# NEUROFIBROMA OF THE HYPOGLOSSAL NERVE

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NEUROFIBROMATOSIS may be found scattered over many different parts of the nervous system. Multiple tumors often in great number are found in the meninges as well as the peripheral nerves, while single tumors are more common in the deeper nerve trunks as well as the cranial nerves. Of the cranials, the acoustic is the most frequent site, while cases have been reported of the vagus, of the cervical sympathetic, and spinal accessory nerves. In the visceral form, these growths have been reported as occurring in the tongue, stomach, jejunum, ileum, colon, vagina and bladder. (Walter E. Dandy, p. 667, Lewis Practice of Surgery, vol. 12.)

The following is a report of a case of neurofibroma of the right hypoglossal nerve. So far as I know, this is the first case on record. It is quite interesting to note that while the growth was of considerable size, occupying more than two-thirds of the proximal portion of the nerve as it emerges from the cranium, it was absolutely symptomless so far as motor function disturbance. Evidently, the few fibres that were left were sufficient to carry on motor function. Soon after resection, however, motor function on that side was entirely interfered with, and atrophy of the right side of the tongue was soon evidenced.

CASE REPORT.—M. S., female, aged thirty. For the past one and one-half years, a slowly growing, painless swelling was noticed on the right side of the neck, at the angle of the jaw. Conscious of some difficulty in swallowing, no interference in mastication; did not notice at any time difference in the thickness or size of the right side of the tongue. At no time any motor disturbance of tongue, or any deviation of tongue when protruded. No interference with speech. Examination showed considerable enlargement of right side of the neck (Fig. 1), caused by a mass occupying entire superior carotid triangle, smooth and globular, the lower boundary reaching to about the middle of the sternomastoid. The mass is felt beneath the sternomastoid, cystic in character, painless and sufficiently large to cause definite bulging of the pharynx and tonsil toward the median line. Pressure on the outside of the mass with the mouth open shows very decidedly the dislocated tonsil and pharynx toward the mid-line. Diagnosis was not made; the probable diagnosis being a large branchial cyst or aberrant thyroid.

*Operation.*—Avertin anæsthesia, incision from the angle of the jaw parallel with the edge of the sternomastoid, four and a half inches long. Sternomastoid retracted, incision continued down to tumor. The entire tumor easily separated from the surrounding structures and shelled out of its bed, after carefully separating it from the carotid sheath, its upper limit tapering far up into the neck to about the level of the transverse process of the second cervical vertebra, which was distinctly felt. The sheath of the nerve at its extreme upper limit was thinned out and here tied and cut, the entire mass delivered,

and, upon doing so, the rounded lower portion of the mass was found to be continuous with a cord-like structure which was identified as the hypoglossal nerve. A portion of the normal nerve was removed with the tumor. The tumor occupied that portion of the nerve nearest to its exit from the skull to about where the nerve normally crosses the tendon of the digastric muscle. At this point the nerve became abruptly normal, that is, at its junction with the globular-shaped, distended part of the tumor. During the removal of the tumor, the cystic portion ruptured, a clear fluid escaping; the rest of the mass consisted of a semi-soft gelatinous-like structure. The operation lasted thirty-five minutes, the patient having an uneventful recovery.

