Chapter 89: Neoplasms

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Benign Neoplasms

The differential diagnosis of a mass in the neck encompasses a broad range of possibilities, including congenital cysts, inflammatory masses, and both primary and metastatic neoplasms. A clear understanding of the normal anatomy of the neck and a high index of suspicion are fundamental to recognizing these pathologic processes and then progressing logically to establish a diagnosis. Under most circumstances the otolaryngologist-head and neck surgeon familiar with the head and neck can make an appropriate differential diagnosis quickly based on the history and physical examination. Adjunctive tests such as computed tomography (CT) scan and angiography have removed much of the uncertainty associated with the diagnosis.

The most common benign masses found in the neck are lymphadenopathy and pathologic conditions involving the salivary and thyroid glands. Benign tumors in the neck are otherwise relatively uncommon.

The preoperative diagnosis of a benign tumor in the neck may be made with some certainty based on an adequate index of suspicion from the history and physical evaluation and appropriate radiographic studies. In all cases, however, excision and histologic evaluation are necessary to establish an accurate diagnosis.

Paragangliomas

Paragangliomas are neoplasms that arise from paraganglionic "bodies" of the autonomic nervous system. Paraganglia are microscopic in size and are composed of granular T cells that contain catecholamines and Schwannlike satellite cells. These cells are of neuroectodermal origin. The carotid paraganglia (carotid body) is sensitive to changes in PO_2 and PCO_2. Batsakis (1980a) speculates that all paraganglias have a similar function.

The paragangliomas of the head and neck develop in the jugular fossa and middle ear; however, they may appear in the neck and parapharyngeal space.

Paragangliomas are encapsulated brownish tumors with a firm consistency. These tumors consist of clusters of epithelioid cells (Zellballen) separated by a highly vascular, fibrous stroma. Reticulin stains demonstrate the relationship between the clusters of cells and the stroma. Pyknosis and giant nuclear forms frequently are encountered, probably representing a degenerative phenomenon.

Early histologic staining techniques employing the chromaffin reaction failed to show the presence of catecholamines; thus reports referred to cervical paragangliomas as nonchromaffin (Berdal et al, 1962). Newer techniques, however, demonstrate catecholamines in low quantities in these cells (Kersing, 1977; Lack et al, 1977).

Neoplastic transformation in the adrenal medulla results in a pheochromocytoma that is histologically similar to a paraganglioma. Pheochromocytoma nearly always secrete catecholamines, whereas paragangliomas of the head and neck are rarely vasoactive. However, isolated reports of secreting laryngeal and carotid paragangliomas have been made (Hamberger et al, 1967; Justrabo et al, 1980; Strauss et al, 1983).

Because catecholamine-secreting head and neck paragangliomas are exceedingly unusual, routine preoperative screening for vasopressors in patients with a solitary paraganglioma of the head and neck is not indicated unless clinical indicators exist, such as a history of hypertension, palpitations, and blushing. Approximately 10% of patients with paraganglioma have a family history of paraganglioma; multiple lesions are present in 26%. Patients with multiple paraganglia are also at higher risk of having a functional pheochromocytoma and should undergo preoperative screening for vasopressor substances. Similarly, patients with a family history but without clinically obvious paraganglioma should undergo arteriography to rule out multiple clinically unrecognized lesions (Coia et al, 1981; Veldman et al, 1980).

A large series estimated the incidence of malignancy in paragangliomas to be approximately 5% (Soeprono and Hodgkin, 1983). Others, however, have estimated malignancy to be somewhat more common (Druck et al, 1976). Distinguishing malignant from benign lesions on the basis of histologic evaluation is not possible. Invasion of surrounding structures and metastasis are indicators of malignancy. Metastasis may be compatible with long-term asymptomatic survival in some patients (Soeprono and Hodgkin, 1983).

Vagal paragangliomas

Clinical features

Approximately 3% of all paragangliomas originate from the vagus nerve. Metabolically active tumors secreting catecholamines have not been described with lesions of vagal origin; however, such lesions could occur.

Vagal paragangliomas (glomus intravagale) most commonly arise in association with one of the vagal ganglia (Black et al, 1977). The superior (jugular) ganglion lies within the jugular fossa; approximately 1 cm caudally lies the nodose ganglion. Tumors arising in the area tend to be spindle shaped and to displace the carotid artery anteriorly and laterally. Paragangliomas of vagal origin develop less often along the superior or recurrent laryngeal nerve.

