LIPOMA OF THE SPINAL CORD ASSOCIATED WITH KLIPPEL-FEIL SYNDROME

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Central nervous system manifestations associated with the Klippel-Feil syndrome are not uncommon, and are probably caused by a congenital myelodysplasia. This naturally raises the question as to whether in some cases the Klippel-Feil syndrome may be associated with an intramedullary tumor, particularly since the central nervous system symptomatology resembles that of syringomyelia in many instances. It is also well recognized that the symptomatology of syringomyelia and intramedullary tumor may be indistinguishable. An intramedullary tumor (ependymoblastoma) was found at autopsy in a patient of Turnbull's, but his case was not one of Klippel-Feil deformity. Du Toit considered the probability of an intramedullary tumor in his patient with Sprengel's deformity. The possibility of an intramedullary tumor was also mentioned in the case reported by Avery and Rentfro. In Nery's case the Klippel-Feil syndrome was most likely associated with an intramedullary tumor. A myelogram disclosed a block at the level of the 7th thoracic vertebra. His patient died rather suddenly from respiratory embarrassment. Unfortunately, a postmortem examination was not obtained. Proven intramedullary tumors, however, have not been reported heretofore, as far as the author is aware, and the following case is therefore presented.

CASE REPORT

A 48-year-old female, referred by Dr. Harry Subin of Atlantic City, N.J., was admitted to Temple University Hospital on Mar. 11, 1949. She had had difficulty in walking and in raising both arms as long as she could remember. The right extremities were more involved than the left. She could never raise her hands above the level of her shoulders but was able to comb her hair with the left hand. During the last year she had experienced progressive difficulty in walking and she complained of pain in both forearms, especially the right. She described a "numb feeling" in both forearms. The numbness appeared to be greater on the left side and prevented her from being able to mend her clothes. She denied any bowel or bladder difficulty.

Examination. She had large, heavy-set features. Her neck was shortened, and the hair line extended to the mid-cervical area. Below the hair line there was a huge pad of fat which extended from C7 to T4 and gave the appearance of a gibbous deformity (Fig. 1). The gait was spastic so that she barely shuffled along. The right lower extremity was more spastic than the left and it was more difficult to raise the right

Fig. 1. Photograph of patient showing low hair line, short neck and the pad of fat posteriorly.
limb. Spasticity involved all four extremities but the lower limbs were more severely affected. The left arm could be raised to the level of the head but the right arm could be elevated only as high as the shoulder. Deep tendon reflexes were hyperactive bilaterally. A Hoffmann sign was present on both sides. Abdominal reflexes were not elicited. The Babinski sign could not be demonstrated on either side. Sensation to pain and temperature was decreased bilaterally from C3 to T5. Touch sensation was decreased over the same areas but particularly over the 4th thoracic dermatome. Position and vibration sensations were decreased in both upper limbs but more so in both lower limbs. There was marked dyskinesia in the right upper limb and dysmetria in both upper limbs.

Spinal puncture revealed a complete manometric block on Queckenstedt maneuver. The only spinal fluid abnormality was a protein of 82 mg. per cent. Roentgenograms showed an anomalous fusion with a deformity in the cervical area. The 3rd, 4th and 5th cervical segments were fused together, and the 6th and 7th segments were likewise fused. Apparently seven cervical segments were present. In addition to the fusion, the involved vertebral bodies were relatively narrower in their anteroposterior dimension. The vertebral canal was quite large. The first two cervical segments appeared normal (Fig. 2). Pantopaque myelography (Fig. 3) was carried out to delineate the extent of the lesion, if possible, and to perhaps determine whether it was intra- or extramedullary. The pantopaque injected into the lumbar region was brought to the cervical region without difficulty. An unusually large vertebral canal was demonstrated. The morphology at the level of the foramen magnum was not well delineated and some of the pantopaque entered the skull. The caliber of the cervical vertebral canal was well shown; the canal measured 3½ cm. in transverse dimension and 3 cm. in the anteroposterior dimension. In the supine position it was not possible to bring the pantopaque any higher.