3 Mayon-white R. Protection for the asplenic patient. *Prescrib J* 1994;**34**:165–70

4 Zarrabi MH, Rosner F. Rarity of failure of penicillin prophylaxis to prevent post-splenectomy sepsis. *Arch Intern Med* 1986;**146**:1207–8

5 McMullin M and Johnston G. Long term management of patients after splenectomy. *BMJ* 1993;**307**:1372–3

6 Gregg CR, Stuart Tucker W. Acute endogenous endophthalmitis after splenectomy. *J Tennessee Med Assoc* 1985;Jan:25–6

7 Cheesborough JS, Williams CL, Rustom R, Bucknall RC, Trimble RB. Metastatic Pneumococcal endophthalmitis: Report of two cases and review of literature. *J Infect* 1990;**20**:231–46

8 Katten HM, Flynn HW, Pflugfelder SC, Robertson C, Forster RK. Nosocomial endophthalmitis survey. *Ophthalmology* 1991;**98**:227–38

9 Bacon AS, Davison CR, Patel BC, Frazer DG, Ficker LA, Dart JKG. Infective endophthalmitis following vitreo-retinal surgery. *Eye* 1993;**7**:529–34

(Accepted 24 December 1994)

Erythropoietic protoporphyria presenting in an adult

C A Henderson MRCP[1] S Jones PhD[2]
G Elder FRCPath[3] A Ilchyshyn FRCP[1]

J R Soc Med 1995;**88**:476P–477P

CASE PRESENTED TO SECTION OF DERMATOLOGY, 7 MARCH 1995

Keywords: *erythropoietic protoporphyria; prophyrins; cirrhosis*

Erythropoietic protoporphyria is an inherited disorder of porphyrin metabolism, in which reduced activity of the enzyme ferrochelatase leads to accumulation of protoporphyrins in erythrocytes. Protoporphyrins are photoactivated by ultra-violet light causing tissue damage by release of free oxygen radicals, which manifests as photosensitivity. The majority of cases of erythropoietic protoporphyria present in childhood although sometimes symptoms are delayed until the second decade. We report here a case presenting in adulthood and discuss the risk of liver disease in the condition.

CASE HISTORY

A 33-year-old man presented with a long history of pain and itching of the hands and face, together with a rash, following exposure to sunlight. Symptoms were kept to a minimum by sun avoidance, until 12 months prior to presentation when his tolerance to sunlight appeared to deteriorate. He also developed blisters on the backs of his hands for the first time. An erythematous and papular crusted rash was seen on sun exposed sites (Figure 1). He drank less than 40g of alcohol per week.

Departments of [1]Dermatology and [2]Biochemistry, Walsgrave Hospital, Coventry, CV2 2DX, UK [3]Medical Biochemistry, University of Wales College of Medicine, Wales, UK

Correspondence to: C A Henderson

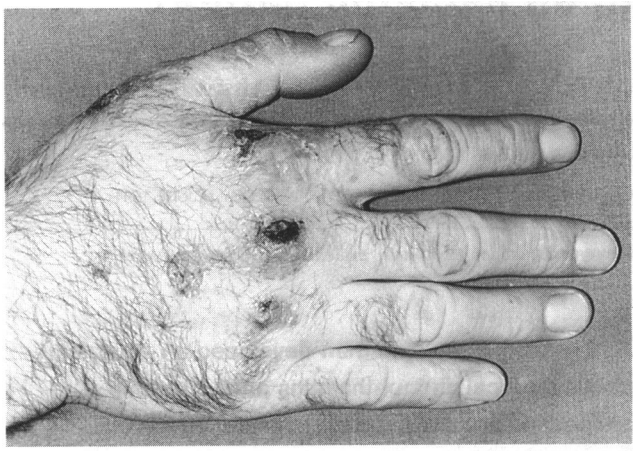

Figure 1 **Crusted lesions over the knuckles of the right hand**

Table 1 **Reports of liver disease in erythropoietic protoporphyria in various studies (Refs 3–7)**

Study	Patients	Abnormal LFT	Cirrhosis	Death from liver disease
1	55	19	7	2
2	32	1	—	—
3	29	—	—	—
4	20	2	—	1
5	402	1	—	2
Total	538	23	7	5

LFT=Liver function test

Investigations revealed normal urinary and faecal porphyrins, but raised erythrocyte porphyrins of 34.7 μmol/L (normal range 0.4–1.7 μmol/L). Fluorescent emission spectroscopy of plasma showed a peak at 627 nm. Liver function tests were normal.

DISCUSSION

Presentation of erythropoietic protoporphyria in adulthood has only been reported on two previous occasions[1,2], one occurring at 62 and the other 69 years. Typically, it presents in childhood with pain on exposure to sunlight, followed by an erythematous crusted rash. Blisters are uncommon and occur in 3–14% of cases.

