THE SPASMODIC TYPE OF SYRINGOMYELIA

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Syringomyelia was divided by the great authority on this disease, Hermann Schlesinger, into six main types, namely, (a) typical syringomyelia, (b) motor, (c) sensory, (d) trophic, (e) tabetic, and (f) pachymeningitic syringomyelia.

In 1900 Dr. Pierre Marie presented and identified the first five cases of this type, and shortly afterwards his pupil, Guillain, wrote an elaborate thesis, giving a complete description of these same patients under the name of the spasmodic type of syringomyelia. In 1906, with Alquin and again with Raymond, Guillain added two more cases to the series, while Raymond with Francois reported the eighth example in the same year. Verger followed another case, which showed similar symptoms during life, to autopsy and records a central glioma as the actual pathological condition, while in 1908 Alexander Bruce in the *Review of Neurology*, wrote concerning a patient with some resemblance to Guillain's series, who may probably be regarded as either possessing the same or a related type of the disease.

In presenting this patient, whose symptoms correspond to the typical instances of Guillain, the different features of the case are each described, followed by immediate reference to the corresponding condition in the cases reported.

I. The most important characteristic of the spasmodic type of syringomyelia is undoubtedly what may be termed *the attitude of the patient*.

This little girl is sixteen years old and the disease has steadily advanced since the age of five, the slow progress being a feature of this condition.

As she stands up her deformity is apparent. The head slightly bent forward nestles between the strongly marked borders of the trapezii. The thorax, if viewed posteriorly, arching forward in its upper part, shows a marked concavity in its superior part in front, and this has been termed *thorax en bateau*.

The spine exhibits a most extreme degree of scoliosis, the curvature being to the left side.

The arms are drawn to the side of the body, the right, which

is at present the most affected, lying across the body with the hand towards the symphysis pubis, and exhibiting marked flexion at the elbow, a position in which it is rigidly fixed.

On the left side the elbow is already commencing to present the same signs, but is far less marked in rigidity than the right.

The shoulders are pushed forward and upward, slightly rotated inward.

Contrasting this attitude with that described by Guillain one notes the similarity in all respects as regards head, shoulders, thorax en bateau and position of the arms. Also the unilateral advance was present in four of his five cases, while the scoliosis was marked in one and present in four others.

II. Turning from the attitude of the patient to the minute study of the arms, emphasis must be laid on the characteristic position at each joint on the right side.

1. The shoulder raised by the trapezius pushed forward on the thorax and rotated slightly in by the pectoralis.

2. The elbow in a state of rigid flexion, although if the joint is still further flexed then some extension is possible.

3. A most important sign at the wrist, namely, *hyper-extension of the hand on the forearm*, which will be referred to again.

4. The position of the fingers, namely, the three inner, flexed firmly into the hand, while the fourth finger shows incomplete flexion; this being most marked at the terminal phalanx, and with the thumb, which is adducted, forming a position described as a "pair of pincers."

These striking points are the main diagnostic signs of the disease together with those already referred to under the general attitude of the patient, and they are reported in all the definite cases of the disease and may be said to be invariably the same.

True, the exact position of the fingers may show some alteration due to the period and advance of the disease, so that this may be described as the typical position, and in a more advanced case one may expect to find more marked flexion of the fingers, in which the index will be included, as is well shown in one case of Guillain's. This *hand* has been present in the cases examined post-mortem, and is characteristic of syringomyelia spasmodique, and while it might possibly be present in pachymeningitis, yet to the present time it is diagnostic of the former disease.

The hyper-extended wrist is also striking and equally important, and it may be produced by syringomyelia and conditions copying this, as glioma, pachymeningitis, and by acute poliomyelitis.

It is easily distinguished from the common cerebral type of hand seen in diplegia, or hydrocephalus, in which marked flexion frequently is found.

III. Considering now the muscular power of the right arm, movement at the shoulder is limited; elevation is possible, but

adduction and abduction are very weak, while no power of rotation is present. At the elbow a very small extent of flexion and extension is possible, and, strange to say, if the power is tried, despite the contracted condition of the biceps, yet the extension of the triceps is greater than the biceps power, and if the arm be still more flexed it is noticeable that the extension is the stronger, so that the muscle in contraction is weaker than the muscle which has given way. The same fact applies to the other arm.

