Chapter 43: Neoplasms

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Tumors of the nasal cavity are intriguing growths because so many dissimilar pathologic conditions can exist in a small place. Since the nose and sinuses are really inseparable, limiting the discussion entirely to nasal cavity tumors is not possible.

Nasal tumors are quite rare. Because they involve the complex anatomy of the midface, the otolaryngologist - head and neck surgeon involved in treatment planning must understand how this anatomy and the pathologic features are related to the biologic behavior of the tumors. Treatment of neoplasms of the nasal cavity cannot be effectively planned based on what is done in other parts of the body or how similar tumors in other parts of the head and neck are treated. The physician who deals with these tumors must also understand the emotional impact on the patient of such growth in this area.

Epidemiology and Pathogenesis

Rarity and occupational clusters make nose and sinus tumors special in the study of cancer causation and prevention. In the USA the incidence of nasal tumors is less than one case in 100.000 persons per year. In Japan and Uganda the rates are more than twice that (Roush, 1979). Reports of large series of nasal tumors are more common from the UK than from the USA. Except for nonepithelial tumors, the malignant growths are nonexistent in children.

Roush (1979) has estimated that up to 80% of all malignant tumors in humans are related to environmental causes. That estimate is probably high, but some of the tumors of the nose and sinuses are related to environmental factors. The evidence for an environmental agent in the causation of nasal tumors is strongest for nickel exposure. Results of retrospective cohort studies of nickel-refining workers in Wales, Norway, and Canada imply a risk factor 100 times greater in persons in this occupation than in other persons. The risk increases with length of exposure and age at first exposure. These countries, which had recognized the environmental risk before epidemiologic studies confirmed that it existed, have worked effectively to reduce exposure (Barton, 1977). The risk from nickel appears to involve only those who work in the refining process, not those whose jobs are in the manufacturing processes.

Epidemiologic evidence also exists of an increased risk of nasosinus cancer in woodworkers, boot and shoe workers, and furniture makers. Because of the complexity of the environmental exposure in these groups, the responsible agents have not been identified.

Hydrocarbon exposure is also thought to carry an increased risk of nasal cancer. Other agents suspected are mustard gas, isopropyl oil, and hydrocarbon gas. All these agents are related *only* to squamous cell carcinoma. The other tumors occur too infrequently for clustering of cases to be found.

Several nonoccupational agents may cause cancer of the nose and sinus. Maxillary sinus cancer has been reported in persons in whom thorium dioxide (Thorotrast), a radioactive agent, was injected into the sinus and left in place for a long time. Snuff use is

also suspect in parts of Africa, where the snuff is different from that used in Europe and North America.

Frequency by Site

Hospital studies characteristically report that about 55% of nose and sinus tumors originate from the maxillary sinus, 35% from the nasal cavity, 9% from the ethmoid sinus, and the rest from the other sinuses. With large tumors the site of origin can only be guessed at, since all these sites merge.

Diagnostic Assessment

The symptoms of nasal tumors, whether benign or malignant, are not striking. No unfailing signs or symptoms are produced only by malignant tumors. Nasal obstruction, blood-tinged mucus, and epistaxis are the most common symptoms. In an early stage the symptoms are not those that would encourage the person to seek medical attention. Facial asymmetry, loose teeth, and sensory changes around the nose are less frequent late symptoms. At times, a nasal tumor is diagnosed only when a pathologist examines polyps or what he thought to be nasal polyps. Tearing on one side may be an early symptom of a lateral nasal, medial antral, or ethmoid tumor.

Butlin's (1887) classic description of the progression of symptoms of the malignant tumors can hardly be improved:

"Sometimes the appearance of the tumor is preceded by pain, but in many instances there is no pain until the disease is advanced. The first sign of serious disease is the appearance of a swelling of the face over the antrum or of fullness and obstruction of the corresponding side of the nose. With the fullness of the nostril there may be discharge of bloody fluid. The swelling gradually increases, not only in the directions in which it first was noticed, but also up toward the orbit, down towards the mouth, and back into the spheno-maxillary fossa. The eye may be pushed up and the hard palate pushed down, but the swelling in the fossa is not so easily perceived. The nostril on the affected side often becomes completely obstructed. As the disease advances, the bony wall may be destroyed and protrusion may take place, with affection on the soft parts around the bone. The skin of the face in this way becomes adherent to the tumor and immovable over it, and the result may be a vast ulcer, with the thrusting forth of a fungous mass".

Physical Examination

Observation and palpation are the key elements in the clinical examination of a patient with a known or suspected tumor of the nose or sinus. The physician should look for facial asymmetry, eye position in relation to the opposite eye, tooth position and stability, ease or difficulty of nasal breathing, voice character, and mobility of the lower jaw (presence or absence of trismus).

