ANNALS OF SURGERY

VOL. LV APRIL, 1912 No. 4

ORIGINAL MEMOIRS.

CEPHALIC TETANUS.*

WITH REPORT OF A CASE.

BY ALFRED J. BROWN, M.D.,

OF NEW YORK,

Demonstrator of Anatomy, College of Physicians and Surgeons, Columbia University; Clinical Assistant, Department of Surgery, Vanderbilt Clinic.

OUR knowledge of the unusual form of tetanus variously known as cephalic tetanus, kopftetanus, and tetanus hydrophobicus is still so imperfect as to render desirable reports of individual cases, in the endeavor to ascertain the causation of so peculiar a clinical type of an otherwise well-known general disease.

The recognition and first description of the disease is usually ascribed to Rose,[1] by whom the name tetanus hydrophobicus was given; his priority has, however, been disputed by Eastman,[2] who gives the credit for the first description of the condition to Bell.[3] At present the term cephalic tetanus is applied to those cases which present a paralysis or paresis of one or more of the cranial nerves as a prominent symptom, together with more or less well-marked symptoms of tetanus, generally confined to the region of the head and neck, though at times involving the entire body. Some authors have classified the condition as chronic tetanus on account of its usually

* Read before the Brooklyn Society for Neurology, December, 1911.

long incubation period, but this is by no means invariably characteristic of this form. Others have classed as cephalic tetanus all cases in which the wound of entry has been on the head, irrespective of the symptoms presented. The cases mentioned here belong to the class above referred to, in which paresis or paralysis of one or more of the cranial nerves is present.

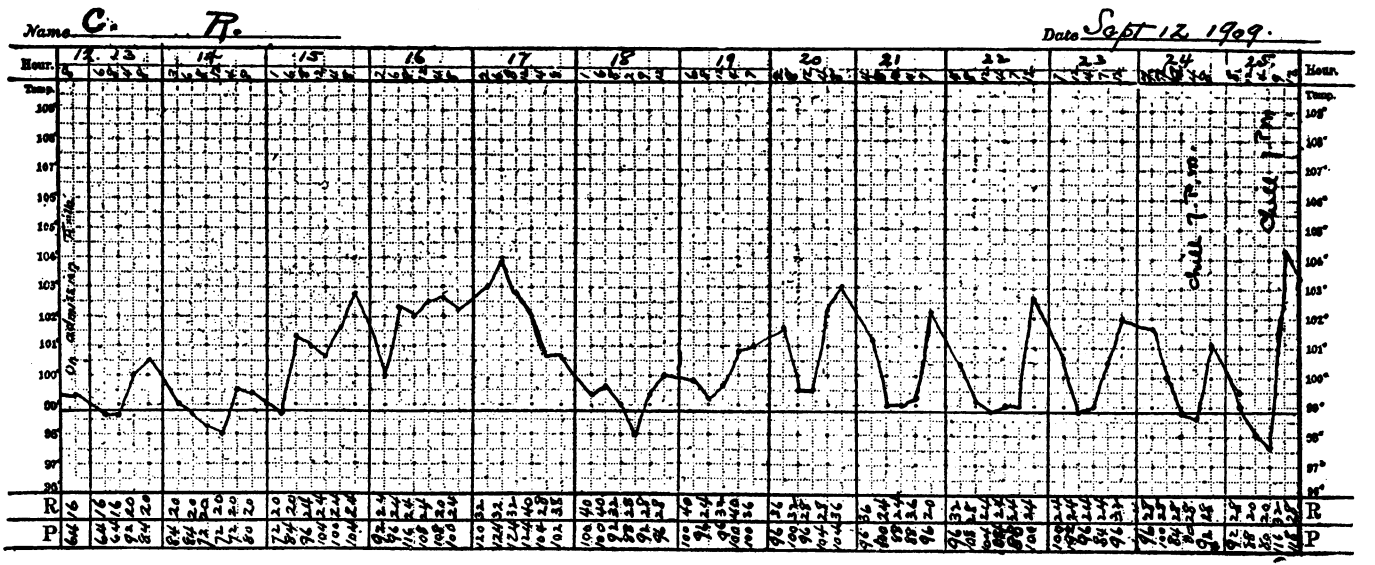
The disease is apparently very rare in the United States. Eastman [2] mentioned three cases as all he found reported in the literature up to 1907, and added three more. I have been unable to find any additional cases reported in this country to date.

