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Cytomegalovirus-induced haemophagocytic lymphohistiocytosis syndrome

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A 24-year-old female presented with fever and abdominal pain for 1 week. Her medical history included seizures secondary to tuberous sclerosis controlled with lamotrigine and Crohn's disease in remission for the last 2 years treated with 6-mercaptopurine. On presentation, she was found to have pancytopenia and elevated liver enzymes. Physical examination was significant for a petechial rash. Peripheral blood smear showed atypical lymphocytes (figure 1A). Serology was positive for Ig M cytomegalovirus with an elevated viral load of 82 392 copies/ml. HIV testing was negative. Her clinical course was complicated by acute respiratory distress requiring mechanical ventilation. The patient denied history of repeated infections during childhood, any relevant family history or sick contacts. A bone marrow aspirate revealed increased benign-appearing histiocytes (figure 1B) with phagocytosis (figure 1C) of erythrocytes, leucocytes and platelets. The soluble interleukin-2 receptor was 25 568 units/ml. There were insufficient natural killer cells to evaluate for perforin

expression. Genetic testing for familial haemophagocytic lymphohistiocytosis (HLH) including Perforin 1, Munc13-4, Syntaxin 11 were negative. Treatment was initiated with intravenous ganciclovir and intravenous immunoglobulin. Simultaneously, immunochemotherapy with HLH-2004 protocol was administered with dexamethasone, etoposide and cyclosporine for 8 weeks with rapid clinical improvement to the point of no longer requiring life support and concomitant resolution of her pancytopenia. The patient remains in complete remission 6 months after her initial diagnosis and is currently being evaluated for possible stem cell transplantation.

Competing interests None.**Patient consent** Obtained.**REFERENCES**

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Figure 1 Peripheral blood smear showed atypical lymphocytes (A). A bone marrow aspirate revealed increased benign-appearing histiocytes (B) with phagocytosis of erythrocytes (C).

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Munoz J, Shareef N, Donthireddy V. Cytomegalovirus-induced haemophagocytic lymphohistiocytosis syndrome. *BMJ Case Reports* 2012; 10.1136/bcr.10.2011.4963, Published XXX

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