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.

REFERENCES

- Evans, W. (1944). *Brit. Heart J.*, **6**, 41.
Goldenberg, N. A., and Sandler, J. G. (1931). *Rev. oto-neuro-oftal.*, **9**, 429.
Maas, O. (1937). *Brain*, **60**, 498.
Parsonage, M. J., and Turner, J. W. Aldren (1948). *Lancet*, **1**, 973.
Spillane, J. D. (1943). *Lancet*, **2**, 532.

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

immediately. His own doctor treated him by diet and alkalis, and the condition subsided after three weeks. Since then he occasionally had a mild recurrence of his epigastric pain for which he did not attend his doctor; he had not adhered to the diet prescribed. He had had no pain before the onset of his present illness. For the last five years he has been having mild attacks of "asthma," which were worse on the hot days of summer. The attacks did not prevent him from doing his work, and he has not received treatment for them.

On examination the temperature was 97.4° F. (36.3° C.), pulse 88, and respirations 26. He was well nourished. He seemed to be in pain but not shocked, and was rather excited. His tongue was moist and slightly coated. General abdominal rigidity was present, but no real tenderness; there was no reduction in liver dullness or any rebound phenomenon. No tenderness was felt per rectum. Respirations were mainly thoracic and wheezy in character. There was slight dullness at the right base posteriorly on percussion, and breath sounds were wheezy all over the chest. The pulse was regular and of good volume. The heart was not enlarged and there were no murmurs. The blood pressure was 130/84. No appreciable disease was found in the central nervous system.

Perforated gastric ulcer was suspected, and in view of the normal temperature and pulse and the patient's general condition it was decided to treat him conservatively under observation. A kaolin poultice was applied to his abdomen and he was put in Fowler's position. A half-hourly pulse chart was kept. At 5 a.m. the temperature was 99.8° F. (37.7° C.), pulse 90, and respirations 24. At 7.30 a.m. the temperature was 100.6° F. (38.1° C.), pulse 112, and respirations 24. The pulse chart showed a rise from 90 to 112 between 5 and 7.30 a.m.

He was re-examined at 7.30 a.m. and his abdomen was found to be still rigid, with generalized tenderness and rebound phenomenon, and in view of his rising pulse and temperature laparotomy was decided upon.

Operation.—At 9 a.m., under anaesthesia induced by thio-pentone followed by gas and oxygen and ether, a supra-umbilical paramedian incision was made and free fluid was found on opening the peritoneal cavity. On exploration the stomach was found to be entirely in the right thoracic cavity. It was in a complete peritoneal sac. There was a small hole under the liver in the right side of the crus of the diaphragm. This opening was a definite rounded orifice admitting three or four fingers. Through it, it was possible to trace the duodenum and pylorus. With peritoneal forceps passed through the opening the stomach could be grasped and pulled slightly downwards, but it could not be drawn through the aperture. A small perforation was identified by tracks of lymph and effusion, and it was closed by passing a needle on a holder through the aperture and making a purse-string suture, covering it with an omental graft from the greater curvature omentum. The liver was very prominent in the wound and seemed to be displaced downwards. It had to be retracted upwards to expose the gap in the diaphragm. When this was done there was a clear view of the opening, which was situated in the right crus of the diaphragm behind the liver and to the right of the vertebral column. A drainage tube was inserted through a separate suprapubic stab wound and the abdomen was then closed in layers.

The patient stood the operation quite well, and was in fair condition when taken from the theatre. Immediately on his return to the ward 5% glucose-saline intravenously was started. The 5% glucose saline was given intravenously for the first 48 hours. A Ryle's tube was passed and the stomach aspirated every two hours. Then he was given glucose water and citrated milk by mouth. This was gradually increased until at the end of the first week he was put on the first week of Sippy's diet. He tolerated his feeds well, and Meulengracht's diet was given at the middle of his second week. His temperature and pulse were raised for the first week; this was ascribed to pulmonary complications after anaesthesia. It subsided after treatment with penicillin injections and inhalations. His temperature and pulse have been normal since that time.

On Nov. 16 a screening examination and a series of skiagrams showed a complete thoracic stomach on the right side

of the thorax and confirmed that the stomach is above the diaphragm and is lying centrally between the anterior and posterior walls of the chest. It is in a complete peritoneal sac, separated from the pleural cavity by omental tissue, peritoneum, and the basal pleura. Screen examination showed full movement of the left diaphragm and slight movement of the right diaphragm. The stomach, the greater curvature of which is uppermost, appears to empty from the right side. The duodenum passes across and behind the base of the stomach to empty through the aperture in the medial side of the right diaphragm. A film taken on Nov. 17 showed the colon displaced to the left side. This is probably due to the downward displacement of the liver.

By the sixth week he was convalescent and was able to take light diet without any discomfort or digestive trouble. He looks and feels extremely well and has returned home.

Comment

This is a case of complete thoracic stomach with a short oesophagus of congenital origin, hence the failure of the attempt to draw the stomach through the opening in the diaphragm at operation and the absence of a sliding stomach at the radiological examination under the screen when in the vertical position, which may occur in some cases.

It is completely enclosed in a peritoneal sac separating it from the pleural cavity. Unfortunately at operation it was not possible to feel the junction of the oesophagus with the stomach, so we cannot say whether the peritoneum was

Radiograph showing stomach on right side of thorax above the diaphragm. The greater curvature is uppermost. Barium is in the oesophagus.

reflected on to the stomach before it reached the cardiac orifice or whether it enclosed the cardiac end of the oesophagus.

As a result of the complete peritoneal sac there was no infection of the pleural cavity from the peritonitis after operation.

The stomach is completely rotated, with the greater curvature uppermost, and the oesophagus curves underneath the cardiac end of the greater curvature to reach the cardiac orifice. There is no stricture or hour-glass contraction, as it is a completely thoracic organ and no portion is nipped by the diaphragm, hence the absence of gastric fullness and distension.

We wish to express our thanks to Sir Thomas Dunhill for his interest in the case, for his great kindness in corresponding with us about it, and also for his account of the condition in the Arris and Gale Lecture delivered by him to the Royal College of Surgeons in 1934, from which we have derived much information. We are also indebted to Dr. F. M. Gordon and Mr. R. J. Walls, M.S.R., for the radiography.