Hematomyelia as a Complication of Syringomyelia: Gowers' Syringal Hemorrhage

Case Report

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Bleeding into a pre-existing syringomyelic cavity in the spinal cord can occur, and the resulting clinical picture may be both catastrophic and confusing.

In 1903, Gowers stated, "I wish to direct . . . attention to some evidence that the existence of these cavities [syringomyelic] occasionally induces the occurrence of one of the gravest lesions of the spinal cord, hemorrhage, and determines special, perhaps characteristic, symptoms. I have seen several cases in which the peculiar character of the symptoms can be best understood by ascribing them to a hemorrhage into such a cavity . . . ." Four patients were discussed, but in only the first was the occurrence of hemorrhage verified pathologically. This was a coachman who developed an ascending paralysis with intense pain and died 5 weeks after. There was no history of symptoms suggesting syringomyelia and no trauma. At autopsy, syringomyelic activation was found in the spinal cord from the cervical to the lumbar region and the cavity was distended with hemorrhage throughout. The remaining 3 cases had symptoms and signs suggesting to Gowers that a lesser hemorrhage had occurred into a pre-existing syringomyelic cavity; however, the picture was much less clear cut than in the first patient and there was no pathological proof for the diagnosis.

Modern text books on neurology mention this possibility as a rare complication of syringomyelia but give no references to verified cases. To our knowledge, no examples of surgical confirmation of such a lesion have been recorded in the literature. Grinker and Bucy state, "An apoplectic-like increase in all symptoms and rapid extension of the lesion to involve new structures may result from hemorrhage into the syringomyelic cavity, either spontaneous or precipitated by trauma. Wilson mentioned in his discussion on hematomyelia that he had made the diagnosis of hemorrhage into a syringomyelic cavity several times but he had never had any chance to prove it. Wilson referred to this condition as, "Gowers' syringal hemorrhage."

We have recently had the opportunity of treating a patient with a sudden onset of monoplegia and loss of sensation in the right arm following trauma. Verification of the lesion at operation together with the fortuitous circumstance of having a myelogram done some years earlier provided a striking confirmation of Gowers' conception of hemorrhage into a pre-existing syringomyelic cavity.

Case Report

M.N.I. #64-1153, J. R., a 39-year-old married business executive was admitted to the Montreal Neurological Institute on July 9, 1964. He complained of pain in the neck and right shoulder with numbness and paralysis of the right arm. These symptoms had appeared suddenly after a fall 6 days before his admission.

History. At age 14, the patient was said to have had a "slipped epiphysis" in the left hip and he had a marked limp favouring the left leg. He wore a cast for one year. In June, 1957, he began to have fairly typical right-sided sciatica. He was seen by an orthopedic surgeon who diagnosed lumbar disc disease and necrosis of the head of the left femur. A Moore prosthesis was inserted into the left femur in October, 1957, and the patient's gait improved. In February, 1958, lumbar discectomy was carried out at L4-5 at another hospital and the patient's sciatica was relieved. In September, 1958, the patient noted the onset of severe neck pain radiating into both shoulders and upper arms. This responded well to physiotherapy. At that time he also found that occasionally when he flexed his neck he felt a sharp shooting pain radiating from the neck down his back into the backs of both legs and feet. This complaint continued up until his present admission. In January, 1959, he experienced intermittent tingling of the tips of all the fingers of the right hand. This became continuous a few months later.

In April, 1960, the patient had a subtotal gastrectomy for perforated duodenal ulcer. He showed no great change until March, 1964, when he again began to have neck pain radiating into both shoulders and upper arms. This was usually aggravated by flexion of the neck. At times the patient felt that his right arm and hand were weak. In March, 1964, he also noted that his right foot would "go to sleep" easily. During the months just before his admission for the present complaint, he was treated with physiotherapy and exercises by several physicians for presumed cervical disc disease.

On July 3, 1964, the patient had slipped and fallen on a tile floor, landing on his abdomen. He cut his lip but did not lose consciousness. It is probable that there had been sudden violent neck extension. He got up immedi-
ately but noted that his right arm was completely without feeling and he felt severe pain in the back of his neck. When he awoke next morning he felt a sensation of burning in the skin over both shoulders, particularly in the right scapular and left clavicular areas, with tingling in the fingertips of the left hand. That morning he also noted that his right arm was extremely weak. He could not tell the position of his fingers or hand without looking at them. He visited his family doctor that day. X-rays of the cervical spine showed some narrowing of the C6-7 disc space but no fracture. He was fitted with a cervical collar and told to rest but there was no improvement. On July 7 he began to have an uncomfortable tingling sensation in the anterior part of the right upper arm. Coughing, sneezing or laughing increased the pain in his arm and shoulder. He was admitted for investigation because of these symptoms.