Careful palpation of the orbital bony contour and the eye for protrusion should be done. The face and the anterior wall of the maxilla under the lip and the alveoli should be palpated as well as inspected. The divisions of the fifth cranial nerve are carefully tested for sensation with a cotton wisp.The nasal cavity is examined with a nasal speculum before and

after shrinkage with phenylephrine or a dilute cocaine-epinephrine solution. Abnormal tissue and its location are observed. Polypoid material may be found along with the tumor. Careful inspection of the posterior choanae and nasopharynx with the mirror or optical instruments is important. One looks for direct extension of the tumor to this area, and in the case of adenoid cystic carcinoma, one can at times see vascular striations over the eustachian tube orifice and into the nasopharynx. When biopsied, these striations may show submucosal tumor.

Radiographic Diagnosis and Treatment Planning

Precise determination of the extent of a tumor is essential in treatment planning and execution. Underestimation of the size and extent can lead to underexcision or failure to irradiate tumor fully. Overestimation can lead to unneeded excision or larger radiation fields than are required. Underestimation, the usual error, is more dangerous. Today, with the extraordinary images obtained by computed scans, the principal purpose of plain radiographs, tomograms, and computed tomography (CT) scans is to determine if a meaningful operation can be done, that is, if surgery is one the treatment options. It is disappointing, expensive, and not helpful to the patient to subject him to a serious operation only to find that gross or microscopic tumor must be left behind. Although some would say that tumor left behind can be managed with some other treatment in a meaningful way (Majumdar and Kent, 1983), as an oncologic principle, that kind of thinking is dangerous. Being completely certain that surgical intervention will remove all of a tumor is never possible, but certain guidelines are available.

An aphorism that has served well in treatment planning for nasal tumors is "the problem is either worse or better than it appears on radiographs". Plain radiographs and tomograms cannot delineate tumor extension that has not destroyed bone. Radiographs and tomograms cannot distinguish between tumor and inflammatory tissue. When no bony destruction exists, xeroradiograms and computed scans cannot differentiate actual tumor from inflammatory reaction or inflammatory polypoid tissue. The new computed scans are far superior to tomograms and plain radiographs for tumor evaluation. Differences in tissue densities permit conjecture about where tumor stops and inflammatory tissue begins, but distinguishing microscope extension is beyond the capability of computed scans.

Robin and Powell (1981) point out the likely sites of radiographic error: the ethmoid complex, the pterygopalatine fossa, the infratemporal fossa, the face, and the orbit. They note that the usual methods of tumor assessment are in error at least one third of the time (this study did not have the benefits of the computed scan in all patients). Robin and Powell suggest that staging surgery is the best treatment planning technique, although the premise of staging surgery is that if tumor is left behind in inaccessible places, something else can be done to complete the treatment.

We do not now have a completely accurate, noninvasive way of evaluating nasal tumors. The best one can do is to use guidelines to operations and to establish indications in advance of when an operation can or cannot be done with the goal of curing the disease.

Complete removal of a nasal tumor is not possible if there is bony destruction of the skull base, except in the cribriform region or the roof of the ethmoid sinus, which can be encompassed surgically en bloc. The farther back in the ethmoid sinus that bone destruction occurs, the less likely complete resection becomes. Cure of malignant tumors by surgery is unlikely if there is bony invasion of the orbital plate of the frontal bone, the posterior wall of the frontal sinus, the wings of the sphenoid bone above the infratemporal fossa, or the pterygoid process of the sphenoid bone.

Bony destruction of the orbital floor does not preclude meaningful surgery. Bony destruction of the roof of the antrum can be contained although complete removal is less likely if bony destruction occurs posteriorly near the orbital apex. Clinical signs of infraorbital nerve anesthesia and eye displacement (forward or lateral) usually attend bony destruction. This bone loss can be seen in the anteroposterior tomogram and computed scan.

Other contraindications to curative surgery are extension into the nasopharynx, extensive bilateral involvement, distant metastasis, and insurmountable regional metastasis. The rapidly expanding field of skull base surgery may change the probabilities of curable resections when tumor involves the skull base, but no conclusive evidence of this exists in late 1991.

Malignant Neoplasms

Squamous cell carcinoma is the most common malignant tumor of the nose. Squamous cancers can originate from the nasal vestibule, the lateral wall of the nose, the turbinates, the meatus, or the septum.

Vestibular cancers are really skin cancers in the anterior portion of the nose. They can metastasize, and when they do, the periparotid and submaxillary nodes are the first groups at risk. Because the lesions are anterior, near the face, and easily seen, diagnosis and treatment planning should not be difficult. The accepted treatment for vestibular cancer is excision or irradiation. When excision is used, a small antral rhinotomy may be helpful for exposure. Multiple flaps designed from the nasolabial area can be used to close the defect. Free skin grafts can also be used.

