REPORT OF A CASE OF SYRINGOMYELIA¹

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Although the symptom complex of syringomyelia is well understood both from the observations of Morgagni, Santorini, Schultze, Schlesinger and other authors, this case is presented as being of some interest. In the first place it again brings before us a typical picture of the disease and also eminently verifies its chronic course, as the patient has been under the personal observation of the writer for twenty-three years, during which time there has occurred little or no material advance in the progress of the disorder.

The patient is sixty-four years of age, married, and a shoemaker by occupation. He was seen for the first time in June, 1886, and since this date he has been under my observation, a period of twenty-three years, without showing any material advance in the progress of the disease.

The personal history is that of a strong, healthy man, without excesses of any kind, leading a quiet home life and attending to his business duties every day for over forty years. There is no history of any traumatism or exposure to wet or cold. Venereal disease is denied and inquiries as to his heredity does not reveal the presence of tuberculosis, syphilis or nervous disease in the family.

The first symptoms were noticed thirty years ago (October, 1878, to January, 1879), which came on gradually, consisting of general debility, a wornout, tired feeling and dizziness. He was inclined to lie down most of the time, but never felt rested. About the same time he observed that his arms and hands were growing weaker and that he could not use the awl so well in making shoes. No change was noticeable at this period in the appearance of the shoulders, arms or hands. In January, 1879, the voice became husky and he has spoken in a harsh, unequal tone ever since. Six months later (June, 1879) sensations of numbness and cold appeared in the left shoulder, which gradually extended to the neck, arm and hand of the same side. This sense of numbness was especially severe and troublesome on the left side of the neck, so that he often resorted to very hot applications for relief. On one occasion after the removal of a poultice it was discovered that his neck had been badly burned, though at the time

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he had not noticed any sense of pain. After this, moreover, he frequently received cuts or injuries on his hands, without knowing it at the time, until his attention was attracted either by the flow of blood or ulceration. About September, 1879, a decrease in size and a difference in shape of the shoulders were noted, the left being more prominent than the right. This atrophy of muscles, first noticed in the deltoids, slowly progressed, involving the trapezii and scapular muscles, the interossei of the hands and still later the latissimus dorsi, the pectorales and extensors of the arms. In October, 1885, a painless ulcer formed in the helix of the left ear, which went on to perforation, some time elapsing before it was healed. During the next three years, while under my observation, there gradually developed an increase in the muscular weakness, with atrophy, a sense of heaviness in the tongue and difficulty in swallowing.

FIG. 1.

Examination.—The patient is a man of medium size, weighing one hundred and forty pounds. He speaks in a harsh, impure and unequal tone. On the left side of the neck is a keloid scar, the result of a burn and on both hands several other scars are noticeable, besides fissures and abrasions which heal very slowly and imperfectly, remaining some time as open sores and he states these cause no pain (Fig. 1).

Nervous System—Motor Functions.—With eyes open he shows no ataxia in gait or on standing, but with eyes closed the equilibrium is not good. There is marked weakness in the upper

extremities and the movements are imperfect and limited. The grasping power of the hands is weak—right 18.20, left 18.11, and compared to the test made twenty years ago, viz: right 30.32, left 28.26, quite a difference is shown. The little and ring fingers of the right hand are slightly flexed and their movements are limited. All the interossei are wasted, especially the third and fourth of the right hand. The muscles attaching the upper extremities to the trunk are also much atrophied and this is more marked in some muscles and in some parts of muscles than in others; the lower part of the trapezius is much more affected than the upper, the upper part of the pectorales more than the lower. The left cheek feels thinner and less firm than the right, and there is marked narrowing of the palpebral fissure of each eye with enophthalmos. There is no appreciable difference between the two sides of the tongue, though the patient states that the left side does not feel as natural as the right. When the tongue is protruded to its fullest extent it deviates slightly to the right. There is a paralysis also of the palate, the uvula being sharply drawn to the right. An examination of the throat shows a complete paralysis of the left superior constrictor of the pharynx and of the left palato-pharyngeus. There is also found a paresis of the adductors of the left vocal cord. The mechanical irritability of the atrophied muscles is increased and fibrillary twitchings frequently occur.

Electrical examination reveals a general reduction in the strength of the contractions produced both with the faradic and voltaic current. The inferior segment of the trapezius fails to contract with the strongest faradic current. Moreover the only muscles which give a true reaction of degeneration are the interossei and the thenar and hypothenar groups. The musculature of the lower extremities is firm, no wasting being present.

Reflexes.—The patellar tendon reflexes are exaggerated; the deep reflexes are normal and the Babinski not obtained. There is no clonus.

Sensory functions.—He complains of feeling cold and states that the left side of face, head, neck and both hands are numb. The skin of the left side of the face, arm, shoulders, forearm and both hands, also left side of trunk and neck, as far forward and backward as the median line and as far downward as the fourth rib in front and the sixth rib behind, is anesthetic; a region corresponding to the distribution of the cervico-brachial plexus, of the intercostal and fifth cranial nerves (Fig. II). This region of insensibility is somewhat ill-defined on the trunk behind, but on the face its limits are sharply defined. On the right side there is no loss of sensation except on the bridge of nose and forehead, just over the median line, but on the dorsum of the right hand, over the anterior and lateral aspects of the arm, the sensation is not natural. There is also marked analgesia occupying the same

distribution as the anesthesia, with the preservation of tactile sensibility in many places. Pressure, localization and muscular senses are normal.

Sensibility to Temperature.—There is a complete loss of temperature sense over entire circumference of hands and left side of neck, partial over both deltoids, anterior and lateral aspects of arms and forearms, and left side of face. Sponges dipped first in hot, then in cold water, water test tubes containing hot and cold water, were made use of in these tests. Warm objects seem cold in various areas and in certain places the test tube of hot water was frequently recognized after prolonged contact.

FIG. 2.

Special senses.—The senses of taste, smell and hearing are not impaired. His vision is not acute and he has to look at an object some little time before recognition is complete. The pupils are small, but active to both light and accommodation. The movements of the eyes are normal and there is no nystagmus. The ophthalmoscope does not show any changes in the fundus of either eye.

Diagnosis.—From the character of the symptoms—motor paralysis with atrophy, trophic changes, anesthesia, analgesia, thermoanesthesia with the preservation of tactile sensibility in many places, there remains no doubt as to the diagnosis being other than that of a syringomyelia from a gliosis. Furthermore, this process at first localized in the cervical enlargement has extended, so as to involve the tenth, eleventh, twelfth and fifth cranial nerves.