LETTER TO THE EDITOR

How long should we treat porphyric patients?

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Acute attack in hepatic porphyrias (acute intermittent porphyria, variegate porphyria, and hereditary coproporphyria) is a rare, but potentially life-threatening, condition. Although every physician should be aware of these metabolic syndromes, the rarity and broad spectrum of the symptoms of porphyrias often lead to a delay in diagnosis. I read with interest an illustrative case report that recently appeared in this journal (1), in which De Block and colleagues reported a 38-year-old woman who suffered acute intermittent porphyria with premenstrual attacks. Intravenous hypertonic glucose with i.v. haem arginate (Normosang) brought about temporary relief of the symptoms. Luteinizing hormone-releasing hormone (LH-RH) analogue combined with a low-dose oestrogen patch was needed to prevent cyclic attacks. When LH-RH analogue treatment was withdrawn, 17 months after its commencement, premenstrual attacks recurred. Another course of combination treatment was therefore reinstituted, and the patient had not required admission to hospital for 3 years (1). It is my opinion that such combination treatment is an excellent way of giving LH-RH analogue in order to avoid drug side effects.

Very recently, we also reported a patient with hereditary coproporphyria, whose premenstrual exacerbation was successfully treated with LH-RH analogue alone (2). Bone demineralization occurred, because we did not include oestrogen in our treatment, and the drug was tapered carefully. Spontaneous menstruation recovered 2 months after cessation of LH-RH analogue

treatment, which was given over a total of 64 months. Surprisingly, since then, the patient has experienced only minor symptoms, for a period of more than 1 year. Clonazepam, to prevent epileptic seizure, is the only medication she is now receiving. We should be aware that the natural course of porphyria is variable and that spontaneous remission frequently occurs (3, 4). As we do not know what is (are) the principal determinant(s) leading to symptomatic porphyria, I believe that we should always be sensitive as to whether current treatment given to each patient might be safely withdrawn.

References

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