At the wrist slight extension and flexion are possible, and here the contracted extensors are more powerful than the flexors of the reverse condition to that in the upper arm.

The last three fingers can be flexed to a certain degree and partially extended; the first finger to a greater extent, and the adducted thumb also to a certain degree.

But the characteristic pincer movement is better shown by the less involved hand, as the right while presenting the same position has passed into a condition of rigidity where the movement of the thumb is difficult.

In Guillain's series the muscular power varied according to the extent and stage of the disease, but he also lays stress on the fact that it is the spastic and rigid condition far more than the atrophic condition which causes the loss of power.

IV. The trophic condition of the muscles is such that no actual wasting of any group of muscles, beyond disuse wasting, can be made out, and the condition conforms to an upper rather than a lower motor condition. True, the biceps is in a peculiar contracted condition and the triceps is extremely small, and the extensors appear wasted, but there is no valid proof that such is the case, with the exception that I have observed fibrillation of the left triceps, and this is characteristic of a lower motor neurone disease.

But to offset this important point it must be stated that despite the equally spastic condition of the legs, yet there is no corresponding atrophic condition there, and they look large and well nourished, although one must bear in mind that the affection is much more recent.

Secondly, it is necessary to add that the cases described by Guillain showed the same positions of the upper extremity, and he claims that such is always a sign of local lesion in the cervical region of the cord and is produced by local processes only.

Thirdly, it appears to me to be characteristic of a type of syringomyelia to have atrophic muscles present with much diminished muscular power; without demonstrable wasting of a lower motor neurone type, and by that I mean for instance with interosseal paralysis without markedly hollowed out spaces between the bones, and with electrical reactions which are not typical of degeneration.

The explanation must be that in this disease either certain

fibers of a muscle waste and those remaining give the electrical reaction, or else there must be a situation between the pyramidal tract and the nerve cells of the muscles, whose destruction causes the spasticity of the first and the peculiar wasting of this type of case without changed electrical reactions.

Guillain's cases evidently were of the same type, for with the same advanced condition of paralysis he only describes actual wasting in a few muscle groups, usually the small muscles of the hand or isolated forearm groups, although late in the disease more marked atrophy occurs.

This condition of spasticity and rigidity with moderate wasting, and with no reaction of degeneration, in a disease making very slow advance, negatives the diagnosis of amyotrophic lateral sclerosis, and may be said to be decidedly characteristic of syringomyelia of this particular type.

V. The electrical reactions are most interesting, for all muscles react to faradism and to galvanism.

There is no polar change and no sluggish contraction, and, in fact, the muscles reacted to a current that was approximately normal.

In no case did the authority prove any R. D., even when marked wasting was present, although a stronger faradic current was required for some of the more advanced muscles or when there was definite atrophy.

VI. The condition of the left arm will not be described, except to say that it is beginning to show in an early condition the same tendencies as the right. The elbow contraction is already present and adhesions at the joint are easily broken down. The hand shows the typical three-finger flexion and pincer position of the first two digits, and the wrist is commencing to show the extension position. Fibrillation has been observed in the triceps.

VII. The muscles of the back must be extremely weak since the scoliosis is so marked, but the abdominal muscles are apparently normal.

VIII. The lower limbs are well nourished and are spastic and rigid, particularly at the knees and ankles. There is no wasting. Movements are possible at all joints, no fibrillation is visible.

The gait is rather that of a double hemiplegic movement, taking place from the spine and sacro-iliac joints by rotation.

Such a gait is perhaps more typical of a syphilitic pachymeningitis, but the scoliosis, the anesthesia, the age, the slow advance of lesion, the arm position and the height of the lesion, together with the absence of cerebral specific manifestations negative that diagnosis. The more marked affection of both pyramidal tracts would also explain its presence here.