Nasal septal cancers are rare. Beatty et al (1982) found only 58 squamous cancers of the septum in their review of all the nasal tumors treated at the Mayo Clinic from 1915 to 1976. Lewis and Castro (1972) reviewed 772 cases of malignancies of the nasal cavity and sinuses, and none cam from the septum. Beatty et al (1982) recommended wide surgical excision by rhinotomy and frozen-section microscopic control of margins. Large lesions may require extensive procedures, including septectomy, rhinectomy, and even maxillectomy with orbital exenteration. Elective neck dissection does not seem necessary.

The overall survival of patients with carcinoma of the nasal septum is good compared with that of patients with squamous cancers of the sinuses. Nearly 7 of 10 patients treated surgically were cured with the initial treatment. The farther back on the septum the tumor originates, the poorer the outlook. Large tumors that have advanced to other sites within the area have a poor prognosis.

Squamous cell cancer of the lateral wall of the nose

Separating squamous cell cancers of the nose from tumors that originate in the sinuses is sometimes very difficult and may be unnecessary, because the lateral wall of the nose is the medial wall of the maxillary sinus and part of the ethmoid complex. The treatment of these tumors is unsettled. What we do today is not much different from what was done 100 years ago. In 1848 Fergusson noted that "for centuries tumors have been removed from this area". The earliest reference he found in the English literature was published in 1671. These primitive operations consisted of cutting, tearing, and scraping away far-advanced tumors and applying hot irons to the tumor base. Fergusson contrasted that treatment to what he called the "modern operation", which began in the early 1800s and consisted of exposing the area with skin incisions, sawing and chiseling the bony attachments, and wrenching the part out with a large forceps. Today the chisels, saws, and large forceps are different, but the concept is not. The incisions are different, and instead of inserting a hot iron, surgeons sometimes use pulse laser beams, ionizing particles, or profound cold. Fergusson's operation took minutes and was done without anesthesia. Today the procedures takes hours and is done with anesthesia. The differences in survival probably relate more to the availability of transfusable blood, antibiotics, and safe anesthesia than to any conceptual or oncologic differences. Fergusson's patients died of shock, sepsis, or sheer exhaustion, whereas patients today who die do so more slowly from cancer. Few surgeons of that time were satisfied with the results of treatment, and that remains true today.

With the introduction of each new form of treatment capable of destroying malignant tissue has been the promise of sophistication and ease of treatment at extra expense. However, the percentages of patients actually cured has not changed much. Electrosurgery was followed by treatment with radium. Radium was followed by irradiation from conventional machines, cobalt source, linear accelerators, and particle emitters. After initial enthusiasm, physicians have tended to return to the surgical procedures using knives, hammers, chisels, and saws.

Each new way of delivering radiation has been an improvement, because uniform delivery could be more nearly precise. Unfortunately, the results of radiation therapy alone for these squamous cell cancers have remained unsatisfactory. The anatomy of the nose and adjacent sinuses and the vagaries of tumor in or near bone make delivery of homogeneous and effective dosage difficult. Tumor hypoxia or necrosis, overlapping fields, and undertreatment of portions of the tumor make actual cure of nasal tumors by irradiation alone unpredictable and unlikely.

The current preferred treatment for large or aggressive lesions is combined therapy with irradiation and surgery used in sequence for potentially curable tumors. The concept is that if the combination is done properly and the sequence and timing are correct, survival will be increased for patients with tumors of comparable stages over what could be accomplished with irradiation or surgery alone. The concept is rational, but it may not be true. If it is true, the percentage increment of cured patients may be so small that it is unmeasurable. It is not known whether irradiation should be done before surgery or after. Irradiation before surgery is appealing from a radiobiologic point of view because the better oxygenated parts of the tumor will be reduced in volume. Unfortunately, so many of these tumors are partially necrotic that hypoxic tumor is irradiated. There is no certainty that

killing part of a tumor that is to be removed is helpful. It may even be harmful, since tumor is left in place longer in a lymphatic field altered in a negative way by irradiation. It is theoretically possible that the positive benefit of tumor destruction by irradiation is counterbalanced by the negative effect on regional tumor resistance. There is no evidence that patients who have no apparent viable tumor in a specimen after preliminary irradiation have a better prognosis than those in whom tumor remains (Ellingwood and Million, 1979; Jackson et al, 1977; Lund, 1983; St-Pierre and Backer, 1983).

Although irradiation before surgery is theoretically ideal, postoperative irradiation is presently advocated. At one time it was said that if irradiation were needed after surgery, it should have been given before. The theoretic basis for postoperative irradiation is the concept of "microscopic disease", which is based on Cohen's (1968) animal experiments with adenocarcinoma. The idea is that "microscopic" cancer (cancer than cannot be seen with the unaided human eye) is more vulnerable to irradiation than is gross cancer and that at certain levels of irradiation below normal tissue tolerance most or all microscopic cancer will be destroyed. This concept has not been verified in humans with squamous cell carcinoma.

