affecting the hands and feet, followed 3–4 days later by symmetrical swelling of the palms and soles of the feet, together with erythema and tenderness, particularly of the distal phalanges. With continued drug therapy the swelling and erythema progresses and a central pallor develops over the tufts of the distal phalanges. Most reported cases have resolved within 7 days of discontinuation of therapy[2], but tend to recur when therapy is re-instituted. Pyridoxine in a dose of 100 mg orally daily has been recommended as treatment that will allow continuation of chemotherapy with 5-FU while maintaining remission of this cutaneous complication of treatment[5].

In our case, not only did the condition only slowly and partially respond to cessation of 5-FU therapy and pyridoxine therapy, but the patient had persistence of abnormal sensation and appearance of the affected digits.

It is apparent that PPEDS as a complication of 5-FU therapy may result in continuing symptoms and signs for many months after withdrawal of the causative agent, and may not respond to pyridoxine therapy. In some cases the degree of degradation of quality of life may be greater from this complication than would be expected, and may warrant prolonged withdrawal of the offending drug.

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Adult-onset congenital erythropoietic porphyria (Günther's disease) presenting with thrombocytopenia

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Cutaneous signs of Günther's disease (congenital erythropoietic porphyria) developing 5 years after the onset of symptomatic thrombocytopenia are described in a 65-year-old man. Persistent thrombocytopenia unresponsive to corticosteroids and immunoglobulin necessitated a splenectomy.

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Table 1 **Patients' porphyrin levels**

Substrate	Porphyrin	Patient	Normal range
Urine	Coproporphyrin (nmol/24 h)	6399	<246
	Uroporphyrin (nmol/24 h)	5881	<36
Stool	Coproporphyrin (nmol/g dry wt)	905	<40
	Protoporphyrin (nmol/g dry wt)	750	<135
Erythrocyte	Free erythrocyte porphyrin (μg/l)	665	<590

CASE REPORT

A 65-year-old man presented with a gastrointestinal haemorrhage and a 2 month history of pruritic haemorrhagic blisters on sun-exposed skin. Five years previously he was noted to have mild thrombocytopenia (platelets=120×10^9/l) and anaemia which led to a diagnosis of atypical myelofibrosis. Two years later ongoing anaemia and thrombocytopenia had necessitated blood and platelet transfusions.

Dermatological assessment at presentation revealed hyperpigmentation and waxy induration of sun-exposed skin. Haemorrhagic blisters, erosions, milia and scarring affected the scalp, face and dorsal surfaces of both hands. Scarring alopecia and hypertrichosis affecting the hands and eyebrows were present. Clinically evident splenomegaly was confirmed by abdominal ultrasonography. The urine

Table 2 **Haematological values**

Haemoglobin	10.6 g/dl
MCV	91.2 fl
MCH	29.4 pg
MCHC	32.3 g/dl
White cell count	10.6×10^9/l
Platelets	42×10^9/l

fluoresced brilliantly with Wood's light and examination of erythrocytes with fluorescent microscopy demonstrated stable fluorescence. Table 1 summarizes the results of porphyrin assays. High performance liquid chromatography of urine demonstrated 97% of the uro- and copro-porphyrin to be of isomer type 1. Haematologic values are recorded in Table 2. A blood film revealed poikilocytosis, anisocytosis and nucleated red blood cells. The reticulocyte count was 1.9%, Direct Coomb's test negative, haptoglobins reduced and LDH (772 IU/L) elevated. Platelet associated IgG was present in high titre (ratio=21, normal range<2).

Radio-isotopic studies demonstrated platelet sequestration in the spleen. The bone marrow was hypercellular with marked megakaryocyte hyperplasia but no evidence of fibrosis or cellular infiltration. Skin biopsy revealed subepidermal blisters with PAS positive material deposited in the upper dermis in a perivascular and periadnexal distribution.

Failure of prednisolone 60 mg for 2 months and immunoglobulin (0.4 g/kg intravenously for 5 days) to increase the platelet count led to splenectomy. Four weeks later the platelet count rose to 102×10^9/l, the highest level over the past 5 years, and remains stable since. Porphyrin excretion values remain unaltered and blistering continues on light exposed sites.

DISCUSSION

Adult-onset Günther's disease is rare[1–5] and tends to be milder than childhood cases. Photosensitivity, haemolytic anaemia and hypersplenism are prominent features. Five reports document thrombocytopenia in adult-onset Günther's disease[1–5]. Platelet sequestration studies in this case report implicate the spleen as the major site of platelet consumption. However, platelet associated IgG antibodies were also present. The significance of platelet autoantibodies in association with thrombocytopenia in this patient is uncertain. Coexistence of two separate diseases is possible: Günther's disease and idiopathic thrombocytopenia together with myelofibrosis leading to hypersplenism. However, there are not sufficient diagnostic features to substantiate the diagnosis of myelofibrosis and all of the haematological features have been previously described as features of Günther's disease.

Previous studies[1,2] reported a positive response to oral steroids in two patients with Günther's disease and thrombocytopenia but steroids were ineffective in this case. Splenectomy in some cases increased erythrocyte life span[6], reduced porphyrin levels[3] and increased platelet count in Günther's disease. In this patient 4 weeks following splenectomy the platelet count rose and stabilized and remains stable 1 year later.

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