PORPHYRIA CUTANEA TARDA
(A case report)

S. P. CHATTOPADHYAY * AND V. K. PAHWA †

Summary

A rare case of porphyria cutanea tarda symptomatica, in a 20 years old muslim female patient, treated successfully with phlebotomy, exchange transfusion and tablet periactin is described.

Porphyria cutanea tarda is a rare disorder[1,2,3,4], manifested by cardinal signs of cutaneous lesions, hyperpigmentation, hypertrichosis and evidence of liver disease. In a classical case, the skin is unusually sensitive both to light and to mechanical trauma. Abnormal fragility of epidermal and dermal tissue leads to blisters on the exposed skin areas, frequent ulceration and finally scar formation. Occasionally the scar formation is very severe, leading to mutilations and deformities of hands and feet. Abdominal pain and neurologic complications are absent in porphyria cutanea tarda symptomatica. Porphyria cutanea tarda hereditaria is a non-sex linked dominantly inherited disease with cutaneous lesions indistinguishable from porphyria cutanea tarda symptomatica, which is non hereditary.

Case Report

A 20 years old muslim female, wife of a service man, attended dermatology OPD with history of developing vesicles and blisters on dorsa of the hands since she became pregnant in February 1976. At the end of the first trimester of pregnancy in May 76, patient noticed that she was passing red coloured urine. She delivered a female child in Nov. 76. On interrogation she gave the history of appearance of vesicles over the hands since the age of 15-16 years on exposure to bright sunlight. She had also noticed hypertrichosis on the face and hyperpigmentation on the malar areas from the age of nine years. There was no history of abdominal pain and no history of similar disease in any other member of the family. There was no history of alcohol consumption or any drug ingestion except that of taking iron tablets for anaemia. Patient's father had died at the age of 35 years, after a short illness of uncertain nature.

On examination the patient was found to be a short statured thinly built lady with mild pallor prominent hypertrichosis of face and mild brownish pigmentation on cheeks and forehead. Skin of the face and extremities was inelastic. There were multiple small blisters and few scars on the sides of fingers. An ulcer 3 cm × 3 cm was present on the right hand. No jaundice

* Classified Specialist (Derm & Ven)
167, Military Hospital, C/o 56 APO
† Classified Specialist (Pathology),
Deptt. of Pathology
Armed Forces Medical College, Pune 1
Received for publication on 19—1—79.

or lymphadenopathy was noticed. Systemic examination showed liver to be enlarged 1.5 cms below right costal margin. It was soft and tender. No other abnormality detected. The urine was red coloured. Microscopic examination of direct and centrifuged deposit of urine showed no red cells and occult blood test was negative. Proteinuria was present. Porphobilinogen was also detected in urine. Spectroscopic examination showed increased coproporphyrin and uroporphyrins in urine. Blood haemoglobin levels varied from 10 gms to 11.00 gms % and serum bilirubin level was 0.5 mgm %. Liver function tests were within normal limits.

Patient was given tablet periactin 1 thrice a day and tablet vitamin C (100 mgm) thrice a day. Wycort ointment and saline dressings were given locally for the wound which showed partial healing. As the patient was pregnant a modified regime of phlebotomy and exchange transfusion was carried out. In the first sitting 300 ml of blood was withdrawn by venesection. Later 300 ml of blood was removed by venesection and 200 ml of fresh matched blood injected intravenously every third day for four weeks. A total of 1100 ml of blood was withdrawn by venesection. Wound on the hand healed and red colour of urine disappeared. Later, patient was put on tablet periactin ½ thrice a day, tablet vitamin C 100 mgm twice a day. Patient was advised to avoid exposure to sunlight. She has been observed for nine months and has shown no signs of any relapse. She is asymptomatic, urine has remained clear and prophobilinogen has been absent in urine.

Discussion

In the present case neurologic manifestations and pain in abdomen were absent and also history of hereditary transmission. Therefore this case fits in as a case of prophyria cutanea tarda symptomatica. Whereas porphyria cutanea tarda is more common in males[3] and usually begins in the fourth to sixth decade, our patient was a young female. No consistently effective treatment has been known for this disease. Epstein et al[6] first reported the beneficial effect of repeated venesections and removal of 2500 to 4500 ml of blood over 3 to 4½ months in prophyria cutanea tarda symptomatica which resulted in marked reduction of urinary uroprophyrin and clinical improvement. In one case induced chemical and clinical remission has persisted for six months without treatment. Rook et al[3] described venesection as an established effective therapy and advocated removal of 500 ml of blood every 2 to 3 weeks on 4 to 6 occasions and reported remissions which have lasted upto 12 months.

As our patient was pregnant, a modified regime of venesection and exchange transfusion was tried. A combination of venesection, exchange transfusion, periactin and vitamin C proved to be beneficial as shown by consistently negative urine for porphobilinogen and an asymptomatic period of follow up for 9 months.

Acknowledgement

We are thankful to Col Dr Sharma, OC MH Meerut for permitting us to use hospital records for publishing this paper.

References

1. Berg AG and Remington C: Diseases of porphyrin metabolism, Springfield, Charles C Thomas, 1962.

2. Tschudy DP, Magnus IA, Kallevas J: "The porphyria" from Dermatology in Internal Medicine, Fitzpatricx TB, Arndt KA, Clark WH, Eisen AZ, Scott EJ McGraw Hill 1971, P 1143.

3. Cairns RJ: Metabolic and nutritional disorders from Text Book of Dermatology; Rook A, Wilkinson DS, Ebling FJG Blackwell, 1972, P 1833.

4. Harrision TR : Principles of international medicine, 8th ed, McGraw Hill, 1977, P 660.

5. Dacie JV and Lewis SM : Practical haematology, 5th ed, Churchill Livingstone, Edinburgh, 1975, P 193.

6. Epstein JH, Redekar AG: Porphyrin cutanea tarda A study of effect of phlebotomy New Eng J Med 279 :1301, 1968.

Announcements . . .

Pediatric Dermatology Seminar VIII – Preliminary Notice

The 8th annual Pediatrid dermatology seminar will convene at the Eden Roc Hotel, Miami Beach, Florida, February 26 — March 1, 1981. It will be followed by a twelve day post-seminar tour to Tahiti and New Zealand with an optional extension to Australia.

For information, contact : Guinter Kahn, M.D., 16800 N.W. 2nd Ave., Miami, Florida 33169. (305-652-8600)

International Congress of Dermatology

The XVI International Congress of Dermatology will be held in Tokyo, Japan, May 23 to 28, 1982. The Congress includes a scientific program (special lectures, case presentations, advances in dermatology, symposia, courses, workshops, informal discussion groups, free communications, poster communications, Japanese Dermatological Association seminars, and a scientific exhibition) and social events (performance of traditional Japanese Kabuki drama, a concert with a world-famous conductor, a short suburban sightseeing tour, and programs for accompanying persons). The Congress site is the Hotel New Otani, Tokyo's prestige hotel which has been the site of many international congresses. English, French, Spanish, German and Japanese may be used in the Congress, and simultaneaus interpretation will be provided during the main educational sessions.

The First Circular including detailed information regarding registration, hotel accommodations and group travel is now available on request to :

Prof. Makoto Seiji, M.D., Secretary General, the XIV International Congress of Dermatology, C.P.O. Box 1560, Tokyo 100-91, Japan.

All interested persons are cordially invited to participate in the Congress.