ANSWER TO PHOTO QUIZ (PAGE 193)

AN UNUSUAL CAUSE OF FEVER AND CYTOPENIA IN MULTIPLE MYELOMA

DIAGNOSIS

The bone marrow examination showed 20% atypical plasma cells with haemophagocytosis of erythrocytes and leukocytes by these plasma cells (figure 1). The atypical plasma cells expressed CD138 and showed lambda monoclonality. Acquired haemophagocytosis or hemophagocytic lymphohistiocytosis (HLH) is usually characterized by haemophagocytosis by histiocytes and mostly associated with (viral) infections and haematologic malignancies, including (rarely) multiple myeloma. The pathogenesis of HLH is not fully elucidated but characterized by increased production of pro-inflammatory cytokines and activation of cytotoxic T cells and macrophages, reflected by increased serum levels of soluble Interleukin-2 receptor and ferritin, respectively.[1] Haemophagocytosis by neoplastic plasma cells, as seen in our case, is extremely rare and only a few reports describe phagocytic plasma cells in patients with multiple myeloma or plasma cell leukaemia.[2-5] Normal plasma cells are immunoglobulin-producing cells and do not have the ability to phagocytise. The mechanism of acquisition of phagocytic capacity by malignant plasma cells is unclear. Malignant plasma cells isolated from the bone marrow of one of the reported patients revealed no *in vitro* phagocytic capacity.[2] The occurrence of haemophagocytosis is not related to a specific immunoglobulin or light chain subtypes,[3] but occasionally associated with aberrant immunophenotypic (CD15+) features.[4] Haemophagocytosis by plasma cell seems more frequently in female patients and it appears that mature erythrocytes and platelets are predominantly phagocytised. The pathophysiological mechanism of myeloma-mediated haemophagocytosis is different from that of the HLH. Thus, the normally-associated biochemical clues, such as a strongly elevated ferritin level, directing the diagnosis of haemophagocytosis as cause of the pancytopenia may not be present. Response to treatment has only been reported in nine patients, of which six showed clinical improvement.[3] In our case, treatment with high-dose dexamethasone was started, and within days, the fever and cold shivers disappeared and the patient felt better. However, he refrained intensive chemotherapeutic treatment and died a few weeks later.

DISCLOSURES

All authors declare no conflicts of interest. No funding or financial support was received.

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