Steroid-induced benign intracranial hypertension

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Summary

A 41-year-old man whose systemic lupus erythematosus (SLE) had been successfully treated for 15 months with a daily maintenance dose of 5 mg prednisolone, developed benign intracranial hypertension (BIH) when the steroid was increased to 60 mg daily for recrudescence of SLE symptoms. The BIH remitted when the steroid was discontinued.

Introduction

Relationship of steroids with benign intracranial hypertension (BIH) is well documented. It has been reported to occur either on withdrawal of steroids or change to another preparation (Walker and Adamkiewicz, 1964; Hagberg and Sillanpää, 1970; Weisberg, 1975). Development of BIH while on maintenance steroid therapy or increase in its dose is an uncommon phenomenon in adults, although well reported in paediatric populations (Cohn, 1963; Gordon and Kelsey, 1967). Such a case is now described from an adult who was being treated for systemic lupus erythematosus (SLE).

Case report

A 41-year-old man with SLE had had this condition successfully controlled for 15 months with a daily maintenance dose of 5 mg prednisolone. When he had a recrudescence of symptoms (arthralgia, purpuric spots; strongly positive L.E. cells and antinuclear factor), the daily dose of prednisolone was increased to 60 mg. Within a few days, arthralgias and purpuric spots cleared, but he developed intense headache with occasional vomiting, and was admitted to hospital. Physical examination revealed

bilateral papilloedema. There was no focal neurological deficit and no signs of meningeal irritation. Steroid-induced benign intracranial hypertension (BIH) was considered as the most likely diagnosis, although SLE and an intracranial space-occupying lesion were also thought to be the possible causes of papilloedema. Haematological studies, urinalysis, blood urea nitrogen, serum creatinine, X-rays of skull and chest were normal. Serum albumin was 21 g/l and globulin 55 g/l. Serum protein electrophoresis showed mild rise of γ -globulin. Perimetry revealed peripheral constriction of fields. Cerebrospinal fluid was clear, the pressure in the lateral recumbent position was 240 mm of water; CSF cytology and biochemistry were within normal limits. Echo-encephalography did not show any shift of midline and electroencephalogram revealed generalized theta activity without localization. CAT scan was normal, ventricular size on both sides was within normal limits.

While in hospital, the patient developed diplopia and was found to have bilateral sixth nerve palsy. Visual acuity at this stage was 6/18 in both eyes. Prednisolone was gradually tapered and stopped. Azathioprine was given to control the SLE and oral glycerol to reduce intracranial tension. The patient gradually improved. Headache disappeared completely over a period of 2 months, papilloedema regressed, visual acuity improved to 6/9 in the left eye and 6/6 in the right. Lateral rectus palsy disappeared on both sides, and the SLE symptoms were absent while on azathioprine therapy. The L.E. cell phenomenon became negative, although the antinuclear factor still remained faintly positive.

Discussion

Although recognized since 1893, a clear description of benign intracranial hypertension has been available only since 1937 (Davidoff and Dyke, 1937;

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Dandy, 1937). Johnston and Paterson (1974) suggested the following diagnostic criteria for BIH: the symptoms should be those of raised intracranial pressure alone; the clinical signs should be those of intracranial hypertension; the major investigations, particularly contrast radiology, should be entirely normal apart from non-specific evidence of raised intracranial pressure; there must be a measurable increase in CSF pressure but the fluid must be of normal composition. The present patient fulfils all these 4 criteria.

Two children are reported to have developed BIH while on long maintenance doses of triamcinolone (Cohn, 1963). Green, Cleveland and Wilkins (1961) reported 4 children, with the congenital adrenogenital syndrome, who were receiving triamcinolone therapy and who developed blurring of the optic discs.

In a review of 28 cases it is observed that the occurrence of BIH is related either to the withdrawal or reduction in dose of a steroid, or to a change to another drug (Walker and Adamkiewicz, 1964).

In the present case, BIH appeared when the daily dose of prednisolone was increased to 60 mg, and the symptoms and signs regressed gradually when the steroid was tapered. This clearly indicates that the development of BIH was related to the increase in the dose of steroid.

Benign intracranial hypertension has also been

reported as a manifestation of SLE (Bettman *et al.*, 1968; Weisberg, 1975). At one stage in the present case, SLE was considered as the cause of the BIH, but the evidence very strongly favoured it to be steroid-induced.

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