PONTO-BULBAR CRISES ASSOCIATED WITH SIALORRHŒA IN SYPHILIS OF THE NERVOUS SYSTEM.*

BY ERNST P. BOAS AND WALTER M. KRAUS, NEW YORK.

I.—INTRODUCTION.

THE objects of presenting this case are to emphasize the fact that crises may occur in cases of cerebrospinal syphilis other than tabes dorsalis, and to describe a group of symptoms referable to the 7th, 9th, and 10th cranial nerves.

It is customary to associate crises with tabes. However, there seems no reason to believe that involvement of nerves or nerve centres sufficient to produce crises cannot occur without concomitant signs of tabes. Nonne¹ emphasizes this in discussing gastric crises. He says, "It is well known and has recently been re-emphasized by Dunger, that gastric crises may be the initial symptoms of tabes. There is no question as to the diagnosis of tabes when pupillary disturbances, absence of Achilles or of knee-jerks or of both, occur in such cases. This is not so in those cases in which 'characteristic gastric crises' occur at short or long intervals in those who have been syphilitic without any objective organic sign as far as the nervous system is concerned. I know of three such cases, and have been able to follow the first case for four years, the second and third cases for three years. All three patients had been syphilitic. Examination of the spinal fluid in all three showed a mild grade of lymphocytosis. I had not begun testing for globulin when I was first consulted by these patients. As you will hear presently, a mild grade of lymphocytosis in the luetic is not sufficient for the diagnosis of tabes."

The case here reported presents a rare type of mixed sensory, secretory, and motor crisis which is scarcely mentioned in the current text-books, and of which the most conspicuous sign—profuse paroxysmal salivation—is only sparingly discussed in the literature of crises.

* From the Medical and Neurological Divisions of Montefiore Hospital, New York. Read before the Section of Neurology and Psychiatry of the New York Academy of Medicine, April 11, 1922.

II.—LITERATURE.

The first note of salivation in tabes which we have been able to discover is in Pierret's² thesis of 1876. Six years later Putnam³ reported six cases of tabes with salivation. The salivation was usually associated with some other form of crisis. One of the patients had occasional attacks during which the saliva "flowed like a fountain" for about fifteen minutes. In the case of another patient each attack of salivation lasted about seven hours. Then Pitres⁴ described a tabetic who had had gastric crises which disappeared after a few years. Some time afterward an intense salivation developed which resisted all treatment. It appeared suddenly in the morning soon after the patient arose, and lasted two to three hours; it was accompanied by a sense of heat in the upper part of the face and in the tongue. For a period of two weeks the salivation was continuous day and night. It finally ceased, but recurred two years later. There was no disturbance of taste, but the sense of smell was completely lost. In 1887 Féré⁵ described a tabetic with ocular and laryngeal crises who also exhibited paroxysmal attacks of salivation. A few years later Féré⁶ observed a taboparetic with epileptic attacks involving the right side of the face and the right arm, in whom each attack was preceded by abundant salivation. Mazataud⁷ was able to report four cases of salivation in tabes. His first patient exhibited a simple sialorrhœa. In the second patient the salivary crisis was announced by a feeling of nausea. The patient would then expectorate three or four large mouthfuls of saliva; with this he complained of a bitter taste in his mouth as well as of hallucinations of sight and hearing. Taste and smell were otherwise normal. The crisis lasted from one to three minutes only, but recurred once to five times a day. A third patient during attacks of gastric crises would experience a bitter taste in his mouth which was followed by sudden salivation lasting from one to two minutes, at the end of which time not only the salivation, but the gastric crisis as well, would cease. These attacks occurred ten times a day. The fourth patient had continuous salivation day and night; with this there was neuralgia of the left side of the face. The salivation continued for over a year, paroxysms occurring during which there was a sudden increase of secretion of saliva.

Klippel^{8,9} reports a case with autopsy findings. The patient complained of a continuous bad taste in his mouth which he likened to the taste of decayed fish. He exhibited changes in sensibility of the 5th nerve, on the face and lips, anæsthesia, formication, and flushing of the cheeks. On autopsy the salivary glands were large and much congested. In the parotid gland there were areas of round-cell infiltration, and the cells of the glandular acini were increased in

number and smaller than normal, with large nuclei. There was a catarrhal inflammation of the larger excretory ducts. In the submaxillary gland some of the acini appeared active and some inactive. He also found degenerative changes in the ganglia of Andersch and Gasser and in the 5th and 9th nerves. Klippel comments on the association of disorders of taste and smell, sensory disorders in the area of the trigeminal, tachycardia, and salivation.

