ISOLATED UNILATERAL SPINAL ACCESSORY NERVE PALSY OF OBSCURE ORIGIN

A REPORT OF THREE CASES

BY

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Injury to the spinal accessory nerve during operation for block dissection of cervical glands is probably the commonest cause of unilateral paralysis of the sternomastoid muscle. In the neck the nerve may be injured by direct compression from enlarged glands and from tumours or abscesses, and of course it may be damaged in penetrating wounds. Within the skull, in the posterior fossa, it may be involved in tumour, abscess, or aneurysm formation. Occasionally a syphilitic process within the skull may implicate the nerve as it runs from the foramen magnum to the jugular foramen. Within the spinal cord or medulla the cells of origin of this exclusively motor nerve are not infrequently involved in degenerative processes such as motor neurone disease and syringomyelia. The same cells are also liable to destruction by the virus of anterior poliomyelitis.

The sternomastoid and trapezius muscles often undergo atrophy in the muscular dystrophies and especially in the affection called myotonia atrophica.

During the past five years I have encountered three patients in whom the sole lesion was an isolated spinal accessory nerve palsy. Weakness and wasting of the sternomastoid muscle and of the upper fibres of the trapezius on the same side were the only abnormalities.

Case 1

A soldier aged 25, serving in the Middle East, was referred to hospital as a possible case of "shoulder-girdle neuritis" (Spillane, 1943). He had always enjoyed good health, but for a year past had been troubled with an aching pain about the right shoulder. This was never acute and did not involve the arm. It was ill defined and dull in character, nearly always

following prolonged exercise. He had noticed that his neck was asymmetrical.

There was considerable wasting and weakness of the sternomastoid and upper part of the trapezius on the right side. The right shoulder had dropped (Fig. 1). The nervous system was otherwise normal. X-ray examination of the skull base and cervical vertebrae revealed no abnormality. The C.S.F. was normal, and the Wassermann reaction in the blood and C.S.F. was negative.

During his stay in hospital repeated questioning and examination

disclosed nothing to suggest that the problem was one of early myotonia atrophica.

Case 2

A soldier aged 31, serving in the Middle East, was similarly referred to hospital because of pain about the neck and right shoulder and obvious asymmetry. His past clinical history revealed nothing of note, and there were no other complaints. He said his right shoulder had not "felt right" for some years, but he had never had any acute pain and his right arm was normal.

The right sternomastoid muscle was apparently absent. There was considerable weakness and wasting of the upper fibres of the trapezius muscle on the same side, so that the right shoulder had dropped considerably (Fig. 2). There was no other abnormality in the nervous or the muscular system. X-ray plates, as in Case 1, showed nothing abnormal. Serological studies were negative.

The patient was detained in hospital, but no clue to the nature of the condition was forthcoming. His family history contained nothing relevant.

Case 3

A married housewife aged 30 who had never left this country was referred to me in 1946 because she complained that "there was something wrong with my neck." She was an intelligent and healthy young woman and said that at the age of 24 she had had acute pain on the left side of her neck. It prevented her sleeping and persisted for about one month. She consulted her doctor; he found local tenderness and believed that it was some form of fibrositis. During that month, however, she also had another symptom. This consisted of periodic tic-like movements of the head on the neck. Many times a day, for a few moments at a time, her head would tilt quickly, quite involuntarily, to the left. The whole condition seemed to have subsided spontaneously.

For several years she had forgotten the affair until her husband noticed some asymmetry about her neck when she wore an evening dress. That was in 1943 or 1944. In 1945 she either became self-conscious about it or the asymmetry was more noticeable, for she consulted a doctor. She had also found that certain movements were difficult from some positions. If she lay on her right side in bed and wished to raise her head she experienced particular difficulty.

FIG. 2.—Case 2. Wasting of sternomastoid muscle and upper fibres of trapezius on right side. Considerable dropping of right shoulder.

FIG. 1.—Case 1. Wasting of sternomastoid muscle and upper fibres of trapezius on right side. Tilt of head to right suggests that the lesion is of long standing.

FIG. 3.—Case 3. Wasting of sternomastoid muscle and upper fibres of trapezius on left side.

She sometimes slept without a pillow, and then had some difficulty in raising the head. These symptoms, she stressed, were slight.

On several examinations the only abnormalities to be detected were identical with those found in Cases 1 and 2 (Fig. 3). The sternomastoid muscle and the upper fibres of the trapezius on the left side were noticeably weak and wasted. The nervous and muscular systems were otherwise normal. Faradic responses were not elicited from the left sternomastoid; only weak contractions were obtained from the upper part of the trapezius. Radiographs of the cervical spine and skull were normal. The blood Wassermann reaction was negative.