Two days after operation, there was pronounced evidence of atrophy of the right

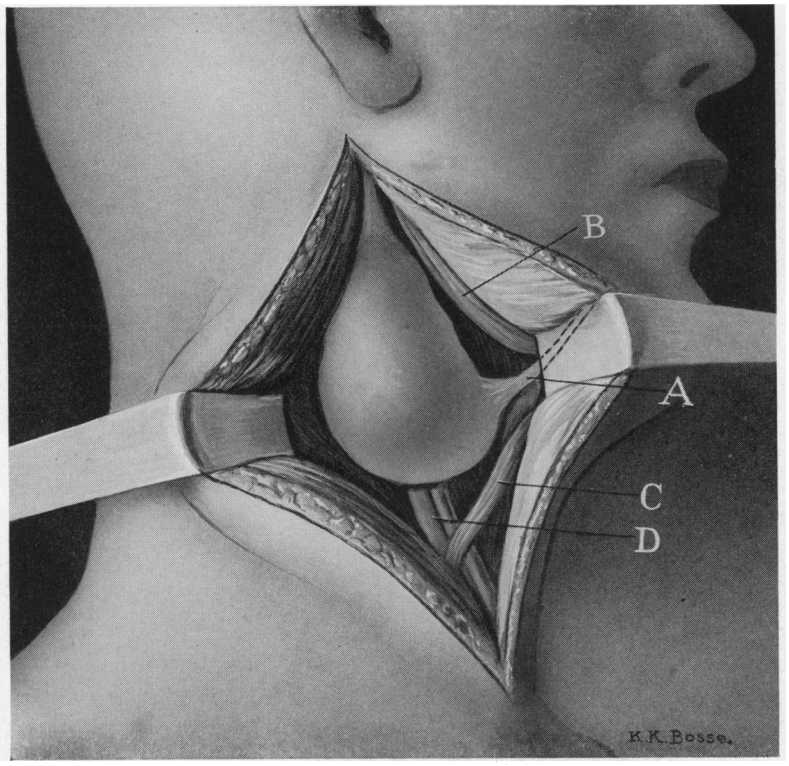


FIG. 1.—Showing neurofibroma of the hypoglossal nerve. (A) Normal part of the hypoglossal nerve. (B) Posterior belly of the digastric. (C) Omohyoid. (D) Deep vessels of the neck.

side of the tongue. (Fig. 2.) The patient complained of some difficulty in managing food on the right side of the buccal cavity. There was some interference with speech for a few days; sensation was normal. When the tongue was protruded, there was deviation toward the right (affected side) while the tip curves to the left side. The raphé formed a distinct curve, the concavity being on the paralyzed side. No deviation was present when the tongue was not protruded. After a month's time speech was normal and complete management of food was possible. Atrophy very decided.

*Pathological Report.—Anatomical.*—Specimen is a piece of tissue measuring five by three centimetres, one part of which is a cyst with a thin wall containing thin hæmorrhagic fluid. The upper part appears to be solid, has a yellowish fatty color and has the consistency of mucinous or gelatinous substance, but on section it has the appearance of firm fibrous tissue.

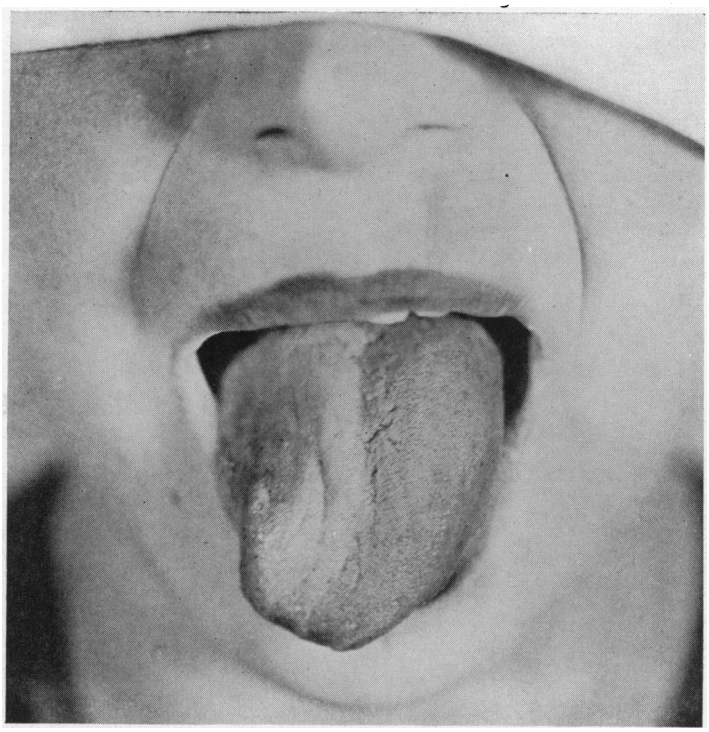

FIG. 2.—Showing atrophy and deviation of right half of tongue following resection of right hypoglossal nerve for neurofibroma.