André's¹⁰ patient had salivary crises lasting from two to three hours in which the flow from the left side was greater than from the right. The patient also had attacks of left-sided neuralgic pains in the face, as well as hypæsthesia of the left side of the face and tongue. Taste was normal.

Umber¹¹ described a patient who experienced intensely disgusting tastes and smells, associated with a sensation of swelling in the neck and throat and a feeling of swelling of the submaxillary glands, with profuse salivation. These symptoms always came at the end of the gastric crisis. In another of his cases the gastric crises were preceded by great salivation and a very bad taste in the mouth. In both of these cases the salivation was apparently secondary to the disturbance of taste.

Alexander¹² in 1911 described a tabetic who had attacks of burning in the mouth with salivation, as well as a bad taste. They never occurred at meals. Taste and smell were otherwise normal. The saliva was thin and watery.

III.—CASE REPORT.

The following is a report of a case of syphilis of the nervous system with salivary crises, which we have studied at Montefiore Hospital.

The patient is a married man, 35 years of age, a furrier by occupation. He entered the hospital complaining of pain in the front of the upper left chest, of loss of weight, and of weakness. He has had no illnesses, with the exception of gonorrhœa at the age of 14 and a chancre at the age of 18. He received antiluetic treatment for four weeks. He states that at this time he lost his hair in 'bunches'. Twenty months ago, while rising from bed one morning, he was suddenly seized with pain behind the suprasternal notch, which radiated to the upper part of the left chest. He describes the pain as in the chest and as a choking, pressing sensation. He then vomited three or four times. Several days later he became dizzy and fell to the floor. For the first four months these attacks of pain occurred about every three weeks. Gradually the intervals became shorter, and during the last nine months the attacks of pain and vomiting have occurred almost daily. Their duration is variable, but they usually last a whole morning, and sometimes extend over several days. The patient was first treated at a gastrointestinal clinic until a blood examination revealed a + + + + Wassermann. This test has been taken at several hospitals. At Bellevue it was + + + +,

at Mt. Sinai + + + +, at Harlem +, and at Montefiore it has twice been negative. After the first Wassermann was reported + + + + he received fourteen doses of salvarsan and twenty mercury injections, and the Wassermann became negative. However, his symptoms continued unabated. Three weeks after the treatment was instituted the patient began to complain of attacks of *severe salivation* lasting many hours. These have persisted, and at present occur almost daily. The salivation commences in the morning and is followed by retching, vomiting, and pain in the upper chest. Food is rarely seen in the vomitus. The salivation and pain last from four to twelve hours and occasionally persist over several days. With each attack the patient feels dizzy, and must lie down or hold on to something for fear of falling. The dizzy spells, however, last only a few seconds. The patient has lost twenty pounds in weight in the last year and a half. There was very slight difficulty of urination some months ago, but this has entirely disappeared. He never suffered from incontinence.

PHYSICAL EXAMINATION.—A man of small stature, weighing ninety-six pounds. Pale, rather poorly developed, and of normal intelligence. Teeth, tonsils, and pharynx in good condition. No evidence of gingivitis such as is seen in mercurial poisoning. Tongue slightly coated. Heart and lungs normal on physical examination. No increase in the area of aortic dullness, although the *x-ray* shows a slight widening of the aortic shadow. Electrocardiogram normal. Pulse-rate continuously rapid, ranging about 100 even when the patient is at rest. Efforts to take the pulse-rate while the patient is asleep have been unsuccessful, because he is such a very light sleeper. Blood-pressure is 135/75. Abdominal viscera normal. Gastro-intestinal *x-ray* negative. Gastric analysis shows the following:—

Fasting contents	..	..	145 c.c.
Free HCl ..	..	..	84
Total acidity	..	..	91
Ewald test meal	..	..	150 c.c. withdrawn
Free HCl ..	..	..	45
Total acidity	..	..	55

This indicates hypersecretion.

The spinal-fluid Wassermann was + while the patient was at Harlem Hospital. At Montefiore it was ±. The colloidal gold at Harlem Hospital was 0133100000, while at Montefiore it was 0111000000. The cell-count after a recent lumbar puncture showed 7 cells, globulin positive.