The following case, the seventh reported in the United States, was seen by me in consultation with Dr. H. D. White, of Rome, New York, and through his courtesy I am permitted to report it.

CASE I.—C. R., male, farmer, aged forty years. His family and previous histories throw no light on his present trouble.

On Monday, September 6, 1909, the patient was struck over the right temporofrontal region by the tine of a mechanical manure spreader, which caused a lacerated wound of the forehead. The blow was severe enough to knock him down, and he thinks he was unconscious for a few minutes, was dazed for an hour or two, and suffered from a severe headache for the remainder of the day. The wound was cleansed and dressed by Dr. White; it was found to extend down to and at one point through the periosteum, but there was no evidence of fracture of the skull. The following day the patient felt practically well, and went about his work as usual on that day and on the day following. On Thursday, three days after the accident, the patient noticed that he could not perfectly control the right side of his mouth while eating and particularly when trying to take fluids. This was followed on the next day by twitching of the muscles of the left side of his face, which increased in severity, and soon tonic spasms of the muscles of mastication of both sides supervened, giving rise to a marked trismus. There were no constitutional symptoms and the patient did not feel sick in any way. The muscular spasms became so severe that six days after the acci-

CHART 1.



Temperature chart.

dent, upon his admission to the Rome Hospital, he was unable to take any nourishment by mouth and his jaws were so tightly clenched that the teeth of his lower jaw were cutting into the gum of the upper, and it was necessary to extract six of them. Subsequent to this he was able to take liquid nourishment by mouth for a day, but on the following day it was necessary to resort to rectal feeding. There was some slight twitching of the extremities.

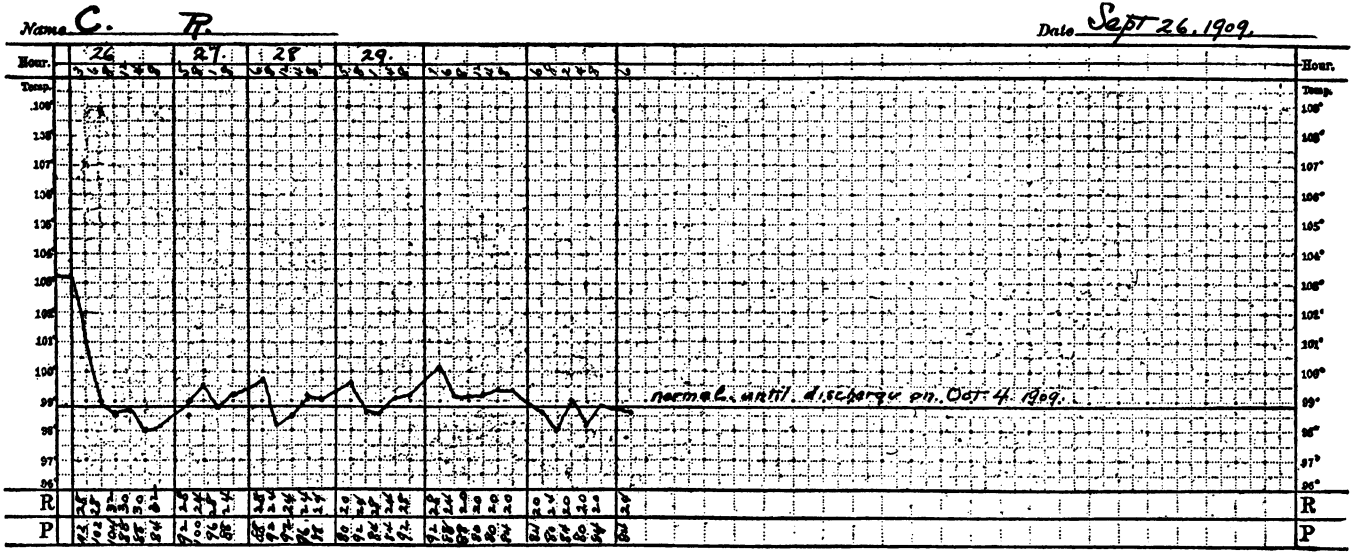
Physical examination at this time showed complete right-sided prosopoplegia determined by marked drooping of the corner of the mouth, obliteration of the nasolabial fold, and inability to close the eye or make a grimace on the right side of his face. There was marked spasm of the muscles of mastication of both sides, the jaws in consequence were tightly clenched, and it was impossible for the patient to open his mouth. When the patient wished to talk he placed his fingers between his lips to hold them apart. Any attempt to test the reactions of the right facial muscles resulted in such an increase of the painful spasm that this was given up after a very brief and unsatisfactory trial. There were occasional slight twitchings noted in the muscles of the extremities. The remainder of the physical examination revealed normal organs. The temperature (axillary) ranged from 98.6° in the morning to 100.5° in the afternoon, with a corresponding range in pulse and respiration from 64 to 92 and 16 to 20, respectively. The patient was given 1500 units of tetanus antitoxin subcutaneously.