The breathy quality of the voice that the patient and physician both interpret as hoarseness may be the patient's first symptom. The history also may reveal aspiration of fluids caused by inadequate glottic closure. Paralysis of the tenth cranial (vagus) nerve causes the most common presenting symptom of vagal paraganglioma (Endicott and Maniglia, 1980; Hirsch et al, 1982). Black et al (1977) noted preoperative vocal cord paralysis in only three of nine patients with glomus intravagale. In a subsequent series, however, Hirsch et al (1982) reported that all six patients with intravagale tumors demonstrated vocal cord paralysis.

These tumors that are allowed to progress undiagnosed commonly result in additional symptomatology, depending on the location of the paraganglioma. Tumors arising in or expanding into the jugular foramen may be associated with symptoms involving paralysis of the ninth or eleventh cranial (glossopharyngeal or spinal accessory) nerves (jugular foramen, or Avellis', syndrome) (Fig. 89-1). This paralysis is manifest by dysphagia and shoulder drop. Paralysis and anesthesia of the palate cause various amounts of nasal reflux of fluids and aspiration. Large jugular foramen paraganglioma may involve the twelfth cranial (hypoglossal) nerve as well; the proximity of the hypoglossal canal to the jugular canal and the course of the nerve along the carotid sheath explains this involvement. When the jugular foramen is involved, patients frequently experience symptoms referable to the auditory system, most commonly tinnitus.

Tumors developing in the area of the superior (jugular) vagal ganglion may be dumbbell-shaped with an intracranial and extracranial component. Identifying with certainty the origin of large paraganglioma involving the jugular fossa, temporal bone, and surrounding skull base is impossible. Tumor expansion and subsequent pressure may cause pain.

Diagnosis of vagal paraganglioma requires a high index of suspicion. Vagal paraganglioma always should be sustected when vocal cord paralysis is observed in a patient with a solitary mass in the neck closely associated with the carotid artery. These lesions characteristically have lateral but not vertical mobility, reflecting their attachment to the vagus nerve. Pulsations, thrills, and bruits usually are not present.

Diagnosis of vagal paraganglioma

The diagnosis of vagal paraganglioma may be suspected more strongly after CT scanning with contrast, an important addition to the diagnostic armamentarium. The finding of an enhancing lesion at the carotid bifurcation is highly specific, and a vascular neoplasm elsewhere in the pharyngeal space must be suspected of being a paraganglioma. A neurolemmoma however, could mimic these lesions, but arteriography will demonstrate the highly vascular lesion characteristic of paraganglioma. A dense vascular stain with early shunting is characteristic (Brismar, 1980; Weber et al, 1982). Medial and lateral displacement of the carotid artery system is the classic pattern found by arteriogram, which we accept as confirmation of the diagnosis.

Incisional biopsy is contraindicated because of the vascular nature of the lesion and the possibility for uncontrolled hemorrhage. In addition, incisional biopsy may make removing the tumor more difficult because of obliteration of tissue planes and increased difficulty in preserving normal structures. To rule out multiple lesions, arteriography is essential especially in patients with a familial history of paraganglioma. Digital subtraction angiography (DSA) is a noninvasive method used to identify these vascular lesions. The detail DSA provided, however, frequently is insufficient to give the surgeon the information needed about the exact vascular supply. The blood supply is usually from the ascending pharyngeal, occipital, and internal maxillary arteries. DSA can be supplemeneted with intraarterial contrast studies, in which case it may be similar to arteriography in its importance and value.

Embolism is seldom feasible or necessary. The vascular supply of paraganglioma frequently arises very close to the carotid bifurcation. Selective cannulation of all the arterial feeders is not possible, and great risk for intracranial embolism exists. Also, the intensity of the inflammatory response following embolism may obliterate the planes of dissection, making subsequent surgical resection more difficult and dangerous. CT scanning must be done preoperatively to determine the superior extent of the tumor. Intracranial extension or involvement of the jugular fossa requires special preoperative planning.

Management

Surgical removal is the treatment of choice for vagal paraganglioma. Radiation therapy is reserved for patients deemed too ill to tolerate surgical management, as well as for lesions incompletely excised, recurrences, and metastases.

