Lumbar Subarachnoid Fallopian Shunt in the Treatment of Pseudotumor Cerebri in a Patient With Narcolepsy

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THE PURPOSE of this paper is to report the simultaneous occurrence of pseudotumor cerebri and narcolepsy. Review of the literature for the last 10 years failed to disclose reports of this combination.

The term 'pseudotumor cerebri' is used to distinguish a syndrome of increased intracranial pressure in the absence of identifiable causes such as intracranial mass lesion, obstruction of ventricles, veins or sinuses, intracranial infection, hypertensive encephalopathy, chronic carbon dioxide retention and certain metabolic-endocrine conditions. It, in effect, represents a heterogenous group of disorders with varying etiologies. Most often the pathogenesis of this syndrome is ill-defined. It is described as benign because, usually, spontaneous recovery can be anticipated. Loss of vision may be a significant factor during the course of this disease.

Narcolepsy is a chronic disorder characterized by recurrent and excessive drowsiness or sleepiness from which the patient may be easily aroused. Frequently there are associated cataplexy, sleep paralysis and hypnagogic hallucinations. There is a high familial incidence of this ailment. The stage of rapid eye movements frequently occurs at the onset of narcoleptic sleep rather than approximately after ninety minutes as occurs normally. Physical and laboratory examinations are normal, with the exception of a frequently low basal metabolic rate in the primary syndrome.

CASE REPORT

In the spring of 1960 this 43-year-old female experienced attacks of numbness over the back of her head with diplopia. Episodes of double vision lasted from two to five minutes. The longest symptom-free interval was two weeks. She was admitted to another hospital and lumbar puncture showed a high CSF pressure. Funduscopic examination revealed retinal hemorrhages. Subsequent lumbar punctures revealed near normal pressure. Her symptoms ceased 18 months after onset—she was pregnant for five of these months. In February 1965 she complained of the same symptoms. In January 1966, she

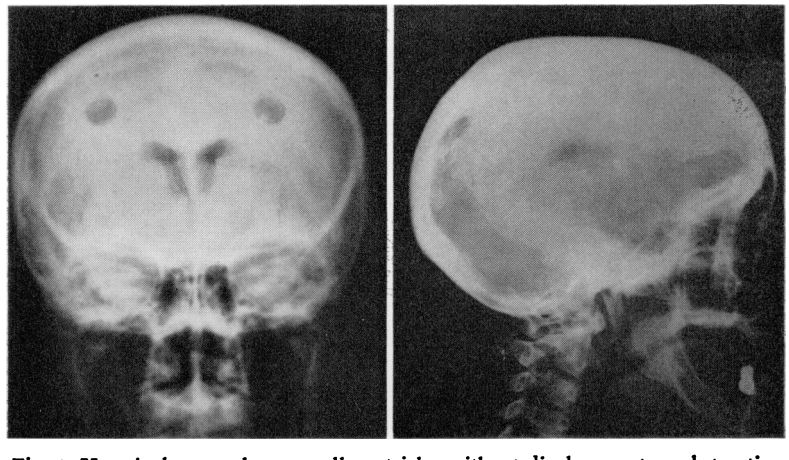

Fig. 1. Ventriculogram shows small ventricles without displacement or obstruction characteristic of pseudotumor.

noted the onset of transient blindness of the left eye, lasting from 10 to 20 minutes; this was associated with incoordination of fine finger movements bilaterally. Of importance in the past history was that from 1940 to 1961 she was able to stay awake for periods of only five hours. In 1961 this waking interval shortened to one and one-half hours. During that period and subsequently she has complained of slowly sinking to the floor when seized by uncontrollable laughter—she was awake and continent during these episodes. She was able to rise after controlling the emotion. In 1961 she was placed on Dexedrine, 10 milligrams daily. This was discontinued two years later when she developed visual hallucinations. At present, without specific medication, she is able to remain awake for periods of four to five hours. She continues to have cataplectic attacks.

Physical examination revealed a mildly obese white female with one diopter of papilledema bilaterally. The remainder of the examination was unremarkable.

She was admitted to Arlington Hospital in March 1966 and a ventriculogram (Fig. 1) showed small ventricles without any evidence of obstruction or displacement. She was given a trial of 20 per cent Osmitrol, 500 cc's intravenously, daily for two weeks and was discharged on Prednisone, 20 milligrams a day and a low sodium diet. On this program she soon became cushingoid.

