Syringobulbia

Case Report with 19-Year Follow-Up

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Syringobulbia refers to a cavity within the medulla and/or pons and may be an independent entity or the rostral extension of a syringomyelic state. The cavity distorts surrounding structures and generally has its largest diameter in the longitudinal axis.

Syringomyelia and syringobulbia usually are considered as neurological entities for which surgery sometimes is of excellent temporizing value. This case, however, is presented more as an anatomical curiosity in which long-term postoperative survival resulted in spite of a complete dysraphia of the medulla in association with the syrinx.

Case Report

A.H., the 54-year-old widow of a deceased army officer, entered Walter Reed General Hospital on Jan. 27, 1962 complaining of bilateral pain and weakness of the upper extremity, weakness of the neck and generalized moderate loss of body control. The original facets of this woman’s problem began many years prior to her admission to hospital in 1948, as reported previously. At that time a syrinx of the bulb was diagnosed and a surgical decompression was accomplished. The patient was discharged from the hospital after 2 months and returned to gainful self-employment after 9 months.

She remained well for fully 10 years, following which she noted that she would drag her left leg and experienced weakness of both hands. She was hospitalized again, evaluated and reoperated upon by Dr. John Martin. In his operative report he stated the following: “... When the dura mater was opened we found that what probably had been the former obex was now down at the level of the posterior arch of the C2 vertebra. The spinal cord was opened out into two lateral shell-like halves, which continued on up to the medulla which was in the same general condition and very badly adherent everywhere to the cerebellum. In this wide cleft, which appeared to be an extension downward of the open fourth ventricle, the space was filled with a semitransparent, tough, jelly-like substance which, no doubt, is the remnant of the degenerated cord and brain stem. When this was aspirated, we got clear urine-colored fluid at a depth of about 8 mm. It was then opened along what had been the posterior median raphe with fine tissue forceps, and we came upon a tremendous cavity which filled the large, hollowed-out upper cervical cord and extended up into the medulla which was, as the cord, only a shell of viable neural tissue. Out of this we sucked any quantity of yellow fluid which coagulated on contact with the air. We left a large ostium in the gelatinous material leading into this cavity at the level of the atlanto-occipital membrane and slightly below...”

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Again, convalescence was long, but the patient felt that she had regained her preoperative level within 1 year. Since then she has remained well and self-supporting until 2 months prior to this admission. The pain and weakness described in the chief complaint were of insidious onset and attempts at physiotherapy were carried out; however, no lasting effects were achieved and the patient was rehospitalized a third time for her disease.

Examination. Pertinent abnormalities were neurological. Testing of cranial nerves revealed subjective diplopia on lateral gaze bilaterally although gross defect was not seen. Horizontal and vertical nystagmus was present on lateral gaze and on visual fixation respectively. Hypalgesia over the maxillary and mandibular divisions of the right trigeminal distribution was noted. Gag reflex was decreased and swallowing ability was considerably impaired. Motor system revealed fair strength throughout with the only weakness present being manifested distally in the extremities. The left side was slightly more involved than the right. Unstained knee and ankle clonus was present bilaterally. Tendon reflexes were hyperactive in the upper extremities. Bilateral Hoffmann, Babinski and Chaddock responses were elicited. Abdominal reflexes were absent (exogenous obesity?). Sensory testing revealed the patient to be intact throughout to all modalities excepting the right Vth nerve, as mentioned above. Gait could not be tested properly since the patient felt too weak generally to walk without assistance. However, her gait appeared to be of a shuffling, wide-based variety.

In view of a previously well established diagnosis and relatively rapid deterioration within the past 2 months, re-exploration was recommended and accepted by the patient.

Operation. On Feb. 2, 1962, under general endotracheal anesthesia, the suboccipital cranieotomy was enlarged and a total laminectomy of C1 and C2 was carried out through a midline incision. After opening the dura mater dense arachnoidal adhesions were divided. The cerebellum appeared normal as did the exposed cervical cord from C1 downward. Upon retracting the cerebellar tonsils upward there was seen to be a multiloculated semicystic-semisolid mass occupying the region of the fourth ventricle which extended into the cerebellar vermis above and the medulla oblongata below. Upon dividing the mass in the midline and allowing most of the contained thin, clear, amber fluid to escape, the solid components were then removed by tedious dissection from the adjacent neural structures. There remained thereafter a large defect extending through the entire thickness of the medulla with only the most lateral aspects of this structure remaining. The clivus was readily apparent and the vertebral-basilar arterial system was visualized (Fig. 1). No alterations of vital signs were manifested throughout the procedure. Routine closure was accomplished.

Postoperative Course. Recovery from anesthesia was
Antibiotic therapy was begun and cultures subsequently grew out a pneumococcal organism sensitive to penicillin, the therapeutic agent already selected. Oxygen tent and moist air were also added to the therapeutic regimen. On February 8, the patient requested and was provided with a bedpan. At approximately 9:00 p.m., when the bedpan was to be reclaimed, the patient was found dead in bed. Postmortem examination was not permitted.

Discussion

Many students of neurology and neuropathology feel that the pathologic process as it is known results in all probability from a congenital heterotopic inclusion of cells at the time of closure of the dorsal raphe. It is felt that this theory is strengthened by the oft-noted multiple congenital anomalies in such patients. These hamartomatous masses may then be considered as tumors, but unlike ordinary gliomas they do not infiltrate extensively. They do, however, manifest a definite and peculiar proliferative process with ultimate formation of cavities. Destruction of adjacent neural tissue usually is related to compression rather than to infiltration. Rosenthal fibers are encountered frequently, thereby removing this as a pathognomonic feature of astrocytomas in the usual sense; but, by the same analogy, mitigating for the placement of the associated glial lesions of syringomyelia and -bulbia into a tumor classification. Microscopic sections of tissue removed in this case support this contention (Fig. 2).

Our case represents a 19-year follow-up of what

Fig. 1. Operative sketch made after removal of semicystic-semisolid hamartomatous tumor.

Fig. 2. Photomicrograph of surgical specimen consisting predominantly of well differentiated protoplasmic astrocytes. X110.