If the concept is accepted, the oncologic implications are that one can safely leave microscopic tumor behind (have positive surgical margins) and then irradiate afterward with predictable safety. To take the concept one step further, one could say that less-than-radical removal might be tolerated if irradiation follows. This idea has been proposed. Robin and Powell (1981) ask whether "radical" resection is ever appropriate, because "radical" removal, they believe, rarely succeeds. They propose subradical surgery followed by irradiation. *Radical* means removal of all tumor tissue; *subradical* is partial removal or what some might call debulking. The argument is that if a complete operation is not satisfactory, a less radical one followed by irradiation might be more acceptable. This oncologic concept is difficult to accept.

The use of radiation therapy in combination with surgery for early curable lesions must be carefully considered in light of the long-term problems associated with combined therapy. Morita and Kawabe (1979) observed that in all their patients who received over 5800 rad to the eye, severe panophthalmopathy with corneal ulceration developed within 2 years of radiation therapy. Of 21 eyes exposed to from 2800 to 5400 rad, 18 experienced visual disturbances and radiation-induced cataracts.

In summary, surgical excision is still the mainstay of treatment of squamous cancers of the nose. If removal is complete, the patient should do better than if tumor is left behind. Small tumors have a better prognosis than large ones. The best way to improve survival still seems to be early diagnosis. Combined therapy is acceptable but unproved as a concept, and nobody really knows the most effective way to combine irradiation and surgery.

Adenocarcinomas

Neoplasms that come from the minor salivary glands of the nose form an interesting and diverse group that comprises less than 10% of the malignant tumors. There are two types of adenocarcinomas: adenoid cystic carcinoma and "other" glandular tumors. In each group are high-grade and low-grade tumors.

Adenoid-cystic carcinomas

Low-grade adenoid cystic carcinomas are those in which the histologic pattern is a mixture of tubular and cribriform formations of epithelial cell with few solid cellular areas. The cribriform pattern is the classic "Swiss-cheese" pattern of cells arranged in elongated tubular structures. High-grade adenoid cystic carcinoma is the pattern of the classic tumor, but solid areas of malignant cells make up a significant volume of the tumor mass. This distinction between high and low grades seems important. Vascular invasion is more common, distant metastasis more frequent, and death as a consequence of the tumor more likely in patients with a high-grade tumor (Goepfert et al, 1983). Perineural invasion is a feature common to both types, as is frequent local recurrence.

The treatment for adenoid cystic carcinoma is surgical resection when reasonable. The more thorough the excision, the longer a patient can expect to be free of recurrence. Although considerable pessimism exists about the curability of adenoid cystic carcinoma, ample justification for curative attempts exist. Five-year survival is not adequate to define cure. The 10-year recurrence-free rates for patients with nasal fossa tumors are in the 60% range, an outcome that encourages vigorous treatment. Some patients live their normal life spans with or without recurrences, even with pulmonary metastasis.

The role of radiation therapy is not clearly defined. No evidence suggests that preoperative or postoperative irradiation decreases the frequency of recurrence. With low-grade adenoid cystic carcinoma I prefer to reserve irradiation for patients with recurrence and pain. The carcinoma responds to irradiation, and pain can be lessened or eliminated, although the result is seldom permanent. Some of these tumors are so extensive that no surgical procedure short of removal of the midface and both eyes is feasible. In the very elderly patient with an extensive tumor but little pain, the decision not to treat may be proper. I have used support without surgery to care for several elderly persons with extensive nasal adenoid cystic carcinoma. They lived out their lives and died of something other than the tumor.

Other adenocarcinomas

The "other" glandular carcinomas are classified in several ways. One method uses the same terminology as that used to describe tumors of the major salivary gland. The diagnoses used are mucoepidermoid and acinic cell carcinoma. Batsakis et al (1980) described a classification based on growth forms: papillary, sessile, and alveolar-mucoid. Mucin production is variable in the first two forms, and the level of cellular differentiation may range from modest to poor.

Papillary adenocarcinomas are usually the most localized. A special type of mucus-producing papillary adenocarcinoma is described that looks like colonic carcinoma microscopically. These tumors are composed of tall columnar cells that form a single layer containing numerous goblet-shaped, mucus-producing cells. The importance of this variant is to know that it exists so that an extensive evaluation of the digestive system can be avoided.

A *sessile* adenocarcinoma has a broad surface, and the tumor cells retain little resemblance to the cells of their origin. These tumors have greater invasive properties and carry a worse prognosis than the papillary adenocarcinomas.

