THE SIGNIFICANCE OF TEMPORARY AND ALTERNATING
PTOSIS, MIOSIS AND ANHIIDROSIS*

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When the spinal cord of man is injured, a wide variety of complex
patterns of activity may develop. Those such as the reflex actions of
the somatic musculature,\textsuperscript{4,9} have been the subject of many recent
writings. In those patients with injury above the fourth thoracic spinal seg-
ment, hypertension and peculiar sudomotor reactions\textsuperscript{7} result from reflex
stimulation of the afferent limbs of the sympathetic system—most potently
induced through the segments supplied by the sacral nerves.\textsuperscript{8} Occasionally
such reflexes may be of such a disabling nature that they require surgical
elimination.\textsuperscript{2,10} In the same general realm of interest are those sympathetic
reflexes concerned with the face and eyes. A careful study has been done in
regard to the ciliospinal reflex in patients with injury of the cervical spinal
cord,\textsuperscript{1} indicating that the afferent pathway is changed after injury. From all
indications, the cervical sympathetic impulses operate independently of the
abnormal reflex responses of the trunk and limbs.

Perhaps the most intriguing finding made in an extensive experience with
patients who have had injury to the cervical spinal cord has been a condi-
tion of ptosis, miosis, and anhidrosis which has been unilateral and tem-
porary and which could be varied by altering the position of the patient.
This report deals with 6 such patients in whom explanations for the changes
appear to have been found.

ILLUSTRATIVE CASES

Case 1. #159001. C.W.H., a 15-year-old boy, was injured in an automobile acci-
dent on June 16, 1951. He believes that he was able to get up to one knee, then
lapsed into unconsciousness. Two days later he was moving all extremities volun-
tarily. Two days after this, a fracture-dislocation of C5 on C6 was manipulated, after
which he was placed in traction. He did not move any extremity after this procedure.
In 3 weeks he was transferred to another hospital where a laminectomy was per-
formed.

On July 23, 1951, he was admitted to the James Whitcomb Riley Hospital with
eleven decubiti, a hemoglobin of 8.5 gm. per cent, blood pressure of 100/60, and an
absolute sensory and motor level at C5. He also had a urethroscrotal fistula and a
severe paraphimosis.

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584
After much supportive therapy and routine investigation, a laminectomy was carried out on Aug. 25, 1951. The spinal cord was found to be compressed laterally by bone fragments. There was a strip of yellowish tissue lying under the arachnoid for a distance of approximately 4 cm. along the right side. This was removed, apparently representing cord material that had extruded through a pial tear at the time of injury. The rent in the pia mater was found at the C6 level where a cavity was present in the cord. This tissue revealed a microscopic picture of pale-staining glial nuclei and nerve cell bodies lying in a fibrillar and loosely arranged stroma, with no evidence of inflammation. The cavity was irrigated and was seen to extend distally.

Following the operation he did very well and in the subsequent months multiple skin procedures were done for the purpose of eradicating the decubiti. After developing an automatic bladder, he was discharged in November 1951.

He was readmitted on Oct. 10, 1952, for repair of a newly formed coccygeal decubitus. At this time, his sensory level was incomplete at C6, with some scattered areas below, and he had active motor elements in the trunk. Of particular interest was the observation that when the patient was in the left lateral position, the right side of the face and neck showed obvious perspiration whereas the left side was dry. On closer examination it was also seen that the left pupil was distinctly smaller than the right, and that the left palpebral fissure was narrower than the right. When he was turned to the right lateral position, the situation was reversed. That is, the right side of the face was now dry and the left showed perspiration, the right pupil was smaller than the left and the right palpebral fissure was narrower than the left. When lying on his back, these differences were not in evidence.
Using the Quinizarin test of Guttmann (Fig. 1), these differences are clearly seen. The time for development of these changes varies from patient to patient and is usually measured in minutes. It is also essential that there be a demand for sweating, although this demand need not represent a great one. Subsequently this patient has been followed further. The sign was still in evidence at the last examination.

Case 2. #126671. D.B., a 26-year-old housewife, incurred a broken neck in an automobile accident on Mar. 4, 1948. She was treated for 8 weeks with traction and then was discharged on a Stryker frame.

