PACHYMENINGITIS CERVICALIS (LUETICA) WITH UNUSUAL FEATURES*

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A. H., 44 years old, white, a native of Austria, was admitted to the neurological service of the Mt. Sinai Hospital on August 11, 1920, complaining of pain and weakness of both upper extremities, inability to use his arms, poor vision, especially in the left eye, tremor of the hands, frequent headaches and diminished sexual power.

His *family history* is of no significance.

His *previous history* except for an attack of gonorrhoea at 28 is negative.

Personal History.—Married at 38; his wife gave normal birth to three normal children, all of whom are alive and well. No miscarriages. Syphilis denied by name and symptom.

His *habits* are good. He never smoked or drank to excess.

Present Illness.—About 10 years ago he began to have gastric disturbances, the exact nature of which cannot be determined; these ceased completely three years ago. Five years ago he began to have a "gnawing" pain and weakness in the right shoulder and arm. The pain was very severe and always worse at night; it lasted one year. The weakness was progressive and greatest on raising the arm. He also noticed that his shoulders and upper part of the arm began to grow thinner, the maximum thinness being reached at the end of two years. Eighteen months ago he began to have similar pains and weakness in the left shoulder and arm, which are also becoming progressively thinner, and at the time of admission the pain in them still persists.

The patient states that during the last three years he has on numerous occasions seen "double," and that during the last year his eyesight has not been as good as before, especially in the left eye. For the last three years his sexual power has not been as good

* From the Neurological Service of the Mt. Sinai Hospital, New York city. Presented at a clinical conference, Nov. 3, 1920.

as before. He frequently has headaches. These have no special localization and are not influenced by anything. Recently he has greatly been annoyed by a tremor in both hands. He occasionally has difficulty in controlling his bladder and rectum.

Examination reveals a poorly nourished individual with no evidences of cardio vascular, renal or respiratory disease. There are no petechiae and no icterus. When he extends his arms a fine tremor is noticeable in the hands and fingers. This is probably due to muscular weakness. Fibrillary twitchings are noticeable in whatever musculature is left of his deltoids, pectorals, trapezii, and spinati. The myotatic irritability of the muscles of the shoulder girdle and arms is somewhat increased.

His mentality is apparently unimpaired. There is no aphasia and no dysarthria.

His visual acuity roughly tested shows no gross defects. There is no hemianopsia. No tests were made for color fields.

Ophthalmoscopic examination by Dr. Wolf: Left eye: Large areas of medullated nerve fibers. Right eye: Small areas of medullated nerve fibers upward and nasally; otherwise fundi are normal. *Pupils*.—Both are small, miotic and irregular in outline but equal in size; the left is fixed to light; the right reacts slightly and sluggishly to light. Both react promptly to accommodation. There are occasional nystagmoid movements in both eyes, in the extreme horizontal position. No paralysis of convergence. No external ocular palsies.

The right naso-labial fold is not as marked as the left one. There are no evidences of involvement of the remaining cranial nerves.

Sensation.—Objectively there are no disturbances of superficial or deep sensibility, and no astereognosis. Subjectively he complains of sharp pains in the arms, and of headache.

Motor Functions.—Deltoids weak, left more than right. Pectorals weak, right more than left. Rhomboidei completely atrophied. The strength of the levator anguli scapulae cannot be determined. Trapezii both weak. Sterno-cleido mastoids are apparently normal. Subclavius cannot be tested. Supra and infraspinatus wasted on both sides. Subscapularis, wasting cannot be determined. Latissimus dorsi, weak on both sides. Teres major, weakness cannot be determined. Serratus magnus, right atrophied, left not involved. Triceps, wasted on both sides. Biceps and coraco-brachialis, wasted on both sides. Supinator longus,

both wasted. Extensors of the wrist and fingers, weak on both sides. Pronators, not involved. Flexors of wrist and fingers, apparently not involved. Thenar, hypothenar, interossei and lumbricales not affected on either side. Both hand grips are weak.

The motor functions of the remaining musculature are apparently normal. General myotatic irritability somewhat increased.

F to N test poorly carried out, but this is due to the great muscular weakness in the arms. No ataxia. No cerebellar symptoms. Station and gait normal. No Romberg.

Reflexes: Superficial.—Corneals: present.

Palatal: present.

Abdominals: all but the lower right are markedly diminished.

Bulbo-cavernosus: present.

Cremasterics: lively and equal.

No Babinski or any of its confirmatories.

Deep.—Jaw jerk: lively.

Pectorals, biceps, triceps, and radial—very lively on the left side and present, but diminished, on the right.

KJ's: extremely lively on both sides.

AJ's: lively; right greater than left. Occasionally ankle clonus can be elicited on the right side.—No Kernig.

Sphincters: Patient states that he has some difficulty in starting urination.

Laboratory Findings.—

Blood Wassermann on two occasions suspicious (Mt. Sinai serological laboratory). Cerebro-spinal fluid: 0.1 to 1 c.c. Wassermann, 4 plus. Globulin, one plus. Five lymphocytes to the cubic millimeter. Fluid obtained under normal pressure.

Urine.—Normal.

Temperature.—Normal.

Blood Pressure.—No difference on the two sides: 105/72.

Electrical Reactions.—Dr. Harris reports: Faradic response in the shoulder girdle muscles and the arm of the left side is prompt but a stronger current is required. Galvanic response in the right shoulder girdle muscles is normal, while on the left side the response is prompt, but the ACC is greater than the KCC.