The *alveolar-mucoid* variant is the most aggressive and has a poorer prognosis than the papillary tumors. These carcinomas are characterized by abundant mucin in which nests of individual cells reside.

All nasal adenocarcinomas other than adenoid cystic carcinoma are considered to be relentless, aggressive tumors with a poor prognosis. An Armed Forces Institute of Pathology (AFIP) study of 50 cases suggests that the low-grade lesions can be distinguished from the other types and treated with optimism. For the high-grade tumors, combined therapy is used, but the results of any treatment are disappointing (Heffner et al, 1982).

Sarcomas

Sarcomas of the nasal cavity are rare; the only ones reported are chondrosarcomas of the nasal septum (Bailey, 1982; Coates et al, 1977). These tumors cause symptoms late and are often so large at diagnosis that determining their exact site of origin is impossible.

Coates et al reported on 13 nasal chondrosarcomas seen in a 25-year period at the Mayo Clinic. The typical chondrosarcoma was of low grade histologically and was a large, pale, glistening, obstructing mass. Local excision was used in seven of the patients, and tumor recurred in five of them. A definitive en-bloc excision cured four of six other patients. The others had protracted courses with local recurrences and finally died from uncontrolled disease that extended intracranially. Radiation therapy did not influence tumor growth after recurrence. I have treated a 32-year-old woman who had nasal chondrosarcoma diagnosed at age 19, wide excision by rhinotomy at age 19, and bilateral maxillectomy 13 years later.

Malignant melanoma

Malignant melanoma is a rare nasal tumor. When it develops, it is more likely to arise from the nasal cavity than from the paranasal sinuses. The sites of origin are the mucous membranes of the septum and the lateral nasal wall. Freedman et al (1973) reviewed the Mayo Clinic experience with 56 melanomas from these sites. Lund (1982) added a review of 36 patients from England.

The symptoms of melanoma are the same as those of all nasal tumors: obstruction and epistaxis. The diagnosis is usually obvious because of pigmentation, although some of the pigmented area may not be tumor. Tissue phagocytes can carry the pigment some distance from the real tumor. The distinction between tumor and submucosal benign pigmentation requires histologic study. If the diagnosis is made early enough and the tumor is completely removed, cure is possible. In the series reported by Freedman et al (1973) nearly 60% of the patients were alive without disease at 5 years. The patients in the Lund (1982) series did not fare as well: nearly two thirds were dead at 5 years. This difference may be related to earlier diagnosis in the former group than in the latter, or it may be the result of different treatment. More radiation therapy, which may have changed local resistance, was used in Lund's series.

The incidence of local recurrence is high. In one series cited by Freedman et al

(1973), 21 of 38 patients treated surgically had local recurrences. More radical excision, including removal of the eye, palate, or external portion of the nose, may not decrease the incidence of local recurrence. The tumor can be multifocal, and safe removal requires the help of a pathologist skilled in diagnosis based on fresh frozen section.

The disconcerting feature of nasal melanoma is that the probability of death does not decrease with time as it does with other tumors. The recurrence curves are linear over time, whereas with other epithelial tumors the risk of death decreases greatly after a patient survives 4 or 5 years without recurrence. Death from melanoma is usually by disseminated disease. The late deaths represent silent disseminated disease held in check by a competent immune system until some other time. Local recurrence and even metastasis do not necessarily imply death within a short time. Patients with both have survived for several years. Vigorous secondary local excision may be worthwhile if doing so is possible.

Olfactory esthesioneuroblastoma

A peculiar and rare neuroepithelial malignant lesion that comes from the olfactory area of the nasal cavity is olfactory esthesioneuroblastoma. Other terms used for this tumor are *esthesioneuroblastoma, olfactory esthesioneuroma, neuroesthesioma, and olfactory neurocytoma.* The growth was described by Berger et al (1924). Neuroectoderm is the presumed cell of origin because of a histologic pattern similar to that of the malignant tumors of the sympathetic ganglion, adrenal medulla, and retina. McGavran (1970) and Taxy and Hidvegi (1977) demonstrated neurosecretory granules of catecholamines in tumors examined by electron microscopy.

Cantrell et al (1977) note that of the 160 tumors reported since 1924, 125 have been cited in the past 15 years. This finding reflects a greater awareness of the tumor; in the past, these tumors were diagnosed as something else. Their histologic resemblance to undifferentiated small cell carcinoma can cause misdiagnosis. The histologic features that identify the esthesioneuroblastoma are neuroepithelial cells arranged in a "pseudorosette" pattern, a surrounding stroma composed of undifferentiated nuclei and fibrillary cords, marked microvascularity, and palisading neuroepithelial cells around blood vessels. Mitotic figures are absent, and there is occasional interstitial calcification. A similarity to adrenal or a sympathetic ganglion malignancy aids the pathologic diagnosis.

