Pseudotumor Cerebri in Children Secondary to Administration of Adrenal Steroids

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Pseudotumor cerebri is a familiar clinical picture to neurosurgeons but the etiology remains obscure. The presence of papilledema without signs of a localized mass lesion, high spinal-fluid pressure, normal protein and absence of cells in the spinal fluid may suggest this syndrome but few physicians would be willing to accept such a diagnosis without a normal air study or angiogram. Some of these cases have been shown to be associated with an obstruction of one of the dural sinuses, as in patients with infections of the middle ear. The cause is not apparent in the majority of cases but in others, an endocrinologic dysfunction appears to be an underlying factor. In support of such a hypothesis is the frequent occurrence of this syndrome in obese, young women with dysmenorrhea or during pregnancy. A disturbance of the pituitary-adrenal axis also is suggested by the number of patients reported with Addison's disease, galactorrhea, and decreased urinary excretion of ketosteroids. Paterson et al.\(^7\) presented an excellent review of the subject and concluded that the underlying cerebral edema may result from several causes, particularly functional hyperpituitarism.

The occurrence of pseudotumor cerebri in children, without disease of the middle ear, was quite a rarity until the use of steroids became commonplace. Dees and McKay\(^3\) were among the first to report the occurrence of benign intracranial hypertension during the treatment of asthmatic children with adrenal steroids. They described 3 patients with varying signs of intracranial pressure including papilledema and cranial-nerve palsies.

One child was receiving Triamcinolone, another Prednisone, and the third, Prednisolone. Valentine\(^4\) reported a child with nephrosis who received 4 mg. of Triamcinolone three times a day. Six weeks later the patient began having headaches and was found to have papilledema. Pneumoencephalograms and carotid arteriograms were normal. After changing to Prednisone, the symptoms subsided. Laurance\(^5\) described a child with asthma who was receiving 4 mg. of Triamcinolone per day. She was discovered to have papilledema and a ventriculogram was interpreted as normal. A second patient was a 5\(\frac{1}{2}\)-year-old male with asthma who also had papilledema 15 weeks after being placed on 4 mg. of Triamcinolone per day. After switching to Prednisolone the papilledema disappeared. Benson and Pharoah\(^1\) described 2 cases. One was that of an 8-year-old child who had papilledema while taking Prednisolone. The youngster had been given 10 mg. per day for a year and when the dose was cut to 5 mg., symptoms of increased intracranial pressure appeared. A ventriculogram was reported as normal. The second case, that of a 5\(\frac{1}{2}\)-year-old boy with eczema, is most unusual, as he was treated only topically with hydrocortisone ointment and after 16 months papilledema developed. Green et al.\(^5\) reported 4 cases of blurred discs and 1 case of pseudotumor cerebri in their discussion of therapy with Triamcinolone in the adrenogenital syndrome. Good et al.\(^4\) reported convulsions both with and without hypertensive encephalopathy in youngsters on prolonged administration of cortisone and adrenocorticotropic.

The discovery of 3 cases during the past 2 years of steroid-induced pseudotumor cere-
Pseudotumor after Rx with Adrenal Steroids

785

bri in the Children’s Hospital prompted this report. Recently a near tragedy occurred during attempted ventriculography in a 6½-year-old boy.

Case 1. C.R. (¶340888) had been under treatment by an ophthalmologist for strabismus involving the right eye since the age of 2. He had been receiving Triamcinolone, 4 mg. per day for about 3 months because of chronic, generalized eczema. A month after beginning this medication it was noted that his strabismus was more pronounced and he became rather poorly coordinated. On several occasions he complained of transient loss of vision and mild headache.

Examination disclosed unsteadiness of his gait with inability to walk a straight line. There was no ataxia of his extremities or dysdiadochokinesia. He had paralysis of both lateral rectus muscles. There was bilateral papilledema with small hemorrhages about the right disc. Roentgenograms of his skull showed questionable broadening of the coronal sutures; the film of the chest was unremarkable. A posterior-fossa neoplasm was suspected and he was prepared for operation by the intramuscular administration of 100 mg. of cortisone the day before and on the day of operation.

Operation. Bilateral occipital burr holes were inserted under endotracheal anesthesia. He seemed to have a partial obstruction of the airway which could not be corrected by altering the position of the endotracheal tube. The ventricles were so small they could not be cannulated. The anesthetist noted that the child felt warm and his rectal temperature was found to be over 108°F.; the exact temperature was not determined because the thermometer did not register above this figure. The occipital wounds were closed rapidly.

Course. Examination with the surgical drapes removed disclosed his pupils to be dilated and fixed. His entire body was rigid and he had bilateral extensor plantar responses. He was given 100 mg. of hydrocortisone intravenously and placed on a cooling blanket. Within 15 minutes his hyperthermia was converted to a hypothermic state. His serum sodium was 128 mEq. per l. After correction of the hyponatremia, the rigidity, coma, bilateral Babinski’s sign and dilated pupils disappeared. During the following week his cortisone was withdrawn gradually without ill effect. The papilledema, hemorrhages and paralysis of the 6th nerves disappeared gradually during the next month.

Case 2 (reported by Calcagno and Rubin3). C.G. (¶244573), a 2-year, 10-month-old girl with nephrosis, was treated with Triamcinolone for 10 months when irritability, vomiting and strabismus developed. Examination revealed bilateral papilledema and palsy of the 6th nerves. A ventriculogram was interpreted as normal.

Two months after withdrawal of the steroids, she had no residual neurologic difficulty.

Case 3. C.C. (¶269388), a 4-year-old female child, was admitted recently because of bilateral palsy of the 6th nerve, headache and vomiting. These symptoms occurred after several months of Triamcinolone therapy for rheumatoid arthritis. The eyegrounds disclosed moderate blurring of both discs but no hemorrhages or exudates. The steroids were withdrawn gradually and the headaches and eye signs began improving. Air studies were not performed. During the next 2 months her strabismus disappeared and her eyegrounds became normal.

Discussion

Case 1 almost expired in an Addisonian crisis despite rather large doses of steroids preoperatively. The occurrence of papilledema and cranial-nerve palsies in a child who has been receiving steroids should suggest pseudotumor particularly if there are not clear-cut signs of ataxia. Avoidance of operation under these conditions may be lifesaving.

Summary

Three children with pseudotumor cerebri induced by prolonged administration of steroids are presented. This syndrome should be familiar to neurologic surgeons so that operation may be avoided. The patients had papilledema and palsy of the 6th nerves but little ataxia. All recovered after withdrawal of the medication.

Addendum

Since this paper was submitted for publication, an article has appeared by Greer, M. Benign intracranial hypertension. II. Following corticosteroid therapy. Neurology, 1963, 13:439-441. The author reported 5 cases of pseudotumor cerebri induced by administration of steroids.

Generic and Trade Names of Drugs

Prednisolone—Ataraxoid, Cordex,
Meticortelone
Prednisone—Arthralgen, Meticorten,
Sigmagen
Triamcinolone—Aristocort, Kenacort