For the next five days the above conditions persisted, and in addition exceedingly painful and distressing spasms of the pharyngeal muscles appeared, which were aggravated by any attempt at swallowing. For two days there were also severe respiratory spasms, and at times a restless delirium with a temperature reaching 103°, pulse of 100 to 124, and respirations 20 to 32.

On the tenth day of the disease a slight conjunctivitis and keratitis developed in the right eye, which promptly subsided under appropriate treatment.

On the twelfth day of the disease the spasms of the muscles of mastication began to decrease; the patient was able to take nourishment by mouth and appeared to be improving. The temperature range was a little lower.

CHART II.



Temperature chart.

On the fifteenth day the patient developed severe paroxysms of coughing, and at the same time complained of a pain in his right shoulder and the right side of his chest which was increased by coughing or taking a deep breath. The attacks of coughing lasted for from 15 to 20 minutes, being induced especially by any endeavor to take food, and in consequence rectal feedings were again instituted. These severe paroxysms continued at intervals until the twentieth day, when they began to decrease in severity and soon disappeared. On the seventeenth day he had a chill lasting a few moments and followed by a rise in temperature to 101.1°, and this was repeated on the eighteenth day, this time being more severe, and followed by a rise of temperature to 104.2°. On the following day the temperature fell rapidly to normal and remained there. Physical examination during the above period elicited the signs of dry pleurisy on the right side, consequently it seems reasonable to assume that the symptoms were entirely distinct from the symptoms of tetanus which had preceded them.

At the time of the fall of temperature to normal, the trismus had so far relaxed that it was deemed safe to take the patient's temperature by mouth. From this time on the convalescence was rapid and the patient was discharged on the twenty-eighth day after his injury, with a very slight prosopoplegia remaining and with the wound healed by granulation after a mild suppurative course.

From the sixth until the fourteenth day of his disease inclusive, the patient received 65,500 units of tetanus antitoxin, injected subcutaneously in ten doses ranging from 1500 to 11,500 units at a dose.

A stab culture taken from the wound was examined bacteriologically by Dr. W. S. Nelson, of Utica, New York, and a preliminary report of bacilli closely resembling tetanus bacilli but without spores was made. Owing to the sudden illness and death of Dr. Nelson, further investigations which were under way were never completed.

REVIEW OF CASES REPORTED IN MEDICAL LITERATURE.

Willard[4] in 1895 was able to collect 74 published cases in the literature, his own case making 75. Ross[5] in 1906 presented an analysis of 81 cases in the literature up to that time,

and since that date I have found 12 cases, the one here reported making 13, thus bringing the total to date to 94 cases.

In 84 per cent. of cases the first sign of the disease has been trismus, either alone or accompanied by paralysis of one or more of the cranial nerves. The paralysis, however, usually follows the trismus. In 14 cases, including the one here reported, prosopoplegia has preceded the appearance of the trismus. Ross[5] asserts that the appearance of this symptom before the trismus determines an almost invariably fatal termination, but this observation does not appear to hold good in the series to date, as of the 14 cases but 9 have died, thus making a mortality of 64.2 per cent., as contrasted with a mortality of 53.2 per cent. for cephalic tetanus as a whole. The 13 previous cases in which paralysis has preceded the trismus have been reported in detail by Stolz.[7]

Neumann[6] claims that the facial nerve is involved most frequently, then the oculomotor, abducens, trochlear, and hypoglossal in the order named, but at the same time states that the cases are too few in number to make any dogmatic rule. This observation has proven true in the series to date.

Prosopoplegia is by far the most common form of paralysis and appears on the side of the lesion. Cases of crossed paralysis have been reported but are open to doubt as to the site of infection.

Bilateral prosopoplegia has been reported in seven cases. In all but one case, that of Roberts,[21] the wound has been across the median line of the head, usually across the bridge of the nose. Not all median line wounds, however, have given rise to bilateral prosopoplegia (Nankivell,[22] Wagner,[24] Oliva,[23] and Nerlich[16]).