Examination. The patient was alert and cooperative, but complained of pain when he moved his neck and a sensation of burning across the tops of both shoulders. The cranial nerves were normal, and there was no Horner's sign. There was complete paralysis of abduction of the right shoulder with severe weakness of the right deltoid, levator scapulae, biceps, triceps, supinator and pronator muscles as well as those of the wrist extensors and flexors. There was weakness of the small hand muscles but this was less marked than in the proximal arm muscles. There was only slight weakness of adduction of the shoulder. The left upper limb was strong except for some weakness in the small muscles of the hand, and movements of the wrists and fingers were normal. Tendons were noted over the right upper arm and shoulder. There was no weakness in the legs. Deep tendon reflexes were all absent in the right arm. In the left arm only the triceps jerk was present. The abdominal reflexes and the knee and ankle jerks were present and equal on both sides. The plantar responses were flexor. The right arm was almost completely insensitive and he had no idea where it was. There was a cloak-like area of disturbed sensation over the upper trunk on both sides including the whole right arm as well as the ulnar half of the left hand, particularly when tested with cold. There was marked impairment of pain sensation in the same areas. The skin over the whole upper part of the right arm and shoulder showed marked hyperalgesia. Position sense was absent in the right arm including movements at the elbow but not at the shoulder. There were no long tract signs and no bladder or bowel symptoms.

In summary, the findings indicated a mixed root weakness of the right upper limb with sensory impairment especially to pain and cold over C5 to T7 or 8 on both sides, plus weakness and reflex changes in both upper limbs. This was felt to be in keeping with a central cord lesion in the lower cervical and upper thoracic region. The possibility of traumatic bleeding into a preexisting syringomyelic cavity was entertained.

Routine laboratory studies were normal. Cervical spine x-rays showed thin disc spaces between C5-6 and C6-7. The chest x-ray was normal. X-rays of the lumbar spine revealed some degeneration of all the lumbar discs with osteo-arthritis of the lumbar apophyseal joints. The head of the left femur had been replaced by a metal prosthesis. Myelography was carried out through T1 (Fig. 1A). Cerebrospinal fluid protein at the time of myelography was 132 mg. per cent.

Operation. On 11 July, 1964, the cervical spinal cord was explored through a bilateral laminectomy from C4 through C7. There was no evidence of fracture or injury to the bones or soft tissues. After the laminectomy had been carried out, the dura was seen to be quite transparent at the C5 to C7 levels and the spinal cord could be seen intimately applied to dura which was bulging and tight. Normal pulsations could be seen above and below the most distented segments at C5 through C7. When the dura was incised the swollen cord bulged into the opening. There was an area of pale yellowish discoloration in the dorsal surface of the cord at the C6 and C7 levels, darker at C7. This seemed to occupy almost the entire dorsal aspect of the cord. On palpation there was a fluctuant feeling and appearance. There was no evidence of any disc protrusion impinging on the cord from in front. Aspiration of the cystic area of the cord was carried out with a #25 short hypodermic needle in an avascular area at the level of C6 about 1 mm. to the left of the midline. When a 2 cc. of reddish-brown old bloody fluid was aspirated there was a marked decrease in tension in the cord mainly at the level of C6 and 7 (Fig. 2). It then became apparent that the most involved part of the cord lay on the right side of the midline at the level of about the lower part of the lamina of C6 and C7. An incision was made into the cord in the midline dorsally at the level of C7 and an opening about 9 mm. in diameter created into the cyst cavity. Following this the dura was left wide open. The closure was carried out in layers as usual.

The postoperative course was one of gradual improvement. There was marked decrease in limitation of neck movement and in pain in the neck and shoulders. A feeling of itching and burning persisted across the tops of both shoulders. When the patient was discharged 20 days after operation, he no longer had any weakness in the left hand or wrist. There had been slight increase in strength mainly distally in the right arm. The sensory deficit noted over the trunk and both arms had decreased in intensity although there was still marked loss of pain and temperature sense in the right arm and hand with a preserved position sense in the right fingers, wrist and elbow.

Later we obtained the myelogram performed at another hospital at the time of the patient's lumbar discectomy in 1958. This clearly showed the presence of a tubular enlargement of the lower cervical spinal cord (which had been recognized and reported in 1958). This indicated that the syringomyelic cavitation had been present at least 6 years before his present illness (Fig. 1B).

At the last follow-up examination, 1 year after operation, the patient was actively employed at his former position, but his right hand was still practically useless. His complaints included a troublesome feeling of itching about the neck mainly across the shoulders and sometimes across the upper chest, a feeling of prickling or tingling in the right hand and forearm, intolerance to anything hot or cold touching the right hand, occasional feeling of tightness in the skin of the right foot and lower leg with occasional leg pains and some persisting discomfort on neck movements. He was still unable to carry out sexual intercourse satisfactorily due to failure of erection.