She was admitted to the Robert Long Hospital on Nov. 15, 1948. There was a large sacral decubital ulcer and a complete motor and sensory loss below C5. Of particular interest was the finding that there were ptosis, anhidrosis, and miosis on the left side when she was placed in the left lateral position, no such changes when she was placed on her back, and the same changes on the right when she was placed in the right lateral position. Upon questioning, the patient stated that this condition had prevailed for several months.

Two days after admission, a myelogram was performed. It showed a large and persistent filling defect at the C4-C5 level. The findings were interpreted as a probable protrusion of intervertebral disc material.

After repairing her blood status, a laminectomy was conducted on Nov. 19, 1948, with removal of the arches of C4, C5, and C6. There were severe and dense adhesions between the spinal cord and the dura mater. These were freed. The C4-C5 disc space was protruberant and much of this material was removed extradurally.
It was noted that although the cord now pulsated, there was a darkened area at the lower end of the exposed field. This area was entered with a 25-gauge needle and about \( \frac{1}{2} \) cc. of viscid fluid was aspirated. The pia mater was sharply sectioned in the midline over this area and a cystic cavity was entered. The cavity was thoroughly irrigated and upon inspection, there appeared to be intact elements of white matter along the sides and bottom with the cavity apparently representing the region previously occupied by the gray matter. A small piece of Gelfoam was inserted in one end of the pial incision in an effort to keep the edges from approximating. The dura mater was closed.

She recovered from this procedure readily and in 2 weeks the sacral decubital ulcer was repaired. On numerous occasions, efforts were made to reproduce the alternating signs to no avail. She was discharged on Dec. 23, 1948.

She was followed at intervals. There was a gradual lowering of the sensory level to T1 by May 1949, with triceps function bilaterally and good radial extension. At this time it was noted that the alternating signs had reappeared. The patient despaired of life in a handicapped condition and refused to take nourishment. She died 67 days later.

At autopsy, the spinal cord was found to be lightly adherent to the dura mater. In the region of previously evacuated cyst there was a bluish discoloration, with the contour of the cord being rather constant as though it were intact. During the dissection of the cord from the canal, this zone collapsed. It could be distended by irrigation, revealing again the cystic cavity. The area collapsed upon suspension in the fixative. On microscopic examination (Fig. 2), the cord is seen to be occupied by a cavity that is lined by a layer of cells.

DISCUSSION

Four additional patients have been observed to have the alternating signs. In each of the 6 cases there were the common findings of dryness of the face, narrowing of the palpebral fissure, and constriction of the pupil on the side that was down. These were observed usually after a short period of time in the lateral position. In general, there was no detectable evidence of anything other than sympathetic balance with the patient in the supine position, although several of the patients could produce the changes by sharply turning the head and neck. When demands were made upon the body as a whole for heat regulation, the face and neck commonly were the main sources of sweating. Moreover, each of these patients had suffered similar injuries in that none of them was anatomically complete. Upon operation (in 4) it was found that the spinal cord bulged at or below the point of maximum insult. It was quite apparent that this bulging was caused by material contained within the pia mater in the central part of the spinal cord. Upon entering the bulge, a cavity containing viscid fluid was encountered. In 1 patient, the spinal cord was not bulged but the central part did contain necrotic material and there were marked adhesions between the cord and the dura mater. In Case 1 there were no dural adhesions and the cavity may not have been a closed one. The signs have been abolished in all patients after operations, returning later in 2.