IX. The reflexes are as follows: Eyes normal. Jaw jerk present. Biceps present, left slightly greater than right. Triceps present, left slightly greater than right, probably due to extreme rigidity of the right side.

Knee joints both increased. Ankle clonus present and double extensor plantar responses, *right more marked than left*.

In the reported cases the reflexes of the lower limbs have been increased in some cases and decreased in others, while those of the upper limbs were absent in three cases and increased in one.

X. The bladder and rectum are at present normal; there was a period when micturition required to be performed more frequently, but she has recovered from this. The bladder is frequently affected in the later stages, as the patient may suffer from incontinence and from cystitis due to trophic disturbances, such as trophic abscess and bleeding.

XI. *Sensation*.—In Guillain's groups four of the cases gave syringomyelic areas, although in one case he had difficulty apparently in making out temperature sensations, which were more markedly defective as regards heat.

In the fifth case, early in the disease no sensory impairment was noted beyond slight anesthesia in the upper part of one thigh. Before death, 21 years later, touch, though retarded, was normal except over the left hand.

Pain was lost over some part of the body and retarded generally. Temperature over the whole body was either interpreted as touch or more frequently as cold.

Turning to this patient one may state that correct judgment of anesthetic areas is difficult, as the child is a foreigner. There is much retardation, particularly in certain areas, and especially of temperature.

No tactile anesthesia was made out.

Analgesia was also absent, though there was an area on the right thigh where it apparently was doubtfully present.

But to temperature sensation much importance is due, as while cold seemed everywhere present, and no definite mistake was made, yet heat, as in Guillain's case, was affected.

Frequently heat was termed cold, at other times touch, and this all over the body; while on another test the mistake was rectified in places, not in others. An extension of the area stimulated in certain cases produced a correct reply.

On the right side one error was practically constant, namely, below the waist, above the thigh; heat was termed touch, but in passing upward gradually it was called cold, while above the nipple it was recognized as heat. If the temperature was raised to a higher point the test was less successful and the reply might be correct.

On the left side the same phenomena occurred, but less clearly. Finally, to a faradic current, the right side was claimed to be more sensitive than the left from the mid line.

Two features require impression here, as they are of great value in these syringomyelic cases: (1) The retardation of sensation; (2) the non recognition of heat as such, but its perception

without temperature association as touch or otherwise as cold. (3) One may note that in early cases sensation may be normal, that temperature disassociation may appear first, and that beyond a difficulty in distinguishing heat as such, that no definitely bounded area may be involved, and that retardation may be a distinctive character.

XII. No trophic areas have been discovered, except a large burn scar on the arm and one on the leg, from which no conclusion could be drawn. In Guillain's group whitlows were present in some cases.

XIII. The cranial nerves are normal, with the exception that an undulating movement is present on the tongue, which may be abnormal. It is interesting to note that the same condition, an undulation, not a fibrillation, is quoted in one other case. Some of the other patients have had cranial nerve signs, as wasting of the tongue, etc., though they are uncommon.

The mental condition is absolutely normal and unusually bright and clear.

XIV. Regarding the diagnosis, it is unnecessary to expand, as each possible case has been referred to as corresponding signs appeared—taking the attitude, the arm condition, the scoliosis, the spastic legs, the fibrillation, the normal electrical reaction, and the irregular disturbances of sensation. There are the two diseases which remain to be differentiated, namely, syringomyelia and pachymeningitis cervicalis. Guillain believes their separation in most cases is impossible, as either by themselves or together they may lead to a similar set of signs. But in this case the marked scoliosis, and the leg condition with the absence of definite atrophic wasting, all point to the correctness of the disease being a definite case of the spasmodic type of syringomyelia.

XV. In conclusion, let the history end where most commence, namely, with the course. It began at the age of five and she is now sixteen. Pain in the back of the head and the curving of the spine were together the earliest signs, and one of Guillain's cases which went to autopsy commenced with the same pain. These disappeared in the girl's case. Gradually at the age of six the arm began to be affected and she complained of difficulty in moving it. The legs were attacked at about ten years of age and the right arm has only shown any affection for three years. For the last year there has been slight difficulty in micturition.