On gross examination, the tumors appear as red, polypoid masses high in the nose. The nose can bleed and become obstructed. Symptoms can be present for months or years. One of my patients had progressive nasal obstruction for 10 years and was completely obstructed for 6 years before diagnosis. The tumor can be multicentric, with separate tumors above and below the cribriform plate. Olsen and DeSanto (1983) describe one patient, who had two separate tumors above and the other below the cribriform plate, with no gross or microscopic connection between the tumors.

Djalilian et al (1977) found that in one of four patients, tumor in the anterior cranial cavity was present at diagnosis or developed later. They noted metastasis in 8 of the 19 patients they studied. Metastatic lesions in cervical lymph nodes were present in two children at diagnosis and in two adults 6 and 8 years after the nasal tumors were removed. I treated one patient in whom bilateral parotid node metastasis, bilateral cervical metastasis, frontal lobe esthesioneuroblastoma, and thyroid gland metastasis developed 6, 9, 11, and 13 years

respectively, after the nasal tumor was removed. The nasal tumor did not recur.

Surgical removal is recommended. Kadish et al (1976) reported on a patient whose tumor responded to irradiation. Olson and DeSanto (1983) as well as Cantrell (1977) advocate combined therapy with irradiation after surgery (Olson and DeSanto) or irradiative radiation before the surgery (Cantrell). The goal of combined therapy is said to be to decrease the incidence of local recurrence. The need for combined therapy has been recently questioned by Biller et al (Biller et al, 1990). Biller's series of 20 patients with extracranial disease were treated by combined craniofacial resection without radiotherapy. There was one local recurrence. The small recurrence rate was attributed to an adequate excision as compared with the rhinotomy alone. The combined approach permits resection of the dura over the cribriform plate, resection of the olfactory bulb, and en-bloc resection of the ethmoid labyrinth, lamina papyracea, septum, and cribriform plate. Lateral rhinotomy may be sufficient for small tumors that originate from the lateral wall of the nose away from the cribriform area. The concept of a combined frontal craniotomy-superior rhinotomy approach, which includes the cribriform area, is sound for the superior based tumors (Chapman et al, 1981).

With so few patients treated, the likelihood that this may be a multicentric tumor, and so many forms of management advocated, no reliable data exist to substantiate which treatment is best for this tumor. The beginning of a rational classification system (Biller et al, 1990) will help sort out the best treatment and ultimately answer whether combined therapy is needed after en-bloc resection. Until then, individualized treatment by adequate resection is important.

Hemangiopericytoma

Nasal hemangiopericytoma is of vascular origin and can arise wherever capillaries are found. Occurrence in the nose is rare. Fewer than 20 cases involving the nasal cavity have been reported.

The histopathologic features of this tumor are complex, with a single common denominator: lack of uniformity in appearance and biologic behavior. In all tumors, a proliferation of capillaries is surrounded by a connective tissue sheath. Outside this sheath are tumor cells that are varied in their individual appearance. Special staining with silver stain blackens the sheath of the capillaries for easy identification and thus allows accurate demonstration of the extracapillary position of the malignant cells (Brown, 1977).

The biologic behavior of the tumor cannot be predicted by its histologic appearance. Nonmitotic tumors can metastasize, and metastasis can occur with a benign-looking primary tumor. Distant metastasis to lung, liver, and bone is common, but regional metastasis to lymph nodes has not been observed. The gross tumor in the nose looks benign and is described as a soft, rubbery, pale gray or tan polypoid mass. Despite the pale, avascular appearance, these tumors bleed vigorously when biopsied.

Treatment consists of wide local excision. *Irradiation* decreases the size of the tumor, but cure with radiation therapy alone in other sites is rare. Recurrence after excision is frequent, but few deaths occur in the first 5 years. Most of what is suspected about these tumors in the nose is inferred from what is known about tumors at other sites.

Benign Neoplasms

Benign tumors of the nose are rare in comparison with malignant growths. In decreasing order of frequency, the benign tumors are osteoma, hemangioma, papilloma, and angiofibroma. The two tumors of greatest interest are the inverting schneiderian papilloma and the nasopharyngeal angiofibroma.

Inverting papilloma

Most authorities consider the inverting papilloma a true neoplasm. Other names used for this growth are *inverted papilloma, schneiderian papilloma, papillary sinusitis, polyp with inverting metaplasia, benign transitional cell growth, epithelial papilloma, inverted schneiderian papilloma, soft papilloma, transitional cell papilloma, squamous papillary epithelioma, papillary fibroma, papillomatosis, and cylindrical cell carcinoma.* These names reflect a variety of histologic interpretations, which range from an inflammatory disease to a malignant neoplasm.

The microscopic features that distinguish a papilloma from an allergic or inflammatory polyp are well known. These features include proliferation of the covering epithelium and extensive fingerlike inversions into the underlying stroma of the epithelium.