Aside from the intriguing nature of the findings, a number of considera-
tions have prompted this presentation. First of these is the predictability of the lesions in each case. Ordinary neurological techniques allow for accurate localization, but usually fail to give dynamic illustration of the pathological situation. These patients had in common a collection of fluid or necrotic material within the spinal cord above the cells of origin of the primary axones of the sympathetic nervous supply to the face and neck. This fluid had resulted in each instance from trauma imposed upon the previously normal spinal cord. In the one specimen that came to microscopic study, a cellular lining was seen to be present. The cystic material appears to have been residue of the degenerated gray matter of the spinal cord with possible additions through mechanisms believed to be operating in the development of other types of fluid collections. There has been intact spinal cord tissue around the cavity in each case. It is apparent from the cystic nature of the cavity that relief would necessarily be surgical in order to be permanent. But more than the simple question of abolishing a neurological curiosity would be the possible effect that evacuation of the fluid might have on the white matter in the region. Since this is the region of maximal insult, and cells in the region can be shown to be responsive, it probably follows that the less susceptible tracts could be expected to function after removal of the noxious influence. Even if the fluid under consideration were to have a specific gravity as great as that of whole blood, its total weight would still be very small. No measurements have been made of the material but its viscosity is certainly less than that of blood. This makes it all the more remarkable that actual physiological changes occur as it is shifted to bring its weight to bear on different portions of the cord. In our laboratories, we have demonstrated that minor blows to the spinal cord of animals usually result in cyst formation such as that observed here. We have not yet seen a comparable group of findings in any other disorder such as suspected hematomyelia from minor injury, syringomyelia, or degenerative disorders.

In attempting to find alternate explanations of the phenomenon, one thinks of some reflex mechanisms which seem to be peculiar to the more or less isolated sympathetic system seen in many patients with injury of the cervical cord. Vascular and sudomotor effects are usually seen in these cases following manipulations of bladder, bowel, or sphincter. These responses may be of a startling nature, but they are clearly reflex and do not depend upon position. It might be that traction could be invoked to explain the findings. The principal argument against this explanation is that the majority of these patients did not have sufficient adhesions to give traction. Also, the more severely injured patients who have dense adhesions between the spinal cord and the dura mater do not show the signs. However, this explanation must be entertained in one case. To date we have not seen this phenomenon in patients with lesions that did not involve the arms, the levels having originally been from C4 to C7 by regular examination. It is probable that the lower extent of each lesion was at least at D1 since the paralysis in the arms and hands was flaccid in nature.

That the abnormal elements lying within the spinal cord are responsible
for the findings is borne out clearly by the patient in Case 2 in whom the
signs disappeared after evacuation of the cyst and returned when the cyst
refilled. The somatic and other sympathetic reflexes were not abolished by
operation and indeed the long tracts had begun to show function, eliminat-
ing any consideration of so-called "spinal shock."

There is another point of interest that these cases bring up. Do similar
fluid collections (or masses) in other parts of the nervous system exert
measurable influences? It is entirely possible that masses with greater
weight, even if only by infinitesimal amounts than the surrounding tissue,
may produce changes only in certain positions and then only when we are
prepared to detect these changes. One is instantly led to wonder about
possible detection of such changes by the electroencephalogram, the pneumo-
encephalogram, or even plain everyday neurological examinations conducted
in unusual positions.

Our original intention after seeing the first case of this phenomenon was
to await an opportunity to present the findings and then defy anyone to
ever find another. It was felt that here at last was an example of neurological
astuteness at its zenith—except that the original observation wasn't made
by us but by the patient. We intended to title the presentation "The Syn-
drome of Temporary and Alternating Ptosis, Miosis, and Anhidrosis" and
thus come into possession of the only syndrome known which existed in only
one patient and thus would never have to be learned by anyone. We hoped
to attempt thereby to contribute in a small way to the discard of the many
so-called convenient terms used to describe combinations of ailments, terms
which are usually unpronounceable and next to impossible to remember from
case to case. This would result in greater ease in teaching neurology to medi-
cal students, most of whom acquire a wholesale distaste to the discipline
because of the many fancy words used. Since we have seen more cases, we
are without the forceful case that we had, but we still have wholesale aver-
sions to the use of proper names for disorders.

In patients subjected to early laminectomy and pial incision, this
phenomenon has not been observed, presumably because, as in the experi-
mental material, the cysts do not form once the necrotic cord tissue and
blood are evacuated. In a sense, this would constitute an additional bit of
evidence in favor of early operation.

**SUMMARY AND CONCLUSIONS**

1. Temporary and alternating ptosis, miosis, and anhidrosis occur when
patients with cystic cavitation of the lower cervical spinal cord as a result of
trauma are placed in the lateral positions. Upon evacuation of the cyst, the
signs disappear.

2. It is postulated that the signs constitute evidence of an incomplete
lesion which will benefit from surgery.

3. The question is raised as to whether it is possible to detect other lesions
of the central nervous system by examination of patients in other than
routine positions.
REFERENCES


