NEURILEMOMAS OF THE LATERAL REGION OF THE NECK

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NEURILEMOMAS of the lateral region of the neck are rare. Five were reported by Stout[1] in 1935 in his series of 52 neurilemomas of peripheral nerves. Since that time only a small number of these tumors has been reported.[2-7] By definition a neurilemoma is a benign neoplasm of the nerve sheath. There has been much controversy about the cellular origin of the tumor, largely because there are two distinct sheaths on peripheral nerves: the sheath of Schwann and the connective tissue sheath. Virchow,[8] in 1863, considered the tumor to be a false neuroma. In 1910 Verocay[9] demonstrated that its origin is in the sheath of Schwann, but applied the inadequate term of *neurinoma*. Mallory[10] and Penfield[11] believed the connective tissue sheath to be the site of origin. Masson[12] supported Verocay's theory, and was able to produce experimentally tumors of the nerve sheath, the origin of which could be traced to Schwann cells. These he therefore termed *schwannomas*. In 1935 Stout[1] introduced the term *neurilemoma*, and in 1940, with Murray,[13] confirmed by tissue culture the origin of these tumors from Schwann cells.

Neurilemomas of the lateral portion of the neck generally present themselves as solitary tumors lying either subcutaneously or deep within the neck. They grow slowly and usually are detected only when they become palpable. Only rarely is pain or tenderness a presenting symptom; such pain results from pressure on adjacent structures and does not originate in the nerve of origin of the tumor. The age and sex of the patient afford no diagnostic help. Neurilemomas vary in diameter from a few millimeters to several centimeters. They may be round, ovoid, or fusiform, and have a smooth or lobulated surface. Grossly, they tend to be firm, discrete, or encapsulated. Often the large tumors are cystic, showing myxomatous degeneration within the lesion. Their color usually is pink, yellowish, or gray, and the cut surface of the tumor is glistening.

Histologically the typical neurilemoma is composed of orderly arranged, long, slender cells with elongated nuclei in a reticulum of long, slender fibers which pass between and parallel to the cells. The cell nuclei tend to lie at the ends of the cells, in palisade formation; this pattern is the so-called *Antoni type A* or *Verocay type*. The *Antoni type B*, or reticular pattern, is characterized by a loosely composed structure with intercellular edema, and vacuoles or microcysts filled with watery fluid. Palisading is absent. Both patterns may be present in the same tumor (Fig. 1). Willis[14] has described a third type of histologic pattern that is essentially epithelioid. The tumor may be grossly cystic (Fig. 2).

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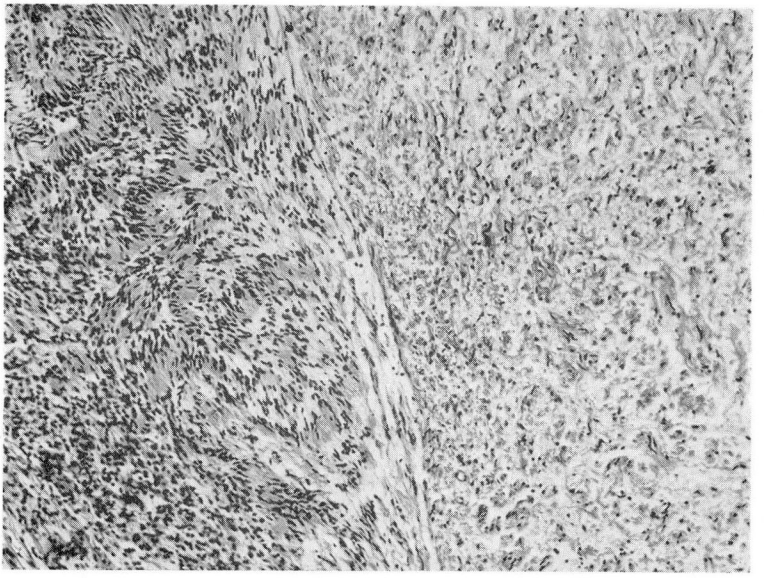

Fig. 1. Microscopic section of a single tumor showing the nuclear palisading of the *Antoni type A* on the left, and the loosely arranged cells without order, *Antoni type B*, on the right; hematoxylin, eosin and methylene blue stain, X85.