She was readmitted in August of 1966 complaining of diplopia. At that time physical examination revealed a weight of 175 pounds, height of 63 inches, a blood pressure of 130 mm. Hg. systolic and 90 mm. Hg. diastolic, a pulse rate of 110 per minute and bilateral papilledema of 1 diopter. The examination was otherwise noncontributory. Laboratory studies showed a hematocrit of 32 per cent, WBC count of 13,300 mm^3, Segs—67, Bands—1, Lymphocytes—30, Monos—2, ESR—26, BUN—11, glucose tolerance test: fasting 96 mgs%, 1 hour—170 mgs%, 4 hours—90 mgs%, 5 hours—74 mgs%, 6 hours—86 mgs%. Sodium 143 mEq, potassium—3.2 mEq, chlorides 107 mEq. Carbon dioxide combining power—24 mEq. Urinalysis was normal with specific gravity of 1.012. Blood serology was non-reactive, serum iron—55 micrograms per cent, 24 hour urine volume was 1000 cc's, creatinine 1.3 gms., 17 ketosteroids 19 mg, 17 hydroxysteroids 8.99 mg, PBI—4.8 micrograms per cent. Chest x-ray was normal and skull series revealed cranial defects at the sites of previous trephination. Lumbar puncture showed an opening pressure of 280 millimeters of water, protein of 39 milligrams per cent, no white blood cells and 4 red blood cells. The serologic test and colloidal gold curve were normal. A pneumoencephalogram showed the ventricular system to be small and not displaced. A right retrograde brachial arteriogram was within normal limits. She was discharged and subsequently re-admitted for the relief of increased intracranial pressure by lumbar puncture. The CSF pressure in September of 1966 was 250 mm of water, in October it was 200 mm of water. Visual fields in December of 1966 showed increased size of the blind spots (Fig. 2). Gliosis of the optic nerve heads and hemorrhages were noted on funduscopic examination at that time (Fig. 3). Bilateral subtemporal decompression was performed to prevent further damage to her sight. The cranial defects were approximately two and one-half inches by one and three-quarter inches. Following surgery the scalp at the decompression sites bulged and were non-pulsating. Intracranial pressure remained elevated.

In January of 1967 she was readmitted because of headaches of sudden onset. Lumbar puncture showed CSF pressure to be 250 mm of water. In March of 1967 the pressure was 310 mm of water and in April it was 320 mm of water and in May it was 280 mm of water. Pneumoencephalogram, done in May, showed no new findings. Visual field examination revealed an increased size of the blind spots since December of 1966. Papilledema had not resolved. A lumbar subarachnoid fallopian shunt was carried out, using an 18 gauge silicone tubing. The sub-temporal decompression sites gradually became flat during the next week. Repeat visual field examination in August of 1967 showed normal blind spots. Papilledema had subsided and visual acuity was normal.

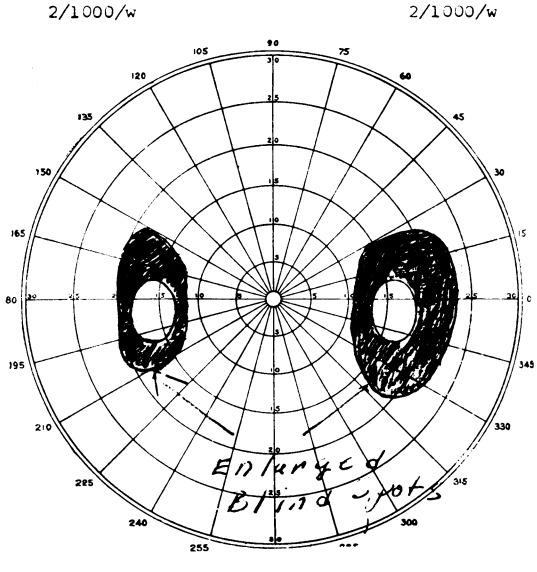

Fig. 2. Visual Fields, O.D. and O.S. showing enlarged blind spots resulting from chronic papilledema.

