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 Neurology Service 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 rightsided 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-

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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 uncomfor-
table 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 ad-
mittted 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, supina-
tor 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 proxi-
mal arm muscles. There was only slight weakness of adduc-
tion of the shoulder. The left upper limb was strong except for some weakness in the small muscles of the hand, and paralysis of the thenar and hypothenar mus-
ters 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 dis-
turbed sensation over the upper trunk on both sides in-
cluding 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 area as referred pain was elicited by palpation in the right upper back. Position sensation was absent in the right arm includ-
ing 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 impair-
ment 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 cen-
tral cord lesion in the lower cervical and upper thoracic region. The possibility of traumatic bleeding into a pre-
existing 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 lumbosacral spine revealed some degeneration of all the lumbar discs with osteo-arthritis of the lumbar apophy-
seal 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 trans-
parent 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 dis-
coloration 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 the cord had 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 improve-
ment. There was marked decrease in limitation of neck movement and in pain in the neck and shoulders. A feel-
ing 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 de-
creased in intensity although there was still marked loss of pain and temperature sense in the right arm and hand with a maintenance position sense in the right fingers, wrist and elbow.

Later we obtained the myelogram performed at an-
other hospital at the time of the patient's lumbar dis-
coideotomy 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 opera-
tion, the patient was actively employed at his former position, but his right hand was still practically useless. His complaints included a troublesome feeling of itch-
ing 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.
Hematomyelia with Syringomyelia

The main findings on examination included marked weakness of abduction of the right shoulder so that he could lift his arm parallel with the floor but no higher. There was severe residual weakness of the right biceps, triceps, brachioradialis, flexors of the wrist and small muscles of the hand. The left arm and hand were strong. Reflexes were still absent in the right arm and present only in the triceps in the left arm. Reflexes in the legs were normal. There was still a cloak-like disturbance of sensation over the upper chest mainly for pain and temperature sense which extended down to the nipple line on the right but only about half as far down on the left. Sensation in the left arm and hand was normal. Sensation in the right arm was reduced to all modalities up to the elbow. From there up to the shoulder it was better preserved on the ulnar surface of the arm than the radial. Position sense was absent in the right fingers, occasionally correct in the right wrist and more often correct in the right elbow. He had learned to write with his left hand.

Discussion

From the history, physical findings and the lesion observed at operation, it seems clear that this patient suffered a traumatic hemorrhage into a syringomyelic cavity that was already present before the trauma. The fact that an enlargement of the cervical cord was demonstrated on a previous myelogram done 6 years before the patient’s traumatic episode would seem to establish this beyond doubt. Unlike Gowers’ coachman, the bleeding in our patient was probably very slight and resulted mainly in disruption of the grey substance on the right side of the cord in the cervical segments supplying the right shoulder and arm.

The mechanism of the hemorrhage in our patient was most likely traumatic rupture of one or several vessels lying in the wall of the syrinx. Netsky has pointed out the frequent occurrence of clusters of vessels lining the walls of these syringomyelic cavities. Impressed with the frequency of these intramedullary vascular anomalies in the walls of syringomyelic cavities, he postulated that they were associated with the development of “true syringomyelia.” Occlusion
hemorrhages from the operative wound. At autopsy the spinal cord was distended with blood from the mid-cervical to the lumbar region and in the upper cervical cord the central canal was wide and surrounded by a glia-like mantle. A central core of similar tissue was found in the lumbar cord. It is possible that this man had an asymptomatic syringomyelia in the absence of trauma into which bleeding occurred because of his hemorrhagic tendency, although this is not entirely certain. A similarly suggestive case was reported by Henneberg and Koch who described a young man with a sudden onset of painful paralysis in both arms with no history of trauma or previous evidence of syringomyelia. The weakness and sensory loss spread to the legs and the patient died in 1 month. At autopsy, an extensive hemorrhage was found in the center of the spinal cord from the low cervical to the mid-thoracic region.

Because there was microscopic evidence of gliosis and cavitation they considered this to be an example of hemorrhage in "latent syringomyelia."

It has not yet been clearly established that surgical decompression and drainage of syringomyelic cavities is of prolonged benefit in patients with this disease. However it would seem that surgical therapy with drainage of the cavity has a definite and urgent place in those patients with syringomyelia who have an acute increase in their symptoms or signs that might indicate the presence of recent bleeding with an increase in tension within the cyst, as in our patient. It is of course to be expected, that only those symptoms due to increased pressure within the syrinx as a result of the bleeding would be favourably affected by surgical drainage, and that those deficits caused by actual tissue destruction would remain.

Summary

We have reported a case of a patient who had symptoms and signs of syringomyelia as well as myelographic demonstration of the syrinx six years before an acute exacerbation of his illness was precipitated by trauma. Operation proved this to be due to hemorrhage into a syringomyelic cavity in the cervical spinal cord. Decompression and aspiration were followed by considerable improvement.

References

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