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# SECONDARY HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS IN A PATIENT WITH DERMATOMYOSITIS, INTERSTITIAL LUNG DISEASE, AND PRIOR MALIGNANCY: A DIAGNOSTIC AND THERAPEUTIC CHALLENGE

General Medicine

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## KEYWORDS

Haemophagocytic lymphohistiocytosis, Dermatomyositis, Autoimmune disease, Interstitial lung disease, Critical care, Secondary HLH

**BACKGROUND**:
Haemophagocytic lymphohistiocytosis (HLH) is an aggressive, hyperinflammatory syndrome resulting from uncontrolled activation of the immune system. It can occur secondary to autoimmune disorders (macrophage activation syndrome), infections, and malignancies. Dermatomyositis (DM) is a rare idiopathic inflammatory myopathy with known associations with interstitial lung disease (ILD) and malignancy, making diagnostic workup challenging in acutely unwell patients. HLH in the context of DM is rarely reported and may be easily misattributed to disease flare or infection.

**Case Presentation**:
A middle-aged woman with known DM and ILD presented with a 16-month history of upper limb pain, 15 months of neck weakness and dysphagia, and more recent onset (10 days) of proximal lower limb weakness and urinary symptoms (urgency, hesitancy, and loss of bladder sensation). She had a history of mucinous colorectal carcinoma, status post right hemicolectomy.

On admission, she had worsening respiratory distress requiring intubation. Chest X-ray and high-resolution CT of the thorax showed evidence of ILD with superimposed pneumonia. She was managed with empirical antibiotics and intravenous immunoglobulin (2g/kg over 5 days) due to concern for a dermatomyositis flare.

Subsequently, the patient developed pancytopenia and hypoalbuminaemia. Immunosuppressive therapy, including azathioprine, was discontinued. A bone marrow biopsy was performed after noting elevated serum ferritin and triglycerides, and low fibrinogen levels. Phagocytic histiocytes were observed, consistent with HLH. A contrast-enhanced CT and malignant cytology from ascitic fluid were negative for recurrent malignancy.

The patient received intravenous methylprednisolone, later transitioned to oral corticosteroids. She underwent tracheostomy for ventilatory support. Multiple consultations were obtained: nephrology (for pre-renal acute kidney injury), gastroenterology (for cholestasis, suspected macrophage activation), oncology, and rheumatology.

Oral steroids were temporarily withheld in preparation for a PET scan to investigate occult malignancy. Over the next 48 hours, her condition deteriorated with worsening cytopenias, elevated inflammatory markers, and hypofibrinogenemia. Sepsis was suspected; empirical antibiotics were continued pending cultures.

she developed refractory hypotension requiring vasopressors. On 4 April, she had a sudden cardiac arrest with concurrent hypoglycaemia and hyperkalaemia. Resuscitative efforts, including defibrillation and advanced cardiac life support, were unsuccessful. She was declared deceased.

**Investigations**:
- **Chest X-ray and HRCT:** ILD with superimposed pneumonia
- **Bloods:** Pancytopenia, elevated ferritin (>2000 ng/mL), hypertriglyceridemia, hypofibrinogenemia
- **Bone Marrow Biopsy:** Haemophagocytosis
- **Ascitic Fluid Cytology:** No malignant cells
- **PET Scan:** Planned but not performed
- **Cultures:** Pending at time of death

**Differential Diagnosis:**
- Dermatomyositis flare
- Secondary HLH
- Infection-related sepsis
- Paraneoplastic syndrome/occult malignancy recurrence
- Drug-induced cytopenia (azathioprine-related)

**Treatment**:
- IV immunoglobulin (2 g/kg over 5 days)
- Empirical antibiotics (meropenem and others)
- **Immunosuppression:** High-dose IV methylprednisolone, later oral steroids
- **Supportive Care:** Mechanical ventilation, vasopressors, tracheostomy
- **Planned:** Rituximab post-stabilisation

**Outcome And Follow-up:**
Despite aggressive therapy, the patient experienced progressive hemodynamic instability and multi-organ involvement. She developed refractory shock and suffered a fatal cardiac arrest. No post-mortem examination was performed.

**DISCUSSION**:
HLH in the context of dermatomyositis is rare but increasingly recognised. The patient met several HLH-2004 criteria, including fever, cytopenias, hyperferritinemia, hypertriglyceridemia, hypofibrinogenemia, and bone marrow evidence of haemophagocytosis.

Overlap with autoimmune disease flare, infections, and previous malignancy delayed recognition. In our patient, early withdrawal of immunosuppression (to investigate possible malignancy) and the inability to rapidly escalate immunomodulatory therapy (e.g., rituximab or etoposide) may have contributed to clinical decline.

Multidisciplinary input was essential, yet the patient's rapid deterioration emphasises the need for early HLH recognition in similar high-risk cases. Diagnosis can be aided by the HScore or HLH-2004 criteria, but clinical judgment remains central.

**Learning Points:**
- HLH can occur as a complication of dermatomyositis and may be life-threatening.
- HLH diagnosis is challenging in patients with overlapping autoimmune disease, infection, and malignancy.
- Delay in immunosuppressive therapy may lead to rapid clinical deterioration.
- Bone marrow biopsy and inflammatory markers are essential in differentiating HLH from sepsis or autoimmune flares.
- Early multidisciplinary management is critical in complex autoimmune-critical care cases.

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The authors declare no competing interests.

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