During the past two years she has been under my observation and no further signs have developed. Her condition is apparently stationary. Two brothers have been examined, and they present no abnormality. Both parents are dead and neither had any disability to suggest myotonia atrophica. She has inquired of cousins and other relatives, with negative results.

Aetiology

In each case all the known causes for such a lesion were considered, but the aetiology remains obscure. In the first two cases the story did not conform to the very characteristic sequence of events in the affection of neuritis of the shoulder girdle. Sternomastoid atrophy has not been recorded in that condition (Spillane, 1943; Parsonage and Aldren Turner, 1948).

The selective nature of the wasting strongly suggests that the lesion is neuritic and not muscular. There was no other abnormality; on careful examination neighbouring muscles were found to be normal.

If the nerve lesion is in the nature of a "neuritis" its causation remains unknown. Cervical adenitis would be unlikely to result in such severe atrophy. Surgeons have often commented on the manner in which the spinal accessory nerve escapes injury in suppurative conditions of the neck. Like the facial nerve the spinal accessory nerve is purely motor in function, but it does not emerge from the skull through a small foramen alone as does the former nerve. It enters the neck accompanied by the jugular vein and by the ninth and tenth cranial nerves through a relatively ample aperture. A lesion at this point would also be likely to involve these nerves.

Poliomyelitis is often suggested, especially in orthopaedic clinics, when the problem arises of explaining an isolated paralysis of one or more muscles. But in this series of cases there is certainly no positive evidence in that direction. From the accounts given by the patients the likelihood of congenital deficiency of the sternomastoid muscle is very remote.

Bilateral atrophy of the sternomastoid muscles is, of course, a usual finding in myotonia atrophica. But the other features of this disease—myotonia, general weakness, diffuse muscular pains, baldness, testicular atrophy, cataract, absent deep reflexes, and reduced vibration sense in the lower limbs—were entirely wanting. Maas (1937), however, has noticed unilateral wasting of one sternomastoid muscle in apparently healthy relatives of patients afflicted with this hereditary malady. One of his published photographs is very similar to that of Case 2. I have been interested in the condition of myotonia atrophica for some years, and inquiry and examination in the present series were conducted with this condition in mind. No evidence of the disorder was present—if one excludes the atrophy of the muscle fibres supplied by the spinal accessory nerve. In Case 3 it was also found that the electrocardiogram was normal. This may be of interest in view of the recent evidence that low-voltage P waves, prolongation of the P-R interval, and slurring of the QRS complex

have been noted in myotonia atrophica (Evans, 1944; Heard and Spillane, in the press).

One may ask whether these cases are examples of the same disease. Although there is no definite proof, it is tempting to assume that they are. The only clue to causation lies in the story of Case 3. She had acute pain in the left side of the neck six years previously and for a time was troubled with spasmodic contractions of the left sternomastoid muscle. The affected area was tender and her doctor diagnosed "fibrositis." This sequence of events was followed by wasting of the muscle. It seems, therefore, that originally there was an irritative lesion of the spinal accessory nerve on that side.

A search of the literature has not disclosed any reports of such a condition, but isolated unilateral hypoglossal-nerve paralysis has been described. The lesion was thought to be due to a periostitis of the anterior condyloid foramen analogous to the lesion of the stylomastoid foramen which may be responsible for Bell's palsy (Goldenberg and Sandler, 1931).

Summary

Three cases are reported in which there was isolated wasting and weakness of one sternomastoid muscle and the upper fibres of the trapezius muscle on the same side.

The cause is not known, but it is probable that the lesion was in the spinal accessory nerve.

The lesion may be similar to that in Bell's palsy.

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A CASE OF DIAPHRAGMATIC HERNIA OF CONGENITAL ORIGIN WITH PERFORATION OF STOMACH

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The following account of a complete right-sided thoracic stomach with a perforated pyloric ulcer is of interest because of (1) the extreme rarity of such a condition—we have found no other recorded case of a similar character; (2) the absence of any previous serious disability—in fact, he was able to earn his living as a general labourer, which involved considerable physical exertion and produced no pulmonary or acute digestive disturbance; (3) the opportunity of seeing and feeling the position of the various organs at operation; and (4) the complete absence in convalescence of any symptoms of discomfort, either above or below the diaphragm.

Case History

At about midnight on Oct. 25, 1948, a man aged 29 was suddenly taken ill with acute abdominal pain, which started at the centre of the abdomen around the umbilicus. It persisted and was extremely severe. He felt sick and vomited once before his admission to Newark Town and District Hospital.

Four years previously he had had "gastric trouble"—pain and fullness in the epigastrium about half an hour to one hour after food, followed by vomiting, which brought relief