The main complication of erythropoetic protoporphyria are photosensitivity, cholelithiasis and liver disease. Photosensitivity is treated with sun avoidance and sunblockers. In severely affected cases, agents which scavenge free oxygen radicals may be beneficial. Beta-carotene helps some[3], particularly at higher doses and trials of N-acetyl cysteine are currently underway.

Liver disease is a rare complication, with reports from different countries[3–7] suggesting a very variable incidence (Table 1). This variation may partly be explained by the different age structures of the groups and the different countries of origin, in view of the fact that recent work has suggested that erythropoietic protoporphyria is probably a genetically heterogeneous disease[8]. The most detailed study of liver disease in erythropoietic protoporphyria followed up 55 cases over 20 years and showed a particularly high incidence[4]. Those that developed liver complications had a significantly higher mean erythropoietic protoporphyrin level (38 μmol/L $\pm$ 8) compared to those without liver damage (13 μmol/L $\pm$ 2). The technique used to measure the proporphyrins was, however, less sensitive than that used in this patient and so is probably not directly comparable.

Mildly abnormal liver function tests are common in erythropoietic protoporphyria, but when symptomatic liver disease develops it tends to progress rapidly as excretion of porphyrins is impaired, leading to further accumulation and liver damage. The average age of 15 patients reported to have died from liver disease was 39 years.

Treatment includes cholestyramine, which reduces the enterohepatic circulation of protoporphyrins. There are now seven reports of liver transplantation in erythropoietic protoporphyria and as yet recurrent liver disease due to accumulation of porphyrins has not been seen, although follow-up is as yet limited.

REFERENCES

1 Fallon JD, Kvedar JC, Margolis RJ, Pathak MA. Erythropoietic protoporphyria presenting in adulthood. *Arch Dermatol* 1989;**125**:1286–7

2 Murphy G, Hawk JLM, Magnus IA. Late onset erythropoietic protoporphyria with unusual cutaneous features. *Arch Dermatol* 1985;**121**:1309–12

3 DeLeo VA, Mathews-Roth M, Harber LC. Erythropoietic protoporphyria. 10 years' experience. *Am J Med* 1976;**60**:8–22

4 Doss MO, Frank M. Hepatobiliary implications and complications in protoporphyria, a 20 year study. *Clin Biochem* 1989;**22**:223–9

5 Schmidt H, Snitker G, Thomsen K, Lintrup J. Erythropoietic protoporphyria. A clinical study based on 29 cases in 14 families. *Arch Dermatol* 1974;**110**:58–64

6 Murphy G, Hawk JLM, Corbett MF, Herxheimer A, Magnus IA. The UK erythropoietic protoporphyria register: a progress report. *Br J Dermatol* 1985;**113**(suppl 29):11

7 Lehmann P, Scharffetter K, Kind P, Goerz G. Erythropoetische protoporphyrie: synopsis von 20 patienten. *Hautarzt* 1991;**42**:570–4

8 Norris PG, Nunn AV, Hawk JLM, Cox TM. Genetic heterogeneity in erythropoietic protoporphyria: a study of the enzymatic defect in nine affected families. *J Invest Dermatol* 1990;**95**:260–3

(Accepted 28 June 1994)

Percutaneous endovascular covered stenting of a distal superficial femoral artery occlusion

Andrew T Goodwin MB BS[1] R Ian Swift BSc FRCS[1]
John D Lewis FRCS[1] Robert Wilkins FRCR[2]
Stephen J D Chadwick MS FRCS[1]

J R Soc Med 1995;**88**:477P–478P

CASE PRESENTED TO CLINICAL SECTION, 11 FEBRUARY 1994

Keywords: *peripheral vascular disease; superficial femoral artery; angioplasty; covered endovascular stenting; intimal hyperplasia*

Departments of [1]Vascular Surgery and [2]Radiology, Northwick Park Hospital, Watford Road, Harrow, London HA1 3UJ, England, UK

Correspondence to: Dr Andrew T Goodwin, 1 Madge Hill, Church Road, Hanwell W7 3BW, England, UK

Peripheral vascular disease is a common problem in the elderly population. This is commonly treated nowadays by percutaneous transluminal angioplasty (PTA). When PTA is used in the treatment of superficial femoral artery (SFA) occlusions 35% reocclude within 1 year[1]. Endovascular stents (uncovered) have been used to try to overcome this problem; however, results have been disappointing with reocclusion rates similar to PTA[2]. In order to try to reduce this problem covered endovascular stents have been developed. We report the first known case in the UK of one of these stents being used in the treatment of SFA disease.

CASE REPORT

A 73-year-old woman presented with intermittent claudication affecting her right calf after walking a distance of 200 yards. Physical examination revealed absent pulses below the femoral artery on that side. Investigations at that time demonstrated an ankle/brachial Doppler index of 0.35 (normal >1.0). She was commenced on aspirin 75mg and underwent an angioplasty of a long distal SFA occlusion.