Paralysis of one or more of the muscles of the eye has been described, and in these cases the wound has involved the orbit. The paralysis has varied from complete ophthalmoplegia to paralysis of one nerve only. Ptosis has been noted in eight cases, strabismus in four, nystagmus in one. Fixed dilatation of the pupil has been described.

Paralysis of half the tongue denoting involvement of the

hypoglossal nerve has been noted in two cases as the trismus was disappearing.

The site of the wound has been on the head or face in all but two cases. Duvergey [8] reports a case in which the site of infection was supposedly under the finger-nail, thus leaving room for doubt as to the site of the lesion, and Zack [9] reports a case of cephalic tetanus following a wound of the foot.

The prognosis depends upon two factors: first, the length of the incubation period; as a general rule it may be said that the shorter the incubation period, the more dangerous the disease; and second, upon the localization of the tetanic spasms. In those cases in which the spasms remain localized in the muscles of mastication and deglutition, the prognosis is good, regardless of the length of the incubation period. If respiratory spasms supervene, the prognosis is more grave, as death may and often does occur during a spasm. If the spasms become generalized, the prognosis is almost invariably fatal.

With regard to the etiology of the condition, a sufficient number of cases have been reported in which tetanus bacilli have been obtained by culture from the wound to make it practically certain that the disease is due to this infection.

Regarding the pathology of the paralyses, however, many theories have been advanced. Rose [10] claims that the paralysis is caused by a local inflammation of the seventh nerve, causing a swelling of its trunk and a consequent constriction at its exit from the stylomastoid foramen. This view is not supported by autopsy findings, as such conditions have never been observed and in all cases reported the nerve-trunk has been found normal on microscopical examination.

Serems [11] has suggested that the paralysis is reflex from involvement of the fifth nerve.

Brünner [12] believes that the paralysis is caused by a selective toxin which is paralytic in its action and affects the end organs of the nerve-fibre. He notes the fact that Fraenkel and Kitasato have been able to produce the facial and ocular palsies in rabbits by the injection of the crude toxin into the facial muscles, but at the same time there has been a reaction

of degeneration present in the affected muscles, while in cephalic tetanus the prosopoplegia is of the character of Bell's palsy and presents no reaction of degeneration. Klemm [18] also advocates the above theory.

Binet and Trénel [14] state that contraction and paralysis of muscle due to nerve impulse are two phenomena of the same order corresponding to two different states of the nerve-cell. The first phase, causing contraction, is excitation; the second, causing paralysis, is complete annihilation. Consequently the effect on the nerve-cell is governed by the amount of toxin transmitted along the nerve-fibre to the nucleus. They argue that the seventh nerve is usually first involved, or if the lesion be in the orbit, the third, and there is sufficient strength in the toxin to cause annihilation of the cells of the nucleus and consequent paralysis. By the time the toxin reaches the motor nucleus of the fifth nerve, however, it is so attenuated that excitation of the cells is caused and spasm results.

Gosselin [15] asserts that there is no true paralysis of the cranial nerves, but the muscles of the opposite side are in excessive contraction, and consequently there appears to be a paralysis of one side. This view is sufficiently refuted by the occurrence of bilateral prosopoplegia.

Nerlich [16] was the first to suggest that the toxin acts on the motor cells in the central nuclei, basing this assumption on the fact that he noted bubble-shaped lesions in the motor cells situated in the nucleus of the twelfth nerve on the side corresponding to the lesion and in both seventh nerve nuclei. The wound in his case was across the bridge of the nose, but the patient exhibited left-sided prosopoplegia only.

Berkhardt [17] reports lesions in the brain stem at the level of the nucleus of the seventh nerve.

Bourgeois [18] states that he has found pigmentation in the nucleus of the twelfth nerve.

The method by which the toxin reaches the central nervous system is by passage along the nerve-fibre from the point of injury as shown by Marie and Morax.[19] The toxin presumably travels by lymphatic paths.

Von Orzechowski [20] reports a fatal case in which he found the nuclei of the third, fourth, fifth, and sixth nerves normal. In the nucleus of the seventh nerve corresponding to the side of the lesion there was a group of cells in the ventromedian portion which showed chromatolysis. There was also vacuolization of cells which corresponded to the description of Nerlich. He quotes Goldscheider and Flatau, Goebel, Matthes, Von Halbau, Sjovall, and Kron as having also found the same lesions. He also notes the same vacuolization of cells occurring lower down in the cord, and the history of his case shows that the patient suffered a wound not only on his face but also on the chest, and suffered from generalized spasm as well as from prosopoplegia. This would tend to show that the lesion in the central nucleus is localized in an area corresponding to the site of the lesion of the nerve, whatever that may be.