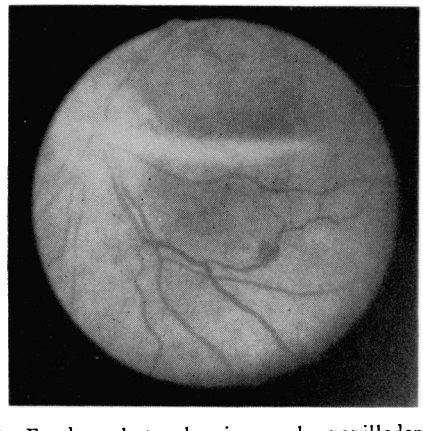

Fig. 3. Fundus photo showing early papilledema and retinal hemorrhage at 5 o'clock.

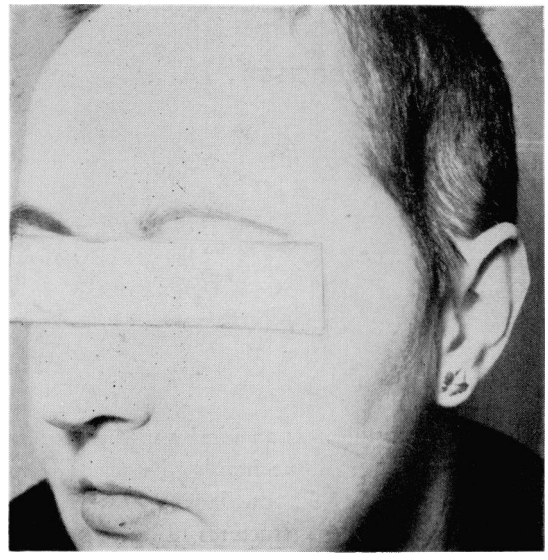

Fig. 4. Sunken temporal scalp at the site of the previous subtemporal bone removal illustrates good decompression afforded by the shunt.

The patient resumed normal household activity and desired cosmetic and protective coverage of the decompression sites. In January of 1968 right tantalum cranioplasty was performed with obliteration of the cranial defect (Figs. 4 & 5) as the first stage of a two stage procedure. Nine months following the shunting procedure the patient was asymptomatic excepting for her narcolepsy.

DISCUSSION

In this patient narcolepsy antedated the onset of pseudotumor cerebri. Diuretic agents, lumbar punctures, and sub-temporal decompression failed to provide relief. Lumbar subarachnoid fallopian shunt was successful in decompressing the optic nerves and thus facilitated the return of normal visual acuity. Pseudotumor cerebri runs an unpredictable course which ultimately resolves spontaneously in the majority of cases. The symptoms of narcolepsy were not affected by the treatment for her elevated intracranial pressure. Lumbar subarachnoid fallopian shunt was chosen in preference to a ventriculo-atrial shunt because of the technical difficulties which would have been encountered with small ventricles. Post-lumbar puncture headache has been ascribed to leakage of cerebrospinal fluid into the epidural space thus facilitating downward traction on the pain sensitive structures in the posterior fossa which are unsupported, especially the veins. It is interesting to note that this patient had a continuous drainage of spinal fluid into the peritoneal cavity and did not complain of headaches. We consider the association of pseudotumor cerebri and narcolepsy in this patient to be fortuitous.

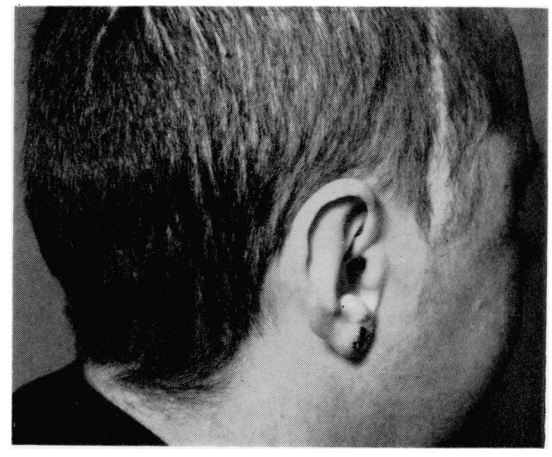

Fig. 5. Tantalum cranioplasty covers previous bony defect with good cosmetic result.

SUMMARY

A case of narcolepsy and pseudotumor cerebri with progressive visual changes has been described. The visual field changes were successfully reversed by a lumbar subarachnoid fallopian shunt after steroid therapy, diuretics, repeated lumbar punctures and subtemporal decompression failed to give relief.

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