Surgical technique

Surgical removal is facilitated by the thorough preoperative evaluation of the tumor and adequate demonstration of its anatomic location (Fig. 89-2). Adequate exposure for most intravagal paragliomas is obtained through a transverse cervical incision made in a normal skin crease. The tumors may extend rostral to the mastoid tip and require a combined transmastoid, transcervical approach. When intracranial extension is noted, a posterior fossa craniectomy is required. When a combined transmastoid, transcervical approach is anticipated, a curvilinear postauricular incision is extended down into the neck along the anterior border of the sternocleidomastoid muscle to provide adequate exposure (Black et al, 1977; Hirsch et al, 1982). Skin flaps are elevated, with care taken to identify and protect all branches of the seventh cranial (facial) nerve. The carotid artery and jugular vein are identified and preserved. The vagus nerve is located low in the neck and followed in a cephalad direction until the caudal border of the tumor is identified. These lesions are characteristically fusiform in shape. The lesion is dissected free of the adjacent structures. The hypoglossal nerve usually is stretched out over the paraganglioma and must be identified with certainty and dissected free and preserved. Injury to the hypoglossal nerve may interfere with deglutition and complicate the patient's postoperative course.

The mastoid process is exenterated with cutting burrs, the facial nerve is identified and preserved, and the mastoid tip is amputated. The lateral sinus is skeletonized, and overlying bone is removed. At this point, the surgeon must decide if reflecting the facial nerve out of its canal anteriorly is necessary to facilitate removal of the tumor. Amputation of the lateral process of the first cervical vertebra facilitates access to the jugular foramen. The posterior auricular, descending pharyngeal, and occipital arteries may require ligation. The lateral sinus then is compressed, and the jugular vein is ligated if necessary. The inferior petrosal vein is plugged with bone wax to allow removal of the entire specimen intact. Employing suction drainage, the surgeon closes the wound primarily. Large tumors requiring extensive dissection in the parapharyngeal space may be associated with varying amounts of postoperative pharyngeal edema; a tracheotomy may be needed.

Extensive tumors involving the middle ear cleft may require reconstruction of the ossicles. Intracranial extension should be identified preoperatively. The presence of extension through the jugular foramen to the posterior fossa or, alternately, medial extension along the

carotid artery to the skull base or middle cranial fossa, requires special skull base techniques, frequently with a craniotomy, to encompass the tumor and provide the necessary visualization. Black et al (1977) noted that two of nine patients in their series had massive intracranial extension that was too close to the brainstem to allow total removal. Thus these procedures frequently are done jointly with neurosurgeons.

Removal of vagal paragangliomas may require the sacrifice of the involved vagus nerve. Paraganglioma intravagale implies that the nerve filaments are wrapped about the lesion, making resection necessary if the tumor is to be removed completely. The term *juxtavagale* describes lesions that develop in the adventitia of the vagus nerve (Hirsch et al, 1982). Such lesions may allow preservation of the nerve's integrity; however, return of function is unusual.

Rehabilitation of the patient's voice is an important consideration. Patients who had mobile vocal cords before surgery but have paralysis of the ipsilateral cord postoperatively may have Gelfoam paste injected under direct laryngoscopy if the surgeon is certain that the vagus nerve was not transected. This injection will improve the quality of the voice and decrease aspiration. If cord movement returns in 6 months, no further action is needed. When function has not returned in 6 months, vocal cord paralysis was present before surgery, or the vagus nerve was sacrificed, Teflon injection may be carried out early in the postoperative period to minimize disability.

Carotid paragangliomas

The carotid body is a paraganglion located near the bifurcation of the common carotid artery. The carotid body represents the largest collection of paraganglionic tissue in the neck. Mulligan (1950) referred to neoplastic degeneration of the carotid body as *chemodectoma,* which remains the popular name for this lesion. The term *carotid paraganglioma,* however, more accurately describes the tumor and its location.

Clinical features

The carotid body normally is responsive to changes in arterial pH, oxygen, and carbon dioxide tensions. Carotid paragangliomas rarely possess vasoactive properties. The most common presenting symptom of carotid paraganglioma is a neck mass located at the bifurcation of the common carotid artery. Very large lesions may produce symptoms of pressure, dysphagia, cough, or hoarseness. Occasionally patients report pain associated with carotid paraganglioma. The pathophysiology of this pain is unclear and most likely is caused by pressure on neighboring structures.