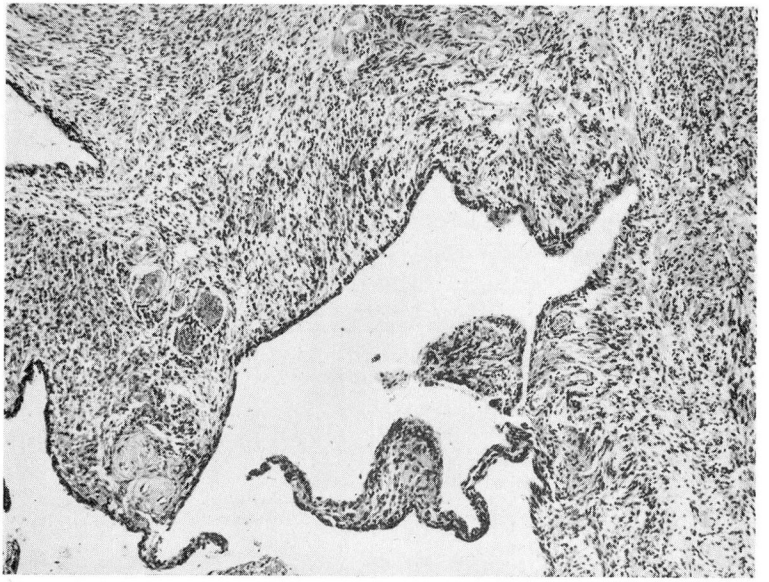

Fig. 2. Cyst formation in neurilemoma; hematoxylin, eosin and methylene blue stain, X100.

Case Reports

Case 1. An 82-year-old man had a large, asymptomatic tumor of the left side of the neck which gradually had increased in size for seven years. On physical examination a firm, rubbery, freely movable mass, 8 cm. in diameter, was noted lying beneath and anterior to the left sternocleidomastoid muscle (Fig. 3). The presumptive clinical diagnosis was branchial cleft cyst.

At operation, the mass was found to be a cystic tumor arising from the vagus nerve 2 cm. below the carotid bifurcation (Fig. 4). A segment of the nerve was removed with the tumor.

Pathologic findings: The lesion was a cystic neurilemoma with a wall 0.5 cm. thick. Microscopic sections showed both Antoni A and B patterns, and cystic areas containing acidophilic granular material.

Case 2. A 38-year-old woman had a presenting complaint of transient pains involving most of the left side of the neck. At the time of onset of the pains, one year prior to examination, she had noticed a mass beneath the angle of the left mandible. This mass subsequently had enlarged slowly. On physical examination a 1.0-cm., slightly tender, freely movable, firm mass was found just below the angle of the left mandible.

At operation, the tumor was excised; no continuity with nerve fibers was noted.

Pathologic findings: The lesion was a neurilemoma, principally Antoni type A with several foci of type B.

Case 3. A 20-year-old woman had a three-year history of generalized peripheral neurofibromatosis. Biopsy of a nodule had shown neurofibroma.

Because of local discomfort, a 1.5-cm. nodule in the right supraclavicular area, diagnosed preoperatively as neurofibroma, was excised.

Pathologic findings: On microscopic examination typical Antoni type B neurilemoma was seen.

Case 4. A 36-year-old woman had the presenting complaint of an asymptomatic lump in the left side of the neck for three weeks. On physical examination, a firm, freely movable tumor, 2 cm. in diameter, was palpated in the left supraclavicular region.

At operation, the tumor was found to be a neurilemoma arising from the brachial plexus. The tumor was removed without damage to the plexus.

Pathologic findings: The tumor was a typical Antoni type A neurilemoma.