No symptoms are unique to these tumors. Obstruction occurs, but bleeding is rare. A history of previous operations, including polypectomy and septal or sinus surgery, is common. Suh et al (1977) noted that 63% of patients who had definitive surgery for papilloma had had nasal surgery for obstruction. In 36 patients who had undergone previous surgery, 76 nasal operations had been performed before papilloma was diagnosed.

Most papillomas originate from the lateral wall of the nasal space. Only 2 of the 57 patients cited by Suh et al had papilloma limited to the maxillary sinus. Secondary involvement of the maxillary and ethmoid sinuses is common. Extension to the sphenoid and frontal sinuses also occurs. Radiographic studies may show bony destruction and erosion, usually in the lateral wall of the nose. Mabery et al (1965) reviewed the issue of malignant change in a papilloma. In the Suh et al series of 57 patients, 4 (7%) had invasive carcinoma associated with papilloma. One of the patients had undergone prior irradiation for papilloma.

Total removal is needed to avoid recurrence; however, complete removal by transnasal excision or simple sinus surgery is not possible with most growths. Lawson et al (1983) proposed that there may be a small subgroup of patients with tumor limited to the inferior or middle meatus or turbinate, as verified by CT scan, who can be treated by intranasal or transantral surgery. For the rest, an en-bloc operation via lateral rhinotomy seems best.

In the past, recurrence rates with simple excision exceeded 75%. The use of nonmutilating but adequate operations, such as rhinotomy, reduces the rate of recurrence. Even after rhinotomy, however, there can be recurrences (Lawson et al, 1983). Careful histologic study of multiple sections of the excised lateral wall of the nasal cavity is recommended to seek areas of in situ or invasive carcinoma.

Juvenile nasopharyngeal angiofibroma

Juvenile angiofibroma is a peculiar benign tumor typically found in young boys who have nosebleeds and nasal obstruction. Symptoms are present for months to years. Most patients are adolescent boys, but some are older and a few are younger. In a series of 120 Mayo Clinic patients with angiofibroma, the ages ranged from 9 to 29 years, with a mean age of 15 at diagnosis; all patients were male (Neel et al, 1973).

Location

The precise site of origin of the angiofibroma has been the subject of much speculation. At onset the growth is a lateralized tumor. It originates in the posterior nasal cavity rather than in the nasopharynx itself. The specific site of origin is the posterior lateral and superior nasal cavity at the point where the sphenoidal process of the palatine bone meets the horizontal ala of the vomer and the root of the pterygoid process of the sphenoid bone. This is near the upper margin of the sphenopalatine foramen.

At the time of diagnosis, most of the tumors have extended beyond the nasal cavity and nasopharynx. They push forward behind the posterior wall of the maxillary sinus, pushing the thin bony wall ahead. Lateral growth puts the tumor in the pterygomaxillary fossa. The nasal septum is crowded to the opposite side of the nose. Extension into the pterygomaxillary fossa can erode the pterygoid process of the sphenoid bone. Further lateral extension can fill the infratemporal fossa and produce the classic bulging of the cheek. Tumor can extend under the zygomatic arch and cause swelling above the arch. From the pterygomaxillary fossa, the angiofibroma can grow into the inferior and superior orbital fissures, erode the greater wing of the sphenoid bone, and leave tumor extradurally in the middle fossa near or adjacent to the cavernous sinus. Posterior extension into the sphenoid sinus through its floor or ostium fills the sinus, pushed upward and back to displace the pituitary, and then can fill the sella turcica. Tumor in the sella or in the orbit can cause loss of vision. The rate of growth of the angiofibroma is not known, but usually growth is slow.

Because the tumor is rarely seen in young adults, spontaneous regression is believed to occur. Clinically, regression after incomplete removal or radiation therapy is recognized. Because regression cannot be assumed, these tumors should be treated.

Radiographic studies

The radiographic findings of angiofibroma are characteristic. Holman and Miller (1965) emphasized the bowing of the posterior wall of the maxillary sinus and the enlargement of the superior orbital fissure. Computed tomography is ideal for precise localization and has essentially replaced conventional tomography in treatment planning (Fig. 43-1).

Some have advocated angiography as useful for diagnosis and treatment planning. Areas of tumor extension may be more clearly identified than with CT scans, particularly intracranially. As CT scans become increasingly detailed, however, the extra risk and cost of angiography becomes less justified unless embolization is used.

Biopsy

Usually diagnosis is so little in doubt that preliminary biopsy is more trouble than it is worth. Excisional biopsy is preferred. If the histologic diagnosis is in doubt, a biopsy specimen can be taken after the patient is asleep and has been prepared and draped for tumor removal. This approach can avoid troublesome bleeding between biopsy and definitive treatment.