NEUROLOGICAL EXAMINATION.—Pupils are irregular; left larger than right. No reaction to light, but good reaction with accommodation. Extra-ocular movements normal. No changes in functions of the 5th and 7th nerves. Pharyngeal reflex markedly diminished. Laryngeal reflex absent. No objective disorder of taste or smell. No abnormalities of the vocal cords. No atrophy or fibrillary twitchings of the tongue. *Deep reflexes all very active.* No Babinski or clonus. Abdominal reflexes present on both sides, and active. Muscle, joint, and vibration senses are normal. Pain on deep pressure over the calves, and Achilles tendon elicits a normal response. There is an approximately circular area of slight hypalgesia over the upper portions of the front of the chest. This is not always present. There are no sensory changes on the back. No ataxia and no Romberg. Gait is normal.

PROGRESS.—During his stay in the hospital the patient has had attacks almost daily. His chief complaint is salivation, which, he says, induces the vomiting and which has been followed by the pain in the chest. At night his pillow has often been soaked with saliva, which drools from his mouth while he is asleep. During the attacks, as well as during the intervals, there are no sensory disorders of the face, mouth, tongue, neck, or pectoral regions. With the salivation the patient experienced a bad taste in his mouth, but there has been no objective disorder of taste or smell.

The following is a description of a typical attack.

Jan. 14, 1922.—This morning, on rising, the patient complained of burning in his throat, passing down to his chest. After dinner he began to complain of salivation and severe pain behind the suprasternal notch, passing downward and to the left. He then became nauseated, and vomited once. On examination $1\frac{1}{2}$ hours after the onset, the patient was still in severe pain. He did not remain in one position, but moved about on the bed and rubbed his hand over his upper chest. At frequent intervals he expectorated a thin saliva. Walking did not aggravate the pain. No characteristic posture. Sublingual glands large. Their openings were prominent, red, and wide open, and saliva could be seen to flow from them. Pulse 126. Blood-pressure on several measurements 200/140 to 220/140, sitting. Heart and lungs showed no abnormality. No abdominal tenderness. Over the sternum and for several centimetres on either side there was a dull-red blotchy appearance, due to the injection of minute telangiectatic vessels. This disappeared when the clothes were removed. Atropine gr. $\frac{1}{60}$ given by hypodermic injection. Half an hour later the mouth became dry and salivation stopped. Pain worse.

Feb. 15, 1922.—The attacks have become more frequent and in the last month have occurred almost daily. Salivation took place almost every night. When asleep, saliva often drooled from his mouth, wetting the pillow and awakening him. On rising in the morning he has usually vomited and the salivation has become worse. If he had not salivated the night before, he did not vomit in the morning. After vomiting, the pain commenced behind the suprasternal notch and radiated to the left. The pain was usually not acute, but very uncomfortable, and was oppressive in character. It has been difficult to draw a deep breath. Nothing but morphine relieved this. When pain has been present, urination became difficult. The attack usually passed away at noon, and in the afternoon he felt well. Sometimes the attack persisted for several days. During the attack he cannot eat or drink.

During one of the intervals the patient was given $\frac{1}{3}$ gr. of pilocarpine hypodermically. This induced a very profuse salivation lasting about half an hour, as well as marked perspiration of the whole body, and abdominal cramps. No pain in the chest, nausea, or vomiting appeared. The saliva could be seen running from the parotid and sublingual ducts. The blood-pressure fell from 130/90 to 100/65. The patient has received no salvarsan or mercury for about six weeks, and shows absolutely no evidences of mercurialization.

IV.—DISCUSSION.

This patient does not present any of the signs of tabes dorsalis, but rather the picture of syphilitic cerebrospinal meningomyelitis.

In addition there are the following signs and symptoms:—(1) A profuse secretion of thin and watery saliva; (2) Gastric hypersecretion; (3) Attacks of vomiting related to salivation; (4) Tachycardia; (5) Attacks of high blood-pressure; (6) A sensation of bad taste in his mouth; (7) Pain in the suprasternal notch radiating downward and to the left; (8) Diminution of pharyngeal and absence of laryngeal reflexes.

Site of the Lesion.—All of these signs and symptoms are referable to involvement of the 7th, 9th, and 10th cranial nerves. If we assume that the involvement lies within the pons and medulla it is

FIG. 1.—Diagram of the visceral afferent and efferent connections in the medulla oblongata. The afferent roots and centres are indicated on the right side; the efferent on the left. Visceral sensory fibres enter by the VII nerve (pars intermedia of Wrisberg, *VII pars. int.*) and by the IX and X nerves. These root-fibres include both general visceral sensory and gustatory fibres, all of which enter the fasciculus solitarius. (Fibres of the IX and X nerves also enter the spinal V tract; but since these are somatic sensory fibres from the auricular branch they are not included in the diagram).

