Departments of Neurology and Neurosurgery, Middlesbrough General Hospital

Familial Benign Intracranial Hypertension

By

J. G. Howe, M. Saunders, and P. R. R. Clarke

Summary

The case histories of two sisters with benign intracranial hypertension are reported. Reference is made to the other recorded familial examples of this syndrome.

Benign intracranial hypertension is increasingly recognized as a cause of raised intracranial pressure. Although there are a number of published series of cases (Foley 1955, Bradshaw 1956, Boddie, Banna, and Bradley 1973), the occurrence of familial examples of this syndrome is rare (Boucheit, Burton, Haag, and Shaw 1969, Wells 1973). The purpose of this paper is to report benign intracranial hypertension in two sisters.

Case 1. L. R., a twenty-seven year old housewife, was admitted to Middlesbrough General Hospital on 27. 11. 1969 with a history of one week's severe generalized headache associated with dizziness and deteriorating vision. She had given birth to her first child eight months previously and had been in good health. There was no history of menstrual irregularity and she had received no medication. There was no relevant family history at the time amongst her nine brothers and sisters. Both parents were well.

At the time of her admission her visual acuity was reduced to light perception. She was not obese and there were no focal neurological signs. The day after admission she became totally blind. She was treated with dexamethasone with relief of her headaches. An air ventriculogram was performed which revealed that the brain was bulging and the ventricles symmetrical but small and pinched.

After one month the papilloedema subsided but the patient was left totally blind. During a six months period following her investigation the patient gradually put on weight despite cessation of the dexamethasone.

Case 2. The thirty-five year old married sister of L. R. was admitted to Middlesbrough General Hospital on 29. 9. 1972 with a one week history
of headache and vomiting. In the past she had had frequent chest infections associated with partially reversible airways obstruction. She had noticed an increase in weight of approximately one stone in the previous year. Her three pregnancies had all been normal and menstruation was normal. There was no history of steroid medication nor had she taken the contraceptive pill or tetracycline.

Examination revealed moderate bilateral papilloedema. The visual acuity was 6/6 in both eyes. A lumbar air encephalogram was attempted but air did not enter the ventricles. She was treated with dexamethasone, her symptoms subsided, and she was discharged. She was re-admitted on 2. 11. 1972 with a recurrence of her symptoms. She was again given dexamethasone and a ventriculogram performed. This revealed slightly dilated symmetrical ventricles. Dexamethasone had little effect on her symptoms which were relieved by lumbar puncture. A spontaneous remission eventually occurred and she was discharged home on 2. 1. 1973 at which time the visual acuity was 6/9 in both eyes.

On 6. 4. 1973 she was re-admitted with a relapse. On this occasion it was decided to treat her with a ventriculo-cardiac shunt using a Pudenz valve. She made satisfactory progress after the procedure and is at present well although now very obese.

Discussion

Boucheit, Burton, Haag, and Shaw (1969) reported the occurrence of intracranial hypertension in two sisters occurring simultaneously. In this instance both sisters were obese. The only other familial occurrence that we are aware of is the family studied by Wells (1973). This involved two generations, a mother and three daughters being affected. In this family pregnancy was associated with the onset of the condition in one case, although the patient was also obese. The menarche was related to the onset in another sister who subsequently relapsed in pregnancy.

In the two sisters described in this report symptoms presented three years apart. Case 1 became very obese following the onset of her illness and this could not be attributed to the use of dexamethasone. This patient became blind after only one week’s history re-emphasising that the syndrome is far from benign. In the instance of Case 2 the patient had put on at least one stone in weight in the year preceding her symptoms. Although the first patient appeared to respond to dexamethasone there was no convincing response in the second case.

In the recorded families obesity appears to be a common link although in the family described by Wells other endocrine factors appeared relevant as well. In this respect there is no difference in the usual factors regarded as important in sporadic cases. It is possible that more detailed studies of familial cases may throw further light on the underlying mechanisms responsible for many cases of this syndrome.