McIlrath and ReMine (1963) reviewed 64 patients with carotid paraganglioma treated between 1907 and 1962. In 60 patients (94%), a neck mass was the initial symptom (Fig. 89-3). They recorded pain or discomfort in 13 patients (20%). Dysphagia and hoarseness were uncommon. Because of its attachment to the carotid artery, the neck mass characteristically is mobile laterally but cannot be moved cephalocaudally. A bruit is frequently apparent (50% of patients). Carotid pulsations may be transmitted through the mass.

Diagnosis

Diagnosis requires a high index of suspicion. These lesions frequently can be confused with branchial clet cysts, metastatic tumors, malignant lymphomas, and tortuosity or aneurysmal changes in the carotid artery. CT scanning with appropriate contrast frequently allows preoperative diagnosis. Arteriographic evaluation of the carotid paraganglioma displays characteristic findings: a vascular mass in the bifurcation of the carotid artery and bowing and displacement ("lyre" sign) of the internal carotid artery. Incisional biopsy should not be undertaken; it serves only to obscure tissue planes and is potentially hazardous. The preoperative diagnosis should be based on radiographic findings.

Management

Cole (1977) reported treating 22 patients with jugular paraganglioma o the temporal bone with radiation therapy. He followed them for 1 to 17 years; typically, visible tumor residual remains unchanged or many years following radiation therapy. Two patients had progressive disease and subsequently died of paraganglioma 15 to 17 years after initial treatment. One patient developed metastatic disease and died 10 years after diagnosis. Two patients developed osteoradionecrosis of the temporal bone, which required surgical debridement; in each case a chronic draining cavity was the result.

Based on his experience, Cole recommends that 4000 to 5000 rads of supervoltage radiotherapy are necessary and concludes that he achieved excellent results. The use of orthovoltage radiotherapy early in the series was associated with several recurrences and a high incidence of radiation-related complications. In a follow-up report, Cole (1979) added three patients to his series and stated, "Wde have not yet encountered a case of glomus jugulare tumor with non-response to high voltage radiation in the doses recommended". The case in this series rarely had histologic conirmation of the diagnosis; Cole (1977) estimated that two patients actually had vagal paraganglioma.

Surgical excision of these lesions remains the treatment of choice in our department. The indolent nature of paraganglioma has led some authors to recommend only surveillance for asymptomatic elderly patients (Rush, 1962). Many of these patients become incapacitated by cranial nerve problems and pain as the untreated tumors become larger (Fig. 89-4). Present understanding of these lesions, skills in vascular reconstructive surgery, and observation of progressive symptomatology have led many surgeons to use more aggressive management protocols.

Carotid arteriography is routinely undertaken in patients with confirmed or suspected paraganglioma. This procedure allows establishment of the diagnosis, identification of the predominant blood supply, and can help in diagnosing multiple paraganglioma. Bilateral carotid arteriography should be used routinely on patients with familial paraganglioma.

Preoperative embolization is not recommended. The multiple vascular connections between the external carotid artery and the tumor are difficult to embolize, which makes accidental intracranial embolization a risk. More importantly, preoperative embolization may precipitate an inflammatory response that can obliterate the normal planes and will make subsequent subadventitial dissection extremely difficult.

Patients with recurrent paraganglioma, patients on whom prior biopsy has been performed, and patients with massive tumors in whom distal control of the internal carotid artery cannot be obtained should undergo temporary external carotid artery balloon occlusion with Xenon computerized estimation of blood flow as part of the arteriography procedure. These tests will allow the adequacy of contralateral cerebral profusion to be assessed. When temporary balloon occlusion is associated with central nervous system changes, or Xenon study demonstrates a decrease greater than 20 cc/100 g of tissue, the risk of permanent sequelae during temporary or permanent carotid interruption is significant. Accordingly, patients who demonstrate inadequate contralateral cerebral blood flow during these studies should undergo intraoperative EEG monitoring and should be managed with arterial shunting if vascular occlusion or repair becomes necessary.

Consulting an experienced vascular surgeon is often appropriate in patients who are operated on for recurrent paraganglioma and in patients with massive tumors in whom temporary carotid interruption may be necessary. This is especially prudent in patients in whom temporary balloon occlusion is associated with CNS changes.

Surgical technique

Surgical excision requires good exposure with identification of the carotid artery both proximal and distal to the tumor. Every effort should be made to identify and preserve the vagus, hypoglossal, and spinal accessory nerves because compromising these cranial nerves contributes significantly to postoperative morbidity.