There is no tenderness on percussion of the skull, spine or any other part of the osseous system.

X-ray examination of skull, cervical and dorsal spinal column is negative.

There are no vasomotor or trophic disturbances.

Bedside Notes.—

Aug. 15, 1920, received intravenously arsenobenzol .4 gm.

Aug. 21, 1920, received intramuscularly Hg. salicylate gr. .i.

Note the wasting of the muscles of the shoulder girdle and both **arms** in contrast to the well preserved musculature of the rest of the body.

Aug. 25, 1920, received arsenobenzol intravenously .4 gm.

Aug. 29, 1920, received intramuscularly Hg. salicylate gr. .i.

Aug. 30, 1920, discharged from the hospital with the following

note: Pains almost entirely disappeared. Beginning increase in power. Transferred to the dispensary for further anti-specific treatment.

Sept. 9, 1920, blood Wassermann in the dispensary negative.

The case may then be summarized as follows: A 44-year-old man began to have some vague gastric disturbances 10 years ago, which lasted until 3 years ago. Then he began to have "gnawing" pains and weakness in the right upper extremity (5 yrs. ago).

Note the atrophy of both shoulder girdles, and the muscles of both arms.

This lasted one year. He then began to have similar pains and weakness in the left upper extremity (18 months ago) which persisted up to the time of admission to the hospital. During the last year his vision (in the left eye) has not been as good as previously. For the last 3 years he has frequently had generalized headaches. His sexual power has been poor during the same period. Some years ago he had frequent urination for one week. His pains are always worse at night. Owing to his muscular weakness he has lately been incapacitated for work.

He presents the following positive objective findings: Pupillary changes, slight nystagmoid movements in both eyes; some facial weakness on the right side; atrophy of the trapezii (slight); marked atrophy of the supraspinati, deltoids, biceps, triceps and supinators, especially the left, with weakness of all movements controlled by these muscles; fibrillary twitchings in the muscles of the shoulder girdle, and partial R.D. in the involved muscles; some sphincteric disturbances; increased tendon reflexes in the lower extremities, with an occasional clonus in the right ankle; a 4 plus Wassermann and an increased globulin content of the spinal fluid.

On this history and findings the diagnosis of cerebro-spinal lues and pachymeningitis cervicalis was established.

In making this diagnosis the following conditions have to be excluded:

1. Amyotrophic lateral sclerosis. Pain in the arms, which is so marked a feature in our patient, is not a symptom of amyotrophic lateral sclerosis. The absence of a spasticity or rigidity in the involved extremities, as well as the absence of bulbar symptoms after five years' duration also speak against such a diagnosis.

2. Spinal tumor. This can be excluded on the grounds that the patient shows evidences of cerebro-spinal lues, that there is no sensory level and no objective sensory disturbances. The duration of the disease, the absence of signs indicating increasing cord compression are all against such a diagnosis. As a general proposition, we may also add that the pain in spinal tumors is much more lancinating in character than in our case.

The same diagnostic criteria are also applicable in the exclusion of circumscribed serous meningitis and circumscribed meningeal cysts.

3. Syringomyelia. Against this diagnosis are, absence of spinal deformity, absence of sensory dissociation, absence of vasomotor phenomena and of trophic disturbances of the skin, bones and joints; absence of oculo-pupillary symptoms, and the absence of a spastic paralysis of the lower extremities, and the fact that the muscular atrophy did not begin in the small muscles of the hands, as is usually the case in the Aran-Duchenne type of progressive muscular atrophy (commonly seen in syringomyelia of the cervical cord.)

4. Multiple neuritis. The absence of any of the usual etiological factors of multiple neuritis, the absence of objective sensory changes

and of nerve tenderness, and the increased deep reflexes in the lower extremities, are sufficient to exclude this condition.

5. Chronic disease of the anterior horns of the cord, such as chronic anterior poliomyelitis, and chronic progressive muscular atrophy are excluded by the absence of pains in the latter, and the presence of exaggerated deep reflexes in the lower extremities in our patient, as well as by the presence of ocular symptoms.

6. Tuberculous pachymeningitis. This condition may present a clinical picture not unlike that seen in our patient. The absence of evidences of tuberculous disease in the vertebral column and in other parts of the body, and the presence of evidences of syphilis, are sufficient to make the differentiation.

7. Muscular dystrophy. In this condition there are no pains and no fibrillary twitchings, no R.D. and no increased deep reflexes, and no cranial nerve involvement.

8. Brachial paralysis from cervical ribs. Radiography of the spine failed to show the presence of any supernumerary cervical rib, nor were there any evidences of inequality of the radial pulses such as are commonly found in this condition.

Note.—Examination in the dispensary on January 27, 1921, after he has received three courses of intensive salvarsan and mercury treatments, show no new neurologic findings. Subjectively he feels much better. He has been free from pain for the last four months. The disease has apparently been arrested. At this writing he is still receiving antiluetic treatment.

We may therefore conclude that this case is one of cerebro-spinal lues, the prominent features of which are a meningomyelitis involving C5, C7 and C8 segments of the cord, with participation of the anterior and posterior roots as well as the anterior horns at the same levels. The exaggerated knee jerks with tendency to ankle clonus on the right side seem to point some involvement of the pyramidal tracts.

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