Blood supply

The main blood supply of the angiofibroma is from the internal maxillary artery. Other vessels that can contribute are the dural, sphenoidal, and ophthalmic branches from the internal carotid system. Vessels sometimes carry blood supply from the thyrocervical trunk. Because of this diverse arterial input, preliminary ligation of the external carotid artery is of little help in decreasing bleeding during excision. Ligation of that artery before definitive surgery can actually increase the problem of bleeding at excision by encouraging arterial input from the less accessible or inaccessible vessels. At times, the internal maxillary artery is pushed forward by the tumor and can be ligated during the exposure phase of surgery before excision. When practical, this ligation of the artery in the course of definitive treatment is worthwhile. The vascularity of the tumors varies. Some are very vascular and others quite fibrous. Thus the bleeding potential of a given tumor is unpredictable.

Techniques to decrease bleeding

Many techniques have been advocated to lessen bleeding. They include arterial ligation, embolization of the arterial supply from the external carotid system, electrocoagulation, interstitial irradiation, hormone therapy, cryotherapy, and external irradiation. Anesthetic adjuncts, such as hypotensive techniques and hypothermia, have also been recommended.

The real value of all these is conjectural, because some angiofibromas bleed more than others. Likewise, much of the blood loss associated with angiofibroma surgery comes from the adjacent bone and soft tissues that are manipulated during exposure and that may be hypervascular because of the tumor. Techniques that theoretically alter the vascularity of only the tumor do nothing to the adjacent structures from which much of the bleeding comes.Personal experience with the use of cryosurgery, hormones, hypotensive anesthesia, and embolization reveals the following: cryosurgery is impractical as an adjunct to excision for large tumors and has been abandoned. Hormones are too much of a problem with too little to support their value. I am enthusiastic about hypotensive anesthesia and embolization. An attempt is made to ligate the internal maxillary artery as early as possible during exposure of the tumor. Surgery on large angiofibromas requires courage, rapid and efficient procedures, alert anesthesia personnel, continuing communication between the surgeon and the anesthesia staff, and multiple routes of access for blood replacement. Surgery on large angiofibromas is dangerous, but the danger can be lessened by an experienced team.

Management

There are many approaches for the removal of an angiofibroma.

Surgical management. Neel et al (1973) noted Hellat's 1911 survey of these tumors. Hellat found 65 different techniques used to remove this tumor. Neel et al suggested eight logical approaches: (1) through the nose, (2) through the palate, (3) through the mandible, (4) through the zygoma, (5) through the bed of the hyoid bone, (6) through the antrum, (7) combined craniotomy and rhinotomy, and (8) the lateral rhinotomy.

Of all these approaches, the lateral rhinotomy is preferable because it is logical and relatively uncomplicated.

The lateral rhinotomy is an incision along the side of the nose (Fig. 43-2). What comes after the incision can be any number of a spectrum of procedures. What is actually done depends on the goal of the operation.

The basic rhinotomy incision will only provide exposure to the anterior septum. Exposure beyond that is blocked by the lateral nasal wall. The most useful extension beyond the lateral nasal incision is the block removal of the lateral nasal wall. This has been defined as the "medial maxillectomy" (Sessions and Larson, 1977).

The purpose of rhinotomy-medial maxillectomy is resections of the lateral wall of the nose. The incision with resection of the lateral nasal wall also provides access to the posterior nasal septum, tumors such as angiofibroma that originate more posteriorly in the nasal cavity, the pterygomaxillary space, and the nasopharynx.

After the skin incision is developed down to bone and the facial flap elevated as necessary, the frontal process of the maxilla, including the bone around the nasolacrimal duct, is removed with heavy bone instruments (rongeurs and Kerrison-type instruments). The medial orbital contents are easily reflected laterally by elevation from the lamina papyracea of the ethmoid bone. This reflection exposes the anterior ethmoid artery, which can be divided. The easiest way to divide the artery is to apply a needle-tip cautery in the cautery mode directly to the bony foramen of the vessel and cauterize the artery as it is divided. This approach is quick and has never been hazardous. The ethmoid sinus is entered through the paper plate and the ethmoids exenterated back to the sphenoid sinus. Removal of the paper plate of the ethmoid will free the superior attachment of the lateral nasal wall. The external nasal structures are elevated and detached from the piriform aperture, and the nasal vestibule is incised to expose the anterior turbinates and septum.

If the entire lateral wall of the nose is to be removed, the lacrimal sac is divided. If, as in the case of a small angiofibroma, the inferior turbinate can be preserved, the nasolacrimal duct can be freed from the bone that surrounds it and be saved. Bony cuts are then necessary from front to back to remove the entire lateral nasal (medial maxillary) wall (Fig. 43-3). The first cut divides the orbital floