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paperhanger by trade. Since June of 1943 he has suffered from a progressively incapacitating disease characterized at the onset by slurring of speech, then by "jumping," weakness and wasting of the muscles of the upper arms and thorax, marked weight loss despite adequate food ingestion, progressive loss of the power of speech, difficulty in swallowing foods, and gradual restriction of food and fluid intake to pultaceous foods to avoid choking and nasal regurgitation. In the last 18 months a habit of a humorless laugh when under emotional stress, a hanging jaw, and a drooping mouth have developed.

The patient always preferred papering to painting, but was boss painter for the Fred Eglehoff Company for 1 1/2 years before World War II. As foreman he mixed the paste into paints and was in the room when spray painting was done by others. During this period of employment none of the men or himself had colic or other symptoms attributable to lead exposure. In November 1942 all occupational exposure to lead ceased when he went into a war plant as a maintenance mechanic. The first symptoms of the present illness began seven months later.*

*The informants for the history were the patient and his wife, and two carbons from the Industrial Commission's file, OD 44675. One of these carbons is a recommendation by Charles F. Barker, who states "...(the patient) first became ill about June, 1942....." This is specifically denied by both the patient and the wife, who state the illness began in June, 1943, months after the patient left theemploy of the Eglehoff Company.

Industrial Commission in December, 1945. With the help of C. H. Knisley and Associates the case was appealed. In June, 1946, the Medical Appeal Board refused to allow the claim. On the advice of Mr. Knisley the case was then referred to us.

On physical examination the patient was found to be a large-framed man with marked wasting and weakness of the muscles of mastication, the neck, the thorax and the upper arms. He made a few grunting noises when trying to talk, and seemed unable to articulate. Most of the time the jaw hung down with the mouth open, the wasted tongue immobile. There was little change in expression, but the folds of the skin were not smoothed. There were constant irregular fisciculations involving particularly the muscles of the thorax, shoulder girdle and upper arm. The jaw jerk was present and gag reflex active. The deep tendon and superficial skin reflexes were all active, with no clonus or pathological reflexes. There was marked myo-edema. No sensory changes, ataxia, or loss of position or vibratory sense were found. He seemed oriented and aware, and annoyed by the humorless laugh that he uttered at frequent intervals during the examination.

Laboratory studies were limited to the study of the spinal ~~puncture~~ fluid, and blood and urine samples for lead. The cerebrospinal fluid dynamics were normal, and the spinal fluid was clear and

Differential diagnosis

The differential diagnosis of the patient's syndrome can be considered as among the many conditions in which progressive wasting and atrophy of muscles occur, and from other diseases in which bulbar signs and symptoms are clearly in evidence. Such illnesses as thyrotoxicosis, arthritic muscular atrophy, syringomyelia, cervical rib, myotonia congenita, progressive muscular dystrophy, peripheral neuritis, infectious neuronitis, cervical pachymeningitis, and secondary lesions of the affected nerves by neoplasm, have certain superficial resemblances to the patient's syndrome, but a clinical knowledge of these syndromes and the studies of the patient rule out such conditions. Myasthenia gravis, tumors of the brain stem, multiple sclerosis, tuberous sclerosis, bilateral vascular lesions of the brain stem as from thrombosis or hemorrhage, central nervous system syphilis, and post-encephalitic and arteriosclerotic Parkinsonismus may cause bulbar or pseudo-bulbar signs; these conditions have all been ruled out by the studies of the patient.

The history, physical findings and laboratory studies are diagnostic of the bulbar type of progressive muscular atrophy.

Etiology of the presenting syndrome

The immediate problem for decision in this case is whether the occupational exposure to lead caused the disease. This problem

in the development of progressive muscular atrophy; (6) the character of the presenting symptomatology; and, (7) the necessary criteria for the diagnosis of lead poisoning.

(1) The occupational exposure to lead

As a painter, the patient presumably had some exposure to lead-containing pigments. Of importance are the facts he preferred papering to painting and did more of the former; the principal exposure to paints was as foreman for the Fred Eglehoff Company, during which time there was no exposure to dry pigments, no spray painting by the patient, and no note of symptoms commonly attributed to lead poisoning in himself or the crew; and the patient has never had such symptoms, other than the paroxyms of abdominal pain at intervals throughout the present illness. It may be inferred that the patient has had less exposure to lead than the average professional house-painter, and that he has neither had symptoms he has attributed to lead exposure other than the present illness, nor had symptoms that might be so attributed.

(2) Time relation between the occupational exposure to lead and the development of the syndrome

The patient had no occupational exposure to lead after leaving the employ of the Fred Eglehoff Company in the fall of 1942, and had none while working as a maintenance mechanic for a

(3) The extent of the present exposure to lead

The illness has been steadily progressive since it began. At the present time the blood and urine lead levels are within the normal range. This is proof that there has been no recent hazardous exposure to lead. It may be concluded that the patient's illness has progressed in the absence of hazardous lead exposure.

(4) The etiology of progressive muscular atrophy

It is generally agreed that the etiology of most cases of progressive muscular atrophy is unknown. The great majority of patients with this disease have had no occupational exposure to lead. Also, the vast majority of patients with hazardous occupational exposure to lead, and with symptoms attributable to such exposure, do not develop this disease. Dr. Kehoe has never seen or had brought to his attention a case of progressive muscular atrophy following undoubted lead poisoning. The experience of James Collier and W. Russell Brain is the same: "I have never seen nor have I been able to find in the numerous records of the National Hospital (in London) any cases in which undoubted lead poisoning has been followed by typical muscular atrophy" (1). However, there are scattered references in the literature in which this question has been elaborated. The first significant reference

it was absent on single determinations. "The (five) cases of progressive muscular atrophy showed no lead (in the urine)."

The first authoritative specific discussion of the problem was by Govers, who stated, "General muscular atrophy may result from lead poisoning, but this form is not, as a rule, progressive in character when the cause has ceased to act. It resembles the ordinary form of progressive muscular atrophy, however, in seat and features and thus differs from the common atrophic palsy of the extensors that is produced by lead." (3) He later described two forms of lead palsy, the first, local muscular paralysis followed by wasting, "the second form, characterized by primary atrophy, occurs especially in the intrinsic muscle of the hand, but is sometimes extensive and irregular in its distribution, affecting many muscles in all four limbs. The wasting is slow, and accompanies, instead of succeeding loss of power" (4). He described no cases with bulbar symptoms. He went on to say "The cases of general muscular atrophy of saturnine origin scarcely ever present any difficulty (in diagnosis) because they usually supervene on severe lead poisoning that has caused characteristic symptoms" (5). He cautioned that "the recognition of lead-poisoning depends, first, on the character of the symptoms of nerve disturbance; secondly, on the existence of other indications of the presence of lead in

lost in an article by S. A. K. Wilson, published in 1907 (7). Because this is the only paper in which the diagnosis of "amyotrophic lateral sclerosis of toxic origin" has been specifically entertained, the cases which were used as examples are summarized below.

Case 1. A compositor, aged 43, began his trade at the age of twelve and continued until incapacitated by wrist drop at thirty-five. He had had colic about three months before. "He ceased work as a compositor, but still had occasionally to do with lead..... Three years ago the weakness and wasting seemed to have commenced afresh, as he began to notice a certain difficulty in keeping his head erect, and a certain feebleness about his shoulders.." On examination the findings included profound muscular atrophy involving principally the upper arm muscles, with some weakness, atrophy and spasticity of the anterior tibial muscles, a steppage gait, a typical extensor plantar response on the left, and a distinct tendency to extension on the right. There were no bulbar signs, "although his articulation has become a little indistinct."

Case 2. A tinsmith of 32 with 16 years experience had never suffered from lead colic, although there was "a good deal of lead in the solder used by him." He developed cramping and weakness in extension in the fingers of the left hand, which progressed to

involvement of other muscle groups of the extremities. Constant painful cramps in the calves continued. The deep reflexes were brisk and the plantar responses remained extensor.

Case 3. The patient was a 37-year-old engine fitter who "had not, apparently, come into direct contact with lead. Twelve years before he (had) contracted syphilis." Eighteen months before he was seen he began to notice weakness and numbness of the extensors of the right middle finger. This progressed to involve the other fingers, and the wrist. At the end of the year there was wasting of the forearm and weakness of the right elbow and shoulder. Three months before he was seen the same changes began in the left hand and arm. He had to hold up his head with his hand to keep it from falling forward as he walked, and he had increasing difficulty in passing water. In addition to the findings of atrophy and slight spasm of the affected muscles he had Argyll-Robertson pupils, a tremulous tongue that appeared slightly atrophic on the right side, and a double extensor plantar response.

