

## NEUROGENIC TUMORS OF THE NOSE AND POSTNASAL SPACE

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### ABSTRACT

Non-epithelial, benign extracranial neurogenic tumors of the nasal cavities are reported in the literature with extreme rarity.

These tumors differ from the more common congenital gliomas, encephaloceles, etc. in that they sometimes are detached from the brain tissues. These tumors have intracranial origins. They are congenital or acquired and may involve any of the nerves inside the nose. These tumors can grow to quite a large size and in such cases, lateral rhinotomy is often necessary for complete excision.

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### INTRODUCTION

Intranasal benign tumors of different histological types are not uncommon. They are either epithelial in origin (adenomas and papillomas), or non-epithelial (fibromas, hemangiomas, nasal glioma, neurilemmoma, chondroma, and osteoma). Odontogenic and fibro-osseous tumors are also rarely encountered.

This paper reports two cases of intranasal schwannoma (who had the preoperative histologic diagnosis upon biopsies), their operative procedures and findings and also the postoperative results after several months. Then, in a short discussion, the interesting points mentioned in the literature about these rare tumors are reviewed.

#### CASE 1

A 3 year old boy was referred to the outpatient clinic with a large right intranasal mass that had occluded the right nostril and caused severe deviation of the nasal contour. He had had a fleshy mass of 2 cm diameter in his right nostril since birth. The tumor did not cause any problem so for the first time, he was operated on by a plastic surgeon at the age of 2 years. The mass apparently was incompletely removed (through an external alar incision). The pathologic report was "schwannoma". The mass had rapidly regrown to a larger size.

On physical examination the patient was healthy and

cooperative. A huge almost white, fixed (to the septum), semi-firm, pedunculated mass (judged from the CT films) filled the whole right nostril.

The tumor was dissected free and removed totally through a lateral rhinotomy incision. The stalk extended proximally to the deep ethmoid cells, but the mass itself was round to ovoid and measured about 4-5 centimeters in diameter.

The patient showed no evidence of recurrence several months after operation.

#### CASE 2

A 36 year old female presented with a 6 month history of vague pain over and behind her left mid-face, gradual proptosis, loss of her left eyesight and repeated colds.

In the past history the patient had undergone a Caldwell-Luc operation for what she was told to be left chronic maxillary sinusitis. The pathology report of the intrasinus debris surprisingly was schwannoma of unknown origin. She had a normal eye at that time.

Clinical examination revealed an obese young lady with a slightly protruded left eye which nevertheless had good (and synchronous with the right eye) movements. No other abnormality was present on physical examination. She had consulted with an ophthalmologist who reported no light perception in the left eye.

Plain radiographs were non-revealing and the left maxillary sinus was even moderately aerated. The CT scan

films showed a large soft tissue mass at the post-antral space on the left side. The tumor had caused bowing of the posterior antral wall, occupied a large space and extended to the left subtemporal fossa. Posteriorly the mass had entered the left orbital cavity and involved the optic nerve. However the tumor had not encroached upon the intracranial brain tissues. She was warned about the dangers of tumor extension and was recommended to have a radical operation through a lateral rhinotomy incision.

The operation was performed on April 20th, 1993. Through a Weber-Ferguson incision and following removal of the posterior antral wall (and portion of the zygomatic bone), a large, dark red, very firm and fixed, non-pedunculated mass was removed from a roomy post-antral and pterygomaxillary space. Bleeding was profuse and the tumor had extended further backwards into the orbital cavity through a small gap in the posterior orbital floor. Here, too, the tumor was dissected free from the optic nerve and the extrinsic ocular muscles. The gap was obliterated by a small silastic sheath.

Postoperatively the patient recovered uneventfully and was discharged from the hospital in 10 days. She has retained movement of her left eye-ball and is in excellent condition.

### DISCUSSION

The peripheral nervous system depends on Schwann cells for support of its neural elements (from an anatomical point of view).

Neural tumors originate from neurons or the sheath cells. Nerve sheath tumors, by far the largest group of peripheral nerve neoplasms, are considered here. A full discussion of their historical and theoretical aspects would be out of place here; for further details the reader is referred to specialized oncologic monographs or related chapters in textbooks.

