septicaemia. Early cases often resolve with antibiotics[1] alone and this treatment should continue for 3 months.

The primary source of this bacteriaemia was undetected but synchronous bone and prostatic symptoms developed. In this patient, the findings of acute prostatitis and its resolution were clearly seen on the CT scan.

Both transrectal ultrasound and CT scan of the pelvis are good investigations in the diagnosis of acute bacterial prostatitis but the former may be painful.

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Long-term sequelae of Palmar-Plantar erythrodysaesthesia syndrome secondary to 5-fluorouracil therapy

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CASE PRESENTED TO SECTION OF ONCOLOGY 10 NOVEMBER 1993

Keywords: *5-fluorouracil; Palmar-Plantar erythrodysesthesia syndrome*

Palmar-Plantar erythrodyasesthesia syndrome has been reported as a temporary complication of 5-fluorouracil therapy consisting of a debilitating erythema and tenderness of the skin of hands and feet. The syndrome has previously been observed to respond rapidly to either temporary cessation of chemotherapy or pyridoxine with little residual disability. We report a case which was characterized by persistent morbidity necessitating prolonged discontinuation of treatment.

Case report

A 55-year-old man first presented in October 1991 with acute appendicitis. At operation an inflamed appendix was removed which had perforated near its base. Histology of the appendix confirmed this was acute appendicitis with no evidence of tumour.

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In May 1992 it was noted that he had developed firm subcutaneous nodules at the site of his appendicectomy scar. Aspiration cytology confirmed malignancy and an abdominal computerized tomography (CT) scan demonstrated the presence of an irregular mass consistent with primary tumour in relation to the caecum.

At laparotomy he was found to have inoperable carcinoma of the caecum, with three palpable metastases in the right lobe of the liver. The tumour was by passed with a stapled side to side ileo-colic anastomosis and the histology confirmed the presence of moderately differentiated adenocarcinoma.

He was subsequently commenced on chemotherapy in August 1992 with 48 h infusions of 5-FU, 1500 mg daily preceded by bolus injection of folinic acid 350 mg. Treatment was continued fortnightly until December 1992 at which time he was placed on 3-weekly cycles, to a total dose of 39 g.

At review in March 1993 he was found to have multiple chest deposits visible on chest radiograph, and he was commenced on continuous 5-FU as palliative treatment. This was started at a dose of 200 mg/m^2/day via Walkmed (410 mg continuously daily). At a total dose of 5-FU of 73.5 g he was noted to have developed erythema affecting both feet and hands with subcutaneous fluid collections and painful paraesthesia affecting the distal phalanges; distal phalangeal pallor was a prominent feature at that time. His treatment was discontinued and he commenced on a 7 day course of pyridoxine 120 mg daily to which there was no response. His pain slowly diminished over the succeeding 3 months, but he continued to report paraesthesia and joint stiffness affecting the fingers of both hands. The skin of hands and feet remained thinned, shiny and atrophic, and distal phalangeal pallor was still evident.

DISCUSSION

Palmar-Plantar erythrodysaesthesia syndrome (PPEDS) has been reported as an unusual complication of high dose bolus or protracted continuous 5-fluorouracil (5-FU) therapy[1–4]. The syndrome consists of a prodrome of dysaesthesia

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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Keywords: *congenital erythropoietic porphyria; Günther's disease; thrombocytopenia*

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