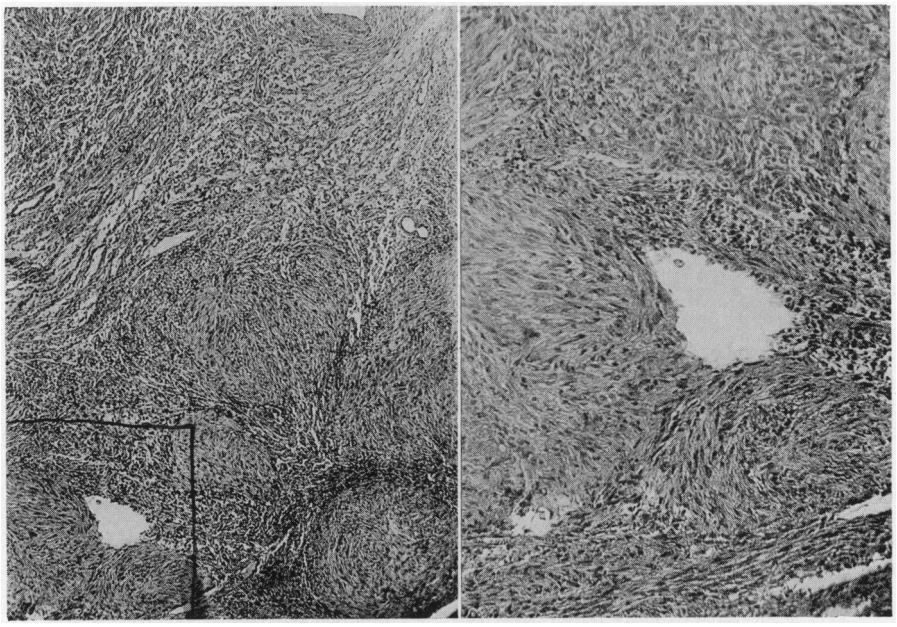

FIG. 3.—Low magnification of the tumor showing its general appearance.

FIG. 4.—Higher magnification (x145) of the area marked off in Fig. 3. The characteristic structure is well shown, including numerous fibrils and nuclei parallel to each other ("nuclei on parade").

*Microscopical.*—Sections show a very cellular structure with areas of œdema and several hæmorrhagic foci towards the periphery of the structure. Throughout the entire section there are seen areas of fatty degeneration. The tumor proper is composed of closely packed fibrils which in some places are parallel and in others perpendicular to each other. In places these are arranged in the form of palisades. Again there are several areas in which these cells are arranged around the small bundles of nerve fibres and here the tumor has an alveolated appearance.

*Some Pathological Features of Neurofibromata.*—Much water has flowed under the bridges since the Old Master of Pathology, Virchow, spoke of "true" and "false" neuromas, designating the tumors composed of nerve structures proper—axis, cylinder and myelin sheath—as true neuromata, and those composed of connective tissue as the false. While the division of neuromata into "true" and "false" may not have led to so much confusion and nomenclatural chaos and welter as was created by leukemias and pseudo-leukemias with their crowning achievement of "true pseudo-leukemia"—true false leukemia—nevertheless an attempt can be made to separate the two neuromata on the basis of their histogenesis, and a very brief review of histological data may not be out of place.

In the structure of a nerve fibre, we have the following elements: (1) The ordinary loose connective tissue: the epineurium which envelops the entire nerve, the perineurium which surrounds each bundle of nerve fibres, and the endoneurium which ensheathes individual nerve fibres. (2) The nerve tissue proper as found in the axis cylinders and myelin sheaths. (3) The neuroglia—the supporting nerve tissue—the cells of the sheath of Schwann.

It is obvious that the tumors derived from (1) are genuine neuro-fibromata, *i.e.,* fibromata of the nerve, just as we have fibromata of the uterus, *etc.* It is equally obvious that the tumors composed of the nerve tissue proper from (2) are neuromata. But what of the tumors composed of neuroglia which is also, embryologically, a nerve tissue, *i.e.,* is derived from the ectoderm (with the exception of the mesoglia of Hortega which is a mesodermal tissue)? Naturally, they must be regarded as neuromata, and, as a matter of fact, in the central nervous system, they are the well-known group of gliomata. It is becoming quite clearly established that pure fibromas, just as pure neuromas and pure gliomas of nerve fibres are very rare and that most of these tumors reported are composed of both connective tissue and of the cells of the sheath of Schwann, and it would be, therefore, desirable to give a new name to these combined tumors, since it is now definitely accepted that the cells of the sheath of Schwann are neuroglia cells.

Such a name has been coined by Verocay.* In discussing these tumors under the name of "neurinoma," Verocay has pointed out that practically all such tumors are derived from the cells of the sheath Schwann (Schwannoma) and are, therefore, ectodermal, not mesodermal, tumors

* Verocay: Ziegler's Beitr., vol. 48, p. 1, 1910.

(even though the neighboring connective tissue participates, to a very limited degree, in the growth). He has further pointed out that these tumors, when stained by the Von Gieson method, do not stain red, but are yellow, and are composed of netlike bundles of fibres (plexiform), with the nuclei frequently in parallel rows ("nuclei on parade," Askanazy). It must be added, however, that several authentic cases of pure fibromata of the nerves (Harbitz, Wegelin, Kaufmann, Ribbert, Antoni) have been reported. The von Recklinghausen disease does not present a constant picture since some cases are pure neuromata while others are pure fibromata.

It is of importance to bear in mind that while a great majority of neurinomas are benign, they may assume sarcomatous properties and that many of the so-called "neurogenic sarcomata" probably belong to what is still described as "neurofibromata." Occasionally, as in our case, the tumor may be cystic.