DYSTONIA MUSCULORUM DEFORMANS ALLEVIATED BY CHEMOPALLIDO THALAMECTOMY AND SUBSTANTI A NIGRALYSIS

CASE REPORT

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(Received for publication October 12, 1959)

Dystonia musculorum deformans, as the name implies, is generally a crippling and progressive neuromuscular disorder. It results in grotesque postures and finally in permanent skeletal changes, such as rotokyphoscoliosis. It is primarily a disorder of childhood and adolescence and therefore the more devastating. With the use of chemopallidectomy and chemothalamectomy introduced by Cooper,1 the majority of such patients can be greatly improved if not "cured" of their dystonic syndrome.

In our 6 cases, 4 patients have been completely relieved and 1 considerably improved, and 1 has received no benefit from surgery. One patient required bilateral thalamic lesions which incidentally were done simultaneously; in a second patient, bilateral pallidal lesions were made at 4 separate operations; in a third, bilateral pallidal and unilateral thalamic lesions were produced in 3 operations; a fourth has had unilateral thalamic lesions on 2 occasions with 75 per cent improvement and soon will have a pallidal lesion made. The fifth patient has been the most difficult to treat, requiring bilateral pallidal and unilateral thalamic lesions done in 6 operations. And, in addition, destruction of the left substantia nigra by a new surgical technique—substantia nigralysis.3

This particular case is worthy of detailed review.

CASE REPORT

Dystonia musculorum deformans alleviated after 7 operations over a 21-month period by left chemopallidectomy and substantia nigralysis, and right chemopallidothalamectomy.

A 38-year-old Jewess of Russian descent, had gradual onset of involuntary and uncontrollable turning of the head to the left in September, 1956. Within a month she noticed a "pulling sensation" in the right arm which soon developed into dystonic posturing and spasms causing inability to hold on to small objects. By December the axial musculature and right lower extremity had become similarly involved with dystonia. It became necessary for her to wear a cervical brace constantly and use a cane to get about.

Examination. The patient was an oriented, intelligent individual with a passive aggressive disorder of character. The cranial nerves functioned normally. In spite of the cervical brace which she wore constantly the head tilted and twisted to the left. It was impossible to rotate it beyond the midline because of increasing spasms of muscles. The right arm was held in a slightly flexed hyperpronated posture with marked flexion of the fingers. Passive supination caused aggravation of the dystonic spasms and pain in the muscles. Moderate scoliosis of the thoracic spine was accompanied by severe lumbar lordosis, most prominent when the patient was in an erect position. The right hip and knee remained slightly flexed with the foot strongly everted. This posture was exaggerated when the patient walked. Although the muscle tone in the right side was increased, no abnormal reflex changes were elicited on either side. Dystonia was not observed in the left extremities. Except for some mild hypalgesia on the right side sensory findings were normal.
From psychiatric examinations and Amytal interviews over a week's period it was concluded that the patient suffered from dystonia musculorum deformans rather than any primary psychogenic disorder, thus substantiating the opinion of those neurologists who had examined her.

Laboratory studies including complete blood count, urinalysis, serum electrolytes and cerebrospinal fluid disclosed no abnormalities. An electroencephalogram was normal. The electromyogram (using disc electrodes) demonstrated fairly constant, extremely high potential activity on the right side.

Operation 1. On Oct. 31, 1957, using transtemporal approach under general anesthesia, two plain Cooper cannulae were placed in the left globus pallidus. The first was in a coronal plane at the posterior wall of the foramen of Monro; the second 5 mm. behind the first. Following injection of 0.3 cc. of 1 per cent procaine into the second cannula the dystonic symptoms abated completely. Therefore 0.8 cc. Ethocel* was injected fractionally over an hour. Subsequently there developed a mild right facial weakness and loss of memory lasting about 5 days. The torticollis and spasms in the right hand and foot returned on the 8th postoperative day. An additional 0.2 cc. of Ethocel in the posterior cannula relieved the dystonia except in the right foot. This procedure was again accompanied by mild facial paresis and loss of memory for 3 days. On the 15th postoperative day 0.3 cc. of 1 per cent procaine injected into the anterior cannula abolished her residual dystonic symptoms; 0.5 cc. of Ethocel was then injected over a 50-minute period.

On discharge from the hospital on Nov. 16, 1957 neurological findings were within normal limits except for slight right facial weakness.

Course. After leaving the hospital she did well, even returning to her former work as a secretary for a few days. Then, during the 2nd week in December, 1957, dystonic symptoms developed on the left side, with torticollis to the right. Except for the fact that the left foot had become strongly everted rather than everted, as had been the case on the right side, the neurologic findings were as described during her first hospitalization. An electroencephalogram on Dec. 27, 1957 was normal, of the low fast-voltage type.

Operation 2. On Jan. 2, 1958, a Cooper cannula was placed by the temporal approach into the right globus pallidus 1 cm. behind the foramen of Monro. Injection of 0.6 cc. of 1 per cent procaine failed to relieve the contralateral dystonic symptoms. The cannula was removed.

Operation 3. This was performed on Jan. 7, 1958. Two cannulae were placed in the globus pallidus in a position similar to those for the left chemopallidectomy, namely in the coronal plane of the foramen of Monro and 5 mm. posterior to it. Immediately following the injection of 0.3 cc. of 1 per cent procaine through the anterior cannula the dystonic postures and spasms ceased. Accordingly, 0.5 cc. of Ethocel was injected over a 25-minute period. The patient had some difficulty in recognizing her physicians and friends and in addition showed lack of affection toward her sons. Dystonic movements began to recur in the left hand. These were abolished by an additional 0.5 cc. of Ethocel. However, the patient experienced severe loss of memory for recent events, which cleared in 72 hours. Eight days after the last injection dystonia had recurred to a moderate degree. Instillation of 0.5 cc. of 1 per cent procaine through the posterior cannula abolished these recurrent symptoms. Therefore, 1.0 cc. of Ethocel was injected over a 60-minute period. She became confused and disoriented. This mental clouding cleared over a 7-day period. However, mild dystonia then returned to the left side.

The patient was discharged from the hospital on Feb. 2, 1958, improved, but with definite reliefs of the dystonia and some blunting of affect. Voluntary muscular strength and sensation were normal. The deep tendon reflexes on the left were somewhat increased over the right side and a positive Babinski's response was present on the left.

Course. The patient was re-admitted to the hospital for the 3rd time on March 23, 1958, because of increasing symptoms of dystonia in the left extremities and torticollis to the right. The symptoms were particularly prominent in the left leg and foot and lumbar spine. The

* Supplied by Ciba Pharmaceutical Company; a solution of 8 per cent celloidin and 95 per cent ethanol.