From the weight of evidence at the present time, it seems fair to assume that the paralysis is due to the action of the toxin upon the nucleus of the nerve which supplies the muscles in the neighborhood of the injury. The toxin is carried to the nerve through the lymphatics, and thence transmitted along the nerve-trunk by the lymphatics to the nucleus. If this point be granted, may it not be true that this condition may be present in any portion of the body? Prosopoplegia would be noted particularly by contrast to the spasm of the opposite side of the face, whereas, on the other hand, paralysis of individual muscles or muscle groups in other parts of the body would easily escape detection by being overshadowed by the general spasm.

In the cases of cephalic tetanus, many which have resulted fatally have shown generalized tetanus and have also shown wounds outside of the facial area, as did Von Orzechowski's. When this has not been the case the fatal issue has often occurred in a respiratory spasm. The early symptom of general tetanus is trismus as the common name of the disease, lockjaw, implies. This in general tetanus is followed by pharyngeal and respiratory spasm, stiffness of the neck, and

gradual passage of the area of spasm down the body, thus denoting a passage of the toxin downward along nerve-trunks. At the same time it is now fairly well established that the toxin gains access to the central nervous system through lymphatic paths.

Turning now for a moment to the consideration of another grave condition which travels by lymphatic paths, carcinoma, we find that there has long been recognized a portion of the body, sparsely supplied with lymphatics, in which carcinoma is quite benign. This area is bounded below and behind by a line drawn from the lower border of the ala of the nose backward to the inferior border of the lobule of the ear, and thence vertically upward through the external auditory meatus to the median line, where it meets a corresponding line of the opposite side. Carcinoma occurring in this area rarely gives rise to metastases or cachexia, and aside from the local lesion is comparatively benign. It seems fair to assume then, reasoning by analogy, that tetanus occurring in this same area, without a wound of any other portion of the body, will, because of the limited absorption of the toxin by the lymphatics, be of a benign form and present only the early symptoms of the disease, namely, trismus, pharyngeal and respiratory spasm, while exactly the same infection, occurring in other parts of the body, owing to the plentiful lymphatic drainage, will be absorbed more rapidly and in greater amount, and will, in consequence, give rise to generalized spasm.

LITERATURE.

[1] Rose: Handbuch der Chir., von Pitha and Billroth, vol. i, ab. 11, No. 11, p. 5.
[2] Eastman, F. C.: Neurographs, Brooklyn, 1907, i, p. 8.
[3] Bell, Charles: Lectures before Royal Society of London, 1830.
[4] Willard: Trans. Coll. of Phys. of Phila., 1895, p. 27.
[5] Ross, T. W. E.: Edinburgh Med. Jour., 1906, n. s. xix, p. 229.
[6] Neumann: Zeitschr. für Heilkunde, 1902, p. 344.
[7] Stolz, A.: Strassburg Med. Zeitung, 1905, ii, p. 259.
[8] Duvergey, J.: Gaz. Hebd. de sc. méd. de Bordeaux, 1903, xxiv, p. 484.
[9] Zack, E.: Centralblatt für inner medecin, 1904, 5 Novembre.
[10] Rose: Deutsche Chirurg., 1897.
[11] Serems: Union Medicale, No. 173.

[13] Brünner: Brun's Beiträge zur klin. Chir., 135, ix, x, xii.
[14] Klemm: Berl. klin. Wchnschr., Jan. 16, 1893.
[15] Binet and Trénel: Rev. de Chir., Paris, 1909, xl, p. 427.
[16] Gosselin: Gaz. de Hop., 1880, ix, p. 65.
[16] Nerlich: Inaug. Diss., Halle, 1892.
[17] Berkhardt: Neurol. Centralblatt, 1898, No. 4, p. 146.
[18] Bourgeois: Gaz. de Hop., Paris, 1900.
[19] Marie and Morax: Annals de l'institute Pasteur, 1903.
[20] Von Orzechowski, K.: Deutsche Zeitschr. für Nervenh., Leipzig, 1909–1910, xxxiv, p. 1.
[21] Roberts: Lancet, London, 1891, ii, 61.
[22] Nankivell: Lancet, London, July 4, 1883.
[23] Oliva: Gaz. de Chir., 1886.
[24] Wagner: Schmidt's Jahrbuch, 1884, p. 139.