Acquired haemophagocytic lymphohistiocytosis secondary to multiple myeloma

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DESCRIPTION

A 56-year-old man who initially presented with immunoglobulin G lambda solitary sacral plasmacytoma 1 year ago treated with radiation therapy was hospitalised with severe anaemia. Oesophago-gastroduodenoscopy performed 2 months prior had shown non-bleeding gastric mucosal involvement by plasma cell neoplasm. Hospital course was complicated by high fevers and bicytopenia, including transfusion-dependent anaemia. Laboratory data were notable for a markedly elevated

Figure 1 (Panel A) Marked histiocyte hyperplasia with haemophagocytosis. (Panel B) Histiocytes confirmed by positive CD 163 on immunohistochemistry. (Panels C and D) Examples of haemophagocytosis.

Box 1 Diagnostic criteria for acquired haemophagocytic lymphohistiocytosis

- ▶ Fever.
- ▶ Splenomegaly.
- ▶ Cytopenias involving $\geq 2/3$ lineages in the peripheral blood: haemoglobin < 9 g/dL, platelets $< 100 \times 10^9$ /L, neutrophils $< 1.0 \times 10^9$ /L.
- ▶ Hypertriglyceridaemia and/or hypofibrinogenaemia: fasting triglycerides ≥ 265 mg/dL and/or fibrinogen ≤ 1.5 g/L.
- ▶ Haemophagocytosis seen in bone marrow, spleen or lymph nodes.
- ▶ Low or absent natural killer cell activity.
- ▶ Ferritin ≥ 500 μ g/L.
- ▶ Soluble interleukin-2 receptor ≥ 2400 pg/mL.

Patient's perspective

I understand that I have a condition that is very high in mortality but I am trying to undergo treatment for the same. If my case can help other physicians learn, it would mean a lot to me.

Learning points

- ▶ Acquired haemophagocytic lymphohistiocytosis (HLH) especially secondary to multiple myeloma is a very rare diagnosis, but we should definitely think about it in the appropriate clinical setting.
- ▶ It is a condition with a very high mortality rate and hence we should maintain a high degree of clinical suspicion to prevent delay in the treatment.
- ▶ Etoposide and dexamethasone are the standard of care for HLH.

ferritin level of 16 123 U/mL. Bone marrow biopsy findings included marked histiocyte hyperplasia with haemophagocytosis represented by the black arrows in panel A (figure 1, viewed with 100 $\times$ objective) and plasma cell involvement of 20%. Panel B (figure 1) shows circumferential brown staining and foamy cytoplasm indicating histiocytes confirmed by positive CD 163 on immunohistochemistry. Further examples of haemophagocytosis are seen in panels C and D (figure 1). The clinical picture of fevers and cytopenias combined with the bone marrow findings and high ferritin established the diagnosis of haemophagocytic lymphohistiocytosis (HLH) which was later confirmed with soluble interleukin-2 receptor level of 10 680 pg/mL. He was treated with dexamethasone and etoposide for 8 weeks but eventually succumbed to multiorgan failure.¹

Acquired HLH can be diagnosed when five out of the following eight criteria are fulfilled as illustrated in box 1.²

It can present secondary to an infection, autoimmune disorders or malignancies. It has rarely been described associated with multiple myeloma, especially involving the gastric mucosa making this case unique. It carries a very high mortality rate, and a high index of clinical suspicion is required in order to make the diagnosis and start treatment in a timely manner.

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