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Immunotherapy-related secondary hemophagocytosis in a glioblastoma patient: response to cytokine-directed therapy

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Abstract

Hemophagocytic Lymphohistiocytosis (HLH) is a severe and potentially life-threatening condition characterized by an excessive and uncontrolled activation of the immune system. ICI-related hemophagocytic lymphohistiocytosis (irHLH) is a rare immune-related adverse event with an incidence of 0.03% to 0.4%. Although rare, it can be potentially lethal, with a high mortality rate of up to 50% in some cases. We present a patient with recurrent glioblastoma who developed Hemophagocytic Lymphohistiocytosis s a result of nivolumab treatment and was subsequently managed with cytokine-directed therapy (tocilizumab). Early diagnosis and treatment of Hemophagocytic Lymphohistiocytosis (HLH) associated with immune checkpoint inhibitors (ICIs) are indeed crucial due to the potentially life-threatening nature of the condition.Cytokine-based treatments (such as anti-IL-6) may be appropriate for patients who do not respond to high-dose steroids.

Keywords: Immune checkpoint inhibitor; cytokine directed therapy; glioblastoma; immune related adverse event; immunotherapy related hemophagocytic lymphohistiocytosis; nivolumab; secondary hemophagocytic syndrome; tocilizumab.

Plain language summary

This case report describes a 44-year-old man who experienced an immune-related adverse event, specifically Hemophagocytic Lymphohistiocytosis (HLH), following immunotherapy for recurrent glioblastoma. He initially presented with persistent fever and lethargy, and further testing confirmed the immune checkpoint inhibitor-related event. High-dose dexamethasone and anti-IL-6 therapy (tocilizumab) were administered, resulting in fever resolution and improvement in laboratory findings. However, his clinical condition and disease progressively worsened. Unfortunately, he passed away due to disease progression.