Tumor removal requires subadventitial dissection. The external carotid artery should be ligated to gain adequate exposure of the tumor as it extends superiorly. Vasospasm of the internal carotid artery may be managed by local infiltration with lidocaine. Every caution should be taken to maintain the integrity of the internal carotid artery. Patients with large or recurrent tumors are best served if plans or vascular reconstruction are made preoperatively in the event that the common or internal carotid artery requires division. Severe ischemia thus can be reduced and its subsequent morbidity avoided.

Peripheral Nerve Neoplasms

The term *neuroma* typically is applied to tumors originating from peripheral nerves. Some confusion exists regarding the proper terminology to use when describing these neoplasms. Research has established that these neurogenous tumors originate from sheath cells of the peripheral nerves. Other terms for these lesions include *neurofibroma, fibroglioma, neurinoma, neurilemoma, neurolemoma, schwannoma,* and *neurofibroma.* Experimental evidence now shows that the parent cell o all these lesions is the Schwann cell. For practical purposes, the terms *schwannoma* and *neurofibroma* now are used most often. Some authors have used *schwannoma* and *neurofibroma* interchangeably.

Several large studies have focused on these relatively rare tumors. Das Gupta et al (1969) reported that in 303 patients with solitary benign neurogenous tumors, 136 (44.8%) had the neoplasm in the head or neck. These tumors most commonly appeared as a mass in the lateral neck. Under most circumstances the diagnosis was established histologically. Pain and neurologic dysfunction were unusual.

Mair et al (1976) noted that 16% of patients with peripheral nerve tumors share a high risk of developing an unrelated malignant tumor. This observation dictates close evaluation and follow-up of these patients.

Batsakis (1980c) argues that clinical differences exist between neurofibroma and schwannoma and that these differences have important diagnostic and treatment implications.

Schwannomas

The schwannoma is typically a solitary lesion; multiple lesions are unusual. It rarely is associated with von Recklinghausen's disease. The lesion is situated along a peripheral nerve; however, the individual axons characteristically do not traverse the lesion but are draped over its surface. Degenerative and cystic changes frequently are encountered. The solitary schwannoma tends to have a centrifugal distribution. Lesions may be painful; radicular pain or unpleasant paresthesias on light palpation may be present.

The schwannoma has a dinstictive pattern on histologic examination. A palisading array of nuclei about a central mass of cytoplasm has been termed *Antoni type A* tissue; a loose, surrounding stroma with no distinctive fiber and cell pattern is termed *Antoni B* tissue. Necrosis, cystic degeneration, and focal thrombosis are prominent in solitary schwannoma. Malignant change seldom occurs.

Schwannomas may develop in the neck from the cranial nerves, cervical sympathetic chain, cervical sensory plexus, and the brachial plexus. They may arise close to the spinal forament and extend into the spinal cord, forming a dumb-bell-shaped tumor. A presumptive diagnosis is based on signs of spinal cord compression and enlargement of the spinal foramen.

Lesions arising from the glossopharyngeal, vagus, spinal accessory, hypoglossal, or cervical sympathetic nerves may appear in the parapharyngeal space. Solitary neurogenous tumors usually are found in the lateral neck. They appear as a visible and a palpable mass (Fig. 89-5). The presence of pain suggests a schwannoma. The size of the solitary tumor varies and may range from a few millimeters to more than 20 cm in diameter. In many cases the nerve of origin cannot be demonstrated clearly.

Mair et al (1976) stressed that neurologic deficit is unusual. This increases the difficulty in making the diagnosis preoperatively.

Neurofibromas

Unlike the schwannoma, the neurofibroma is not encapsulated. Nerve fibers pass through the tumor and frequently are incorporated into it. Cystic and degenerative changes are uncommon. Histologic evaluation demonstrates a spindle cell pattern. Neurofibromas tend to be centripetal in distribution and largely asymptomatic. When associated with von Recklinghausen's disease, multiple neurofibromas are present. Approximately 6% to 16% of patients with von Recklinghausen's disease experience sarcomatous transformation of one of the neurofibromas (Heard, 1963; Holt and Wright, 1948; Preston et al, 1952).

Von Recklinghausen's disease

Von Recklinghausen's disease (neurofibromatosis) is an autosomal dominant trait with variable penetrance (Fig. 8906). Its incidence is approximately 1:3000 births (Fienman and Yakovac, 1970). Positive family histories are obtainable in only 50% of cases because of spontaneous mutation.