On the left side of the figure the general visceral efferent nuclei are indicated by small dots, and the special visceral nuclei by large dots. The latter comprise the motor V nucleus for the jaw muscles, the motor VII nucleus for the muscles related to the hyoid bone and the general facial musculature, and the nucleus ambiguus supplying striated muscles of the pharynx and larynx by way of the IX and X nerves. Three general visceral efferent nuclei are indicated—the dorsal motor nucleus of the vagus under the ala cinerea and the superior and inferior salivatory nuclei. The superior nucleus (*nuc. sal. sup.*) supplies the sublingual and submaxillary salivary glands by way of the VII nerve (pars intermedia of Wrisberg), and the inferior nucleus (*nuc. sal. inf.*) supplies the parotid salivary gland by way of the IX nerve. All of the general visceral efferent fibres are preganglionic sympathetic fibres which end in sympathetic ganglia, whence post-ganglionic fibres carry the nervous impulses onward to their respective destinations.

(From *An Introduction to Neurology*, C. J. Herrick, 1916, p. 156.)

possible to account for all the findings. Involvement of the dorsally placed visceral vagus nucleus (tachycardia, vomiting, gastric hypersecretion), involvement of the adjacent sensory vagus nucleus (pain), of the fasciculus solitarius (taste), the nuclei salivatorii of Kohnstamm (salivation), and the glossopharyngeal and vagus nuclei (diminution of pharyngeal and absence of laryngeal reflexes), account for everything that the patient presents. It is possible that this group of symptoms and signs is produced by neuritis of the glossopharyngeal and vagus nerves. Without post-mortem examination it is impossible to be absolutely certain which of these hypotheses is correct. We incline to the idea that the process is central.

The nervous control of salivation by the autonomic originates in the nuclei of Kohnstamm which lie near the 7th and 9th nuclei.

FIG. 2.—Diagrammatic cross-section through the human medulla oblongata at the level of the vagus nerve, illustrating details of functional localization.

(From *An Introduction to Neurology*, C. J. Herrick, 1916, p. 244.)

The position of the nuclei and the course of their fibres is indicated in the accompanying charts (*Figs. 1, 2, and 3*). The fibres controlling the stomach pass via the vagus and account for the gastric hypersecretion and vomiting. Except at the onset of the disease, the vomiting has always followed an attack of profuse salivation, and might be thought to be due to the filling of the stomach with a large amount of swallowed saliva. In view of the onset with gastric symptoms *unassociated* with salivation and the evidence of hypersecretion by gastric analysis, we feel that a secretion similar to that of the saliva occurs in the stomach. This and the vomiting are therefore separate manifestations, and occur more or less parallel to the salivation.

The tachycardia and the attacks of high blood-pressure are not easy to explain. Much has been written emphasizing the relation of high blood-pressure to gastric crises. However, from a recent piece of work at Dejerine's clinic by Heitz and Norero¹³ it is evident that no close relationship exists between these two manifestations. The rise in blood-pressure and the pain of gastric crises

FIG. 93.—Schematic representation of the vegetative nerve innervation of the head. (Müller and Dahl.)

(From *Vegetative Neurology*, Heinrich Higier, 1919, p. 19. Nervous and Mental Diseases Monograph Series, No. 27. Translated by Walter M. Kraus.)

appear to be separate manifestations dependent upon a common cause rather than causally related to one another. There never was any abdominal pain or tenderness during the patient's attacks. However, pain behind the suprasternal notch has been present. In spite

of this fact, a rise of blood-pressure had not existed during all the attacks of pain, emphasizing the lack of any relationship. The cases reported in the literature include many of the signs and symptoms which we have described. In addition, associated involvement of the 5th nerve has been reported several times.

V.—CONCLUSIONS.

1. Crises occur in patients with syphilis of the nervous system who do not present the picture of tabes dorsalis.

2. A group of signs and symptoms due to disease of the pons and medulla or their nerves occurs in such cases of syphilis of the nervous system, and includes: (a) A profuse secretion of thin and watery saliva; (b) Gastric hypersecretion; (c) Attacks of vomiting related to salivation; (d) Tachycardia; (e) Attacks of high blood-pressure; (f) A sensation of bad taste in the mouth; (g) Pain in the suprasternal notch radiating downward to the left; (h) Diminution of pharyngeal and absence of laryngeal reflexes; (i) Disorders of sensation in the trigeminal distribution.

3. Whether the entire picture described be due to central or peripheral involvement remains to be decided. We incline to the former point of view.

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