Case 4. Was a 63-year old painter and paper-hanger, who had had an attack of lead colic at 60. Six months before he was seen he developed "sciatica" in the right buttock and thigh. Three months later had had cramping of the fingers of the hands, and developed a bilateral wrist drop. On physical examination he had complete double

1. In each there is the same unmistakable (except perhaps, in Case 3) source of poisoning, viz., lead, the action of which had continued over a considerable period.

2. In each the onset is characteristic of an ordinary lead palsy, viz., double drop-wrist. The degree and duration of this lead palsy before the appearance of other symptoms vary greatly in the different cases.

3. The lead palsy.....is followed by progressive amyotrophy of irregular distribution, affecting flexors as well as extensors..... (and) is associated with cramps in the limbs, fibrillations in the diseased muscles, sometimes with involuntary sporadic movements, and with weakness proportional to the wasting....."

We may conclude from study of these cases that Wilson does not follow Gower's criteria in making the etiologic diagnosis, but that he likewise restricts his cases to those "in which the onset is characteristic of lead palsy." In his textbook (7) Wilson adds "two or three more examples have come to my knowledge, since; the latest (1932) concerns a painter who some eight years ago suffered from an attack of drop-wrist (left more than right) and made a partial recovery; on examination I found atrophic palsy of small hand muscles, also of forearms, fibrillary twitches, exaggerated deep reflexes (probable), extensor weakness with little or no spasticity, and unmistakable signs of incipient bulbar paralysis (wasting and

encephalopathy, and one with spastic paraphlegia. The two cases of symmetrical motor neuritis of the upper limbs resemble cases of progressive muscular atrophy. However, Bramwell makes a diagnostic point of the observation that the cases did not progress, after the cause (?) (lead contaminated drinking water) was removed.

These observations may be summarized by the statement that the evidence used in the past to attribute to lead an etiologic role in the development of progressive muscular atrophy is tenuous and presumptive, but that no cases have been so attributed, that have not had peripheral motor palsy the presenting symptom.

(6) The character of the presenting symptomatology

The illness in the present case began with difficulty in articulation. Weakness of the forearm and hands is not prominent, even later in the course of the illness. The abdominal cramping has apparently been considered as evidence of lead poisoning. However, this has continued up to the present, in the proven absence (by blood and urine sampling) of hazardous lead exposure. Cramping of effected muscles may be present in progressive muscular atrophy. In summary, the presenting symptomatology was not peripheral nerve palsy, and was not characteristic of lead poisoning.

(7) The necessary criteria for the diagnosis of lead poisoning

In our opinion the necessary criteria for the diagnosis of lead poisoning are three: (1) there must have been sufficient

ment of the present illness is completely conjectural. No adequate basis for such a conjecture can be found from a study of the patient and the pertinent literature.

Conclusions

1. The patient is suffering from the bulbar type of progressive muscular atrophy.

2. The patient has had less exposure to lead than the average professional housepainter, and prior to the present illness had had no symptoms attributable to lead exposure; the illness did not begin until months after the occupational exposure to lead ceased; the illness has progressed in the absence of hazardous lead exposure; the evidence used in the past to consider lead an etiological factor in the development of progressive muscular atrophy is tenuous and presumptive, but even so no cases have been so attributed that have not had peripheral motor palsy the presenting symptom; the presenting symptom in the present case was not peripheral nerve palsy, and was not even characteristic of lead poisoning; and none of the criteria we consider necessary for the diagnosis of lead poisoning are satisfied in the present case.

3. The assumption that lead has a causative or contributory relation to the development of the present illness is completely conjectural. No adequate basis for such a conjecture can be found from a study of the patient and the pertinent literature.

Henry B. Ryder

8-15-46 - Henry W. Ryder, M. D.

3. Gowers, W. R., A manual of diseases of the nervous system. P. Blakiston Phila., 2nd edition, 1892, 1:474.

4. ib., id. 2:951.

5. ib., id. 2:961.

6. ib., id. 2:959.

7. Wilson, S.A.K., The amyotrophy of chronic lead poisoning: amyothrophic lateral sclerosis of toxic origin, Rev. of Neural. and Psychiat., 5:441-455, 1907.

8. ib., Neurology, edited by A. N. Bruce, Wilkins and Wilkins, 1940, 730.

9. Bramwell, Edwin, Remarks on some clinical pictures attributable to lead poisoning, with special reference to the neurological manifestations of plumbism, Brit. Med. J., July 18, 1931.