flare**. She was found to have **cytomegalovirus viraemia** which improved after starting valganciclovir.

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