The nerve sheaths consist of specialized cells of neuroectodermal origin, which include Schwann cells and capsule cells of the peripheral ganglia. The origin of peripheral cells remains controversial and these cells are not included here. In addition, the sheaths contain connective tissue elements. All sheath tumors are composed of both types of cells, with one or the other predominating. This leads to the temptation to consider them all as a single entity. Yet the differences, both theoretical and practical, are such that division into subgroups with acknowledgement of transitions and overlaps is necessary. The prototypes of the sheath tumors are the solitary schwannoma on one hand and the plexiform neurofibroma of neurofibromatosis on the other.

The benign solitary nerve sheath tumor has been known by a bewildering variety of names during the two centuries

since its first description by Sandifort in 1777.<sup>9</sup> Most of these are now of purely historical interest, but a number are in common usage. The terms "neuroma" and "neurifibroma" are mentioned only to be condemned, as they lead to confusion with other tumors. The acceptable terms include "neurinoma" "neurilemmoma" and "schwannoma". All of these are open to discussion, but only the last one firmly pinpoints the cell of origin of these tumors.

A normal nerve fiber, besides being ensheathed by Schwann cells, is also surrounded by more loosely distributed endoneural fibroblasts. The long historical controversy, reviewed by Baily and Hermann, and by Saxen,<sup>10</sup> between the adherence to fibroblastic and schwannian theories has been settled in favor of the latter by the advent of electron microscopy. It has been shown beyond a doubt that the tumor cells possess basal laminae (basement membranes) and are therefore not fibroblasts. Masson was among the first to indicate that nerve sheath tumors were likely derived from Schwann cells.<sup>6</sup> Schwannian origin has also been supported by Stout,<sup>11</sup> Murray and Stout,<sup>7</sup> Rio-Hortega, Fisher and Vuzenski<sup>8</sup>

The theory for Schwann cells as the origin of these tumors was later objected to by pathologists who could clearly identify collagen in their histopathologic analysis on these tumors. Electron microscopic studies soon thereafter, showed that Schwann cells could produce collagen, and nerves may be found on histopathology of these tumors together with fibromatous structures.

Schwannomas may arise from cranial and spinal nerve roots or from the peripheral nerves. Of the three compartments, the cranial nerves are undoubtedly the most important.

The vast majority of intracranial schwannomas arise from the eighth nerve. A small group originates from the fifth nerve and isolated examples have been recorded in other lower cranial nerves. In general, schwannomas have a predilection for sensory nerves; of the motor nerves, the facial is the most frequently involved.

Solitary schwannomas of peripheral nerves are relatively rare. They occur along the course of nerve trunks, mainly in the limbs, occasionally in the head and neck and rarely on the trunk. These peripheral nerve tumors may grow to large sizes. Degenerative changes such as alterations or hemorrhagic necrosis are usually present in these masses (see case 2).

Neurites do not criss-cross the tumor (schwannomas push the axons aside and do not incorporate them).

Macroscopically, this tumor is firm, well circumscribed, solitary, encapsulated, round or ovoid and almost always benign. It may be painful (the pain is often provoked by pressure). A description of the microscopic appearances of this tumor is beyond the scope of our discussion. Therefore, in order to better study the histologic patterns, the readers should consult pathology texts.

Schwannomas of the cranial nerves are predominantly tumors of middle life, although some occur in young adults. Growing slowly, they escape detection for a number of years, and the average age at diagnosis is therefore considerably greater than the age of onset (improved diagnostic methods that lead to the discovery of more small tumors have shifted the emphasis to the earlier decades of adult life).

Peripheral tumors show no definite age or sex distribution (compare cases 1 and 2), though this may be due to the relatively small number of recorded cases.

Primary schwannomas of the nasal cavities and paranasal sinuses may arise from intranasal nerves; the ophthalmic and maxillary branches of the trigeminal nerve, and branches of the autonomic nervous system. Combined nasal-ethmoid involvement is most frequent (see case 1). This is followed in order of frequency by the maxillary sinus, intranasal, and the sphenoid sinus. Keys to cure are: first, early diagnosis and second, complete surgical removal.

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