

Steroids—A Possible Indication in the Treatment of Bantu Porphyria

BY

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Patients with Bantu porphyria complain mostly of the discomfort produced by the blisters on their fingers and hands, especially as fresh crops of lesions tend to recur continually. Treatment for this condition has been singularly disappointing in my patients in spite of advising them to give up alcohol and trying various drugs. Therefore as the following case seems to have done so well with steroids I feel it is worth reporting this success even though it is only a single case. No doubt steroids have been tried elsewhere for this disorder, but in case this regime may be of value I consider I am justified in recording this case.

Embree (1961) found that chloroquine appeared to be of some benefit to the African suffering from porphyria. After apparently causing a severe reaction in which greater amounts of porphyrins were passed in the urine and at the same time with an increase in the extent of the skin lesions, the disorder appeared afterwards to become quiescent and the patient improved. However Eales (1965) recorded that although patients with symptomatic porphyria apparently improved directly after chloroquine therapy he did not recommend its use since it appeared to aggravate the state of an already impaired liver.

The use of steroids did not seem to be contraindicated, especially as it has been held to be of value in liver disorders, such as infective hepatitis and cirrhosis and, as Bantu porphyria is always accompanied by signs of liver damage in Central Africa (Gelfand & Mitchell, 1957) I thought it worth a trial. Further, as in this disorder there is a sensitivity to light, I decided that for this reason too steroids might have a place in its treatment.

ILLUSTRATIVE CASE

Thomas Marima, an African aged about 45 years, was a Sergeant with many years service in the B.S.A. Police. He spoke good English, was intelligent and showed special interest in his condition. He complained of recurring blisters on his fingers and the backs of his hands for about one and a half years. He was practically never free of them and the longest interval he had been without new lesions was about two weeks. He also noticed that when he gripped

the handlebars of his bicycle, numerous small blood blisters formed on his palms and volar aspects of his fingers. A few blisters occasionally developed on his face, but this caused no great inconvenience. He was employed on patrol duties for which he had to use a bicycle — hence the concern about his hands. He readily admitted to drinking large quantities of African beer over many years but had never associated the blisters with this habit. Otherwise he enjoyed good health. He had a good appetite and experienced no indigestion.

It might be of interest to quote his doctor's words to me: "A/D Sgt. Thomas. This man complains of numbness of his middle finger. His fingernails seem to be degenerating and also he says that when he cycles a lot he gets small blisters filled with blood on his hands. His urine seems to indicate that he is a porphyric."

Thomas looked well except that he had the classical appearance we associate with Bantu symptomatic porphyria. His face was very dark and shiny and both conjunctivae were suffused. The dorsal aspects of his hands were of an intensely dark colour. Many of the finger nails, besides being broken, split or atrophic showed a marked heaping under the nail itself. There were many blisters on his hands and fingers as well as a few raw areas on the backs of his hands as a result of previous ones that had burst.

On examination the patient proved to be well built. His liver and spleen were not palpable. His blood pressure was 110/70. The urine had a dark sherry look and uroporphyrins were reported as being present. Faecal porphyrins were also noted in the stools. His haemoglobin was 115 per cent. and the total white cell count was normal. There was no enlargement of the liver or spleen.

TREATMENT

The patient was told that the blisters as well as his dark skin and the nail dystrophy were probably related to alcohol and that he should stop drinking. I explained that I did not know of any specific remedy that would help him, but I would like him to try the effects of prednisolone and that if he took two tablets regularly and gave up alcohol entirely he might find an improvement and the blisters would stop appearing. As I mentioned previously I had reason to believe that steroid tablets might benefit him, although I could not be certain. Accordingly, on 27th January, 1965, he was put on 5 mg. t.i.d. prednisolone and told to remain on this treatment until the end of February. Thereafter he was

instructed to reduce the dose to one tablet twice a day. He left hospital and carefully carried out these orders. When seen on 28th May, 1965, he was most satisfied with the state of his fingers. He found that shortly after leaving hospital the blisters ceased appearing on his hands. He was seen again on 18th June. At the end of July it was decided to stop his prednisolone for one month and when he was seen in August and again in September he was still most pleased, for, whereas previously he was hardly ever free of the blisters, he was now completely well. He was seen again in December, 1965, when his progress seemed to have been maintained. He had avoided all alcohol from the time he was asked to give up taking it.

COMMENT

In this instance the patient was taken off his alcohol and at the same time given a course of steroid therapy. I doubt whether removal of the alcohol was the reason for his improvement as this measure did not appear to be effective in my other cases. And in any event we have met patients with the disease who deny having taken alcohol or drinking in excess and yet the lesions have continued to appear on the hands. I am not sure why steroids should assist, if indeed they really did, but, as many patients with Bantu porphyria show varying degrees of liver damage or even cirrhotic changes, it occurred to me that this was a good reason for employing them. Secondly as the skin is photosensitive in the disease the lesions may have an allergic basis.

REFERENCES

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