

## Case Report

# A purpuric variant of acute graft versus host disease

## 紫癍形急性移植抗宿主病

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We report a case of acute graft versus host disease (GvHD) in a 31-year-old Indonesian woman diagnosed with acute myelomonocytic leukaemia. She presented with a purpuric eruption of haemorrhagic maculopapules over both palms. This case is unusual in that this is a purpuric form of GvHD that is not commonly seen. It is also an important clinical vignette that reminds clinicians to interpret examination findings in the appropriate clinical context, where initial considerations of pompholyx and eczema should also include GvHD, leukaemia cutis and engraftment syndrome.

一個患有急性白血病的三十一歲印尼籍女子產生了急性移植抗宿主病，在雙掌上爆發出血性丘疹。這篇病例報告的要點是提醒臨床醫師，遇到這種情況除了要考慮汗皰疹、皮膚白血病、植入綜合症以外，也必須想到罕見的血性移植抗宿主病。

**Keywords:** Graft vs host disease, purpura

**關鍵詞：** 移植抗宿主病、血性丘疹

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A 31-year-old Indonesian woman was recently diagnosed with acute myelomonocytic leukaemia presenting with a left breast lump. Due to the aggressive nature of her disease, she had failed

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first-line induction chemotherapy and required salvage chemotherapy which was complicated by pancytopenia and inpatient treatment for severe neutropaenic sepsis. Consequently, she underwent a myeloablative matched umbilical cord haematopoietic progenitor cell transplant.

At three weeks post-transplantation, she developed an intensely pruritic eruption over her palms over a two-day period. On examination, there were multiple purpuric maculopapules and haemorrhagic vesicles on both palms and fingers ranging from 1 to 3 mm (Figures 1 & 2). At that time, investigations revealed that she was bicytopenic with an absolute neutrophil count of 780 cells/ $\mu$ L, and a platelet count of 7,900/ $\mu$ L.

Our consideration for this symmetrical, pruritic and vesicular palmar eruption was that of pompholyx, but given the very recent history of stem cell transplantation, acute graft-versus-host disease (GvHD) was also considered. Leukaemia cutis would be a valid consideration too, though its lesions are more frequently found on the head, neck and trunk.

Skin biopsies were taken from the palms. Histology (Figures 3a & 3b) demonstrated acanthosis, focal parakeratosis, exocytosis, and spongiosis with

scattered apoptotic keratinocytes and intra-epidermal red blood cells in the overlying epidermis. The dermis showed papillary oedema with focal extravasation of red blood cells. There was also discernible vacuolar alteration at the dermoepidermal junction with lymphohistiocytic infiltrates around the superficial dermal vessels. Vasculitic features were absent.

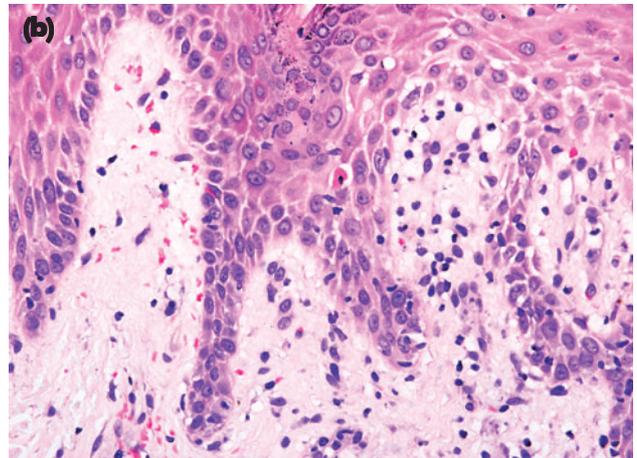
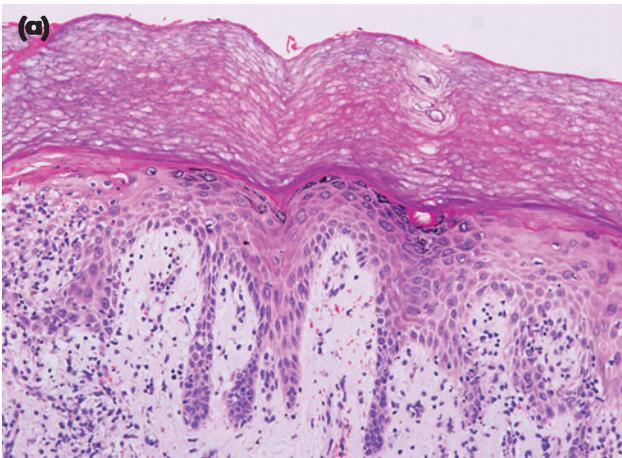
This confirmed the diagnosis of acute GvHD, of a grade 1 histologic stage, presenting as an uncommon purpuric variant. The purpuric



**Figure 1.** Multiple purpuric maculopapules with haemorrhagic vesicles on right palm.



**Figure 2.** This is the patient's left palm revealing a similar morphology.



**Figure 3.** (a) Low power view of the skin biopsy showed acanthosis, spongiosis and exocytosis of the epidermis with papillary oedema and extravasation of the red blood cells in the dermis (Haematoxylin & eosin, original magnification x 200). (b) On higher power, there was discernible focal vacuolar alteration at the dermoepidermal junction with an apoptotic keratinocyte in the epidermis (Haematoxylin & eosin, original magnification x 400).

element is likely contributed by concurrent thrombocytopenia.

She was started on mycophenolate mofetil 500 mg BD initially, but a fine erythematous rash continued to progress and spread over her chest and anterior trunk in patches. Fortunately, the body surface area of involvement was limited to 25%, without further evolution of the rash. She was started on prednisolone 1 mg/kg/day, resulting in resolution over a two-week period.

## Discussion

Acute GvHD usually occurs 10 to 30 days from transplantation, and is attributed to the T-cell response when immunologically competent cells are introduced into an immune-incompetent host.<sup>1,2</sup> This is a common complication and can occur from 20 to 50% of patients who receive cells from a HLA-identical sibling donor, and up to 80% in those where there is a mismatch.<sup>3</sup> Acute GvHD affects five organ systems, the skin, gastrointestinal tract, the liver, the lungs, and lymphoid tissue, of which skin involvement is the most frequently seen,<sup>4</sup> and is associated with a poorer prognosis.<sup>5</sup>

Cutaneous presentations usually affect the palms or soles first, and then progress to a widespread maculopapular rash with desquamation, erythroderma and blistering should the condition become more severe, leaving postinflammatory changes. Rare forms that have been described are scarlatiniform, varicelliform or ichthyosiform in nature.<sup>6</sup>

This case describes a purpuric variant of acute GvHD which is not commonly seen.<sup>7</sup> It also entails an approach to haemorrhagic blistering eruptions that is important to clinical dermatology, where the common considerations include herpes zoster, bullous impetigo, acute eczema, pompholyx, contact dermatitis and autoimmune causes such as pemphigus vulgaris.

Furthermore, this case highlights the importance of interpreting clinical features in the appropriate clinical context. Although the symptomatology and vesicular appearance are suggestive of pompholyx, given the patient history, consideration should be given to differential diagnoses of acute GvHD, leukaemia cutis which may herald recurrent disease, engraftment syndrome, drug eruptions such as palmoplantar hyperpigmentation secondary to chemotherapy e.g. cyclophosphamide, and infective causes such as herpes zoster in the immunocompromised host. Their histopathological findings share many similarities, hence the paramount importance of making clinico-pathological correlation. The medical history of a recent stem cell transplant, the progression and evolution of the eruption, the histological findings and response to therapy were indeed indicative of acute GvHD.

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