Case 5. A 58-year-old woman had had a mixed tumor removed from the left parotid region three years previously. Approximately six months before admission to the Clinic, a mass, presumed to be a recurrent parotid tumor, had been noted deep in the neck beneath the angle of the mandible. On physical examination there was a firm, vaguely outlined, slightly movable, 2-cm. mass deep in the jugulodigastric area.

At operation, a typical neurilemoma was found, arising from an unidentified small nerve close to the pharyngeal wall.

Pathologic findings: Microscopic sections showed neurilemoma, Antoni types A and B, with associated fibrosis and hemorrhage.

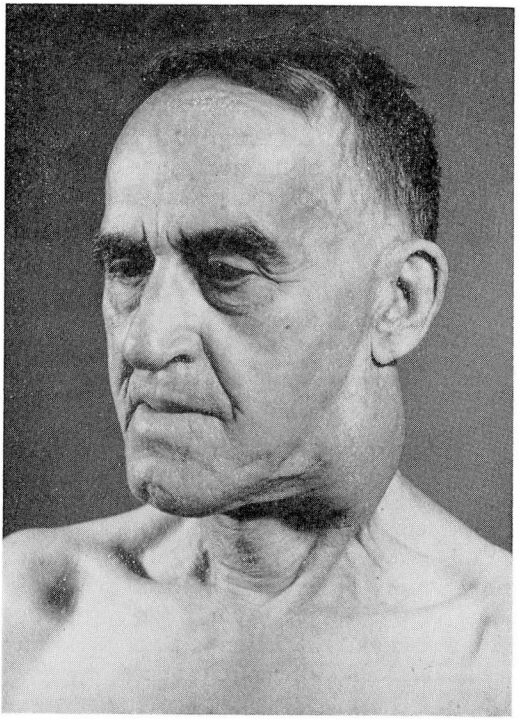

Fig. 3. (Case 1) Patient with large tumor of lateral neck; the mass was freely movable.

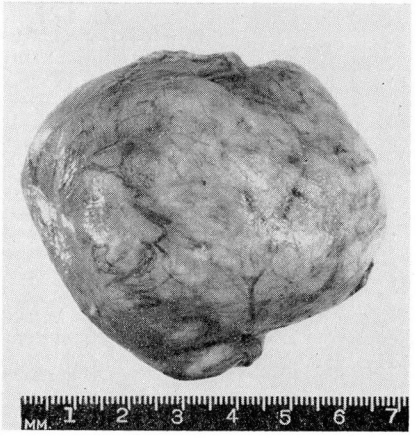

Fig. 4. (Case 1) Ovoid and well-encapsulated tumor removed from patient shown in Figure 1. It contained a single large cyst.

Discussion

In all of the above cases, histologic evidence was necessary to make the definitive diagnosis of neurilemoma. In no instance was it the presumptive clinical diagnosis. At best one can only suspect the lesion preoperatively, since any of the discrete lateral tumors of the neck, including tumors of the carotid body, branchial cysts, neoplasms of the parathyroid, lymphoid tumors, and metastases to cervical nodes are equal possibilities. Although in one of our cases (case 3) neurilemoma occurred with von Recklinghausen's disease, this is a rare association.

In the course of examination of the patient with a mass in the lateral cervical region, it is most important to rule out the presence of a growth in the nose, mouth, or throat, which may be the site of origin of a cervical metastasis.

If, at the time of operation, the tumor appears grossly to be a neurilemoma, a biopsy specimen is taken for definitive diagnosis by frozen section. After the diagnosis of neurilemoma has been established, local excision is carried out, with the plane of dissection close to the capsule. If a major nerve trunk is the site of origin, the tumor is dissected free, and the nerve bundles, generally passing to one side of the tumor or spread out over its surface, are preserved. In most instances there is no serious or prolonged functional deficit of residual nerve. If the origin or location of the tumor precludes its complete removal without sacrifice of the nerve, a segmental resection must be performed. Because of the benign nature of the lesion, results after surgical excision usually are good.

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