The most common initial findings are café-au-lait spots and neurofibroma (Fienman and Yakovac, 1970). Five or more light brown macules more than 1.5 cm in diameter are considered diagnostic. Two thirds of patients manifest physical findings by 1 year of age. The disease may be associated with other neurologic abnormalities or developmental anomalies, such as glioma and spina bifida. The tumors consist of pedunculated, violaceous skin appendages and subcutaneous nodules. The cranial nerves most often involved are the second and eight (optic and acoustic) nerves. Pedunculated skin tumors and acoustic neurofibromas (neuromas) are relatively common in adults; however, they are unusual during childhood. Williams and Pollack (1966) reported a syndrome of mucosal neurofibroma, pheochromocytoma, and medullary carcinoma.

Diagnosis

Tumors that involve the parapharyngeal space most commonly originate from the vagus or cervical sympathetic nerve. The initial finding may be a mass displacing the lateral pharyngeal wall or tonsil into the oropharynx (Fig. 89-7).

Imaging accurately predicts the origin of parapharyngeal space tumors. The parapharyngeal space is divided into a prestyloid and a poststyloid component by the fascia of the tensor veli palatini (Fig. 89-8). This fascial sheath in fact divides the parapharyngeal space into an anterior lateral component and a posterior medial component. Tumors arising from the poststyloid parapharyngeal space are almost always either paragangliomas or nerve sheath tumors. The distinction can be made with enhanced imaging techniques.

Management

Solitary nerve tumors are rarely malignant. Das Gupta et al (1969) note that benign peripheral nerve tumors can be separated easily from their nerve trunks. This is not always possible with cranial nerve lesions. Treatment consists of tumor removal, with every attempt made to preserve the integrity of the nerve of origin. Transoral biopsy or removal should be avoided because of the likelihood of infection and tumor dissemination. Good exposure with identification of vital structures is facilitated through an external neck incision, which allows proper hemostasis.

Surgical techniques

We favor approaching parapharyngeal space tumors through a transverse incision in a naturally occurring skin crease in the lateral neck. The platysma muscle is elevated along with the ramus mandibularis of the facial nerve. The submandibular gland is reflected anteriorly after ligation of the facial artery to provide entry to the parapharyngeal space; retraction of the great vessels posterolaterally provides additional access. Removal of large

tumors has been thought to require disarticulation of the temporomandibular joint anteriorly or a mandibular osteotomy through the body of the mandible. These techniques significantly disrupt the normal physiology of the mandible and temporomandibular joint and in our experience are unnecessary. Biller et al (1981) described a technique employing a midline labio-mandibulotomy, after which soft tissues are divided along the floor of mouth so that the mandible can be retracted laterally, allowing exposure of the great vessels and cranial nerves in the parapharyngeal space. The external carotid artery is divided at the level of the facial artery. The stylohyoid ligament and styloglossus and stylopharyngeal muscles are divided at the styloid tip, which allows access to the carotid canal at the skull base. We have not found this technique to be necessary in our series.

Excision of tumors that extend into the jugular foramen requires a transmastoid, transcervical approach (Hirsch et al, 1982). Following exposure of the cervical component, the mastoid process is exenterated with cutting burrs. The facial nerve is identified and preserved; if necessary, it can be displaced anteriorly out of its canal for added exposure.

The sigmoid sinus is skeletonized and then occluded allowing access to the jugular fossa and permitting removal of the tumor.

Traumatic Neuromas

Traumatic neuromas represent an abnormal attempt by an injured nerve to regenerate (Swanson, 1961). They rarely measure more than 2 cm and appear dense and fibrous with little vascularity, as demonstrated histologically by proliferation of entangled endoneural and perineural tissue, Schwann cells, and regenerating axons.

Persistent paresthesia and an unpleasant tingling in the area surrounding the neuroma are highly suggestive of a traumatic neuroma. Such lesions typically are found following radical neck dissection posterior to the carotid artery near the second cervical nerve. Excision is unnecessary in these cases, but it is diagnostic and therapeutic in patients who have had no previous surgery.

Lipomas

Lipomas are benign, encapsulated, subcutaneous collections of adipose tissue. Whether these represent true benign neoplasms, malformations, or hyperplasias of adipose tissue is unclear. Lipomas are found infrequently in the cervical region. They most often occur in the subcutaneous tissues (Fig. 89-9); however, they may be encountered in any compartment. Most lipomas are noninfiltrating and rarely recur even after enucleation. The infiltrating lipoma (Lin and Lin, 1974), which is nonencapsulated, may be located more deeply and tends to recur following incomplete removal (Figs. 89-10 and 89-11). Malignant degeneration of lipomas rarely occurs.

An unusual variant is the *spindle cell lipoma*, most often found in the supraclavicular and posterior triangles of the neck in older males (Enzinger and Harvey, 1975). These lesions appear as asymptomatic, slowly growing masses. The average lesion measures 4.5 cm. Histologically the lesion may be confused with a liposarcoma. The proliferation of spindle cells is uniform and no lipoblasts are present. In some cases an incorrect diagnosis of

schwannoma or leiomyoma is made; the distinction is important because the spindle cell lipoma is benign and easily cured by simple excision.

Several other disorders involving lipomatous tissue may occur in the cervical region (Batsakis, 1980b), including *benign symmetric lipomatosis (adiposis),* a disorder of unknown etiology in which massive deposits of adipose tissue develop about the neck. *Hereditary multiple lipomatosis* differs in that the adipose deposits involve the arms, legs, and torso, as well as the cervical area.

Lipoblastomas are tumors found in infants that result from abnormal growth of embryonal fat cells (lipoblasts). Kauffman and Stout (1959) have suggested that these represent an anomaly of growth rather than true neoplasms.

Hibernomas are rare tumors that originate from immature or "brown" fat. These lesions tend to be subcutaneous and benign (Mesara and Batsakis, 1967). Local excision constitutes adequate therapy.

Liposarcomas are rarely encountered in the head and neck. Stoller and Davis (1968) reported four cases in a population of 8.5 million during a 7-year period. These tumors are said to arise over again rather than from a preexisting lipoma.

Warthin's Tumors

Papillary cystadenoma lymphomatosum or *adenolymphoma* (Warthin's tumor) most often arises in the tail of the parotid gland of elderly male patients; it rarely may occur in the cervical lymphatic glands (Fig. 89-12) (Dietert, 1975; Handler and Miller, 1978; Norrish, 1935; Tuta and Apfelbach, 1942). These unusual lesions occasionally appear as asymptomatic, solitary neck masses. Diagnosis is made histologically. An adenolymphoma may be identified in cervical lymphatics during the histologic evaluation of a routine radical neck dissection. We know of no cervical Warhin's tumors that recurred following simple excision.

Extracranial Meningiomas

Extracranial meningioma may appear as a neoplasm in the neck. These are extremely unusual lesions; no guidelines concerning symptomatology or physical findings can be inferred from the literature. The diagnosis usually is made on histologic examination of the excised mass.

The source of ectopic meningioma is the subject of much discussion. The most frequently encountered extracranial meningiomas arise primarily from intracranial tumors with direct extracranial extension (New and Devine, 1947). These lesions appear at the skull base; examples include meningioma accompanying the cranial nerves as they exit the skull, extension of the tumor into the carotid canal, and erosion of the temporal bone (Laymon and Becker, 1949). Extracranial meningiomas have been found following erosion of a primary intracranial tumor through the cribriform plate (Tytus et al, 1967), orbital cavity (Stoller and Davis, 1968), and frontal sinus (Hallgrimsson et al, 1970).

Meningiomas may appear in the neck following metastasis of a benign-appearing intracranial meningioma. Verified metastases have been reported in the cervical and mediastinal lymph nodes (Opsahl and Loken, 1965); Shuagnshoti et el, 1971; Tytus et al, 1967).

The cell of origin of the meningioma is still disputed. The most popular theory is that the meningioma develops from meningocytic cells or the arachnoidal cells associated with the pacchionian granulations (Hallgrimsson et al, 1970; Shuangshoti et al, 1971). Extracranial cervical meningiomas may appear without any apparent connection with the intracranial contents, spinal canal, or foramina; these are hypothesized to represent embryonic cells rests. Alternately, the meningocytes may migrate with nerve roots during embryologic development. Hallgrimsson et al (1970) reported such a case associated with the spinal accessory nerve.

The diagnosis of cervical meningioma usually is made following removal for histologic study. No further treatment is necessary unless removal is incomplete. The surgeon must consider connection to an intracranial meningioma or the possibility of extracranial metastases of an intracranial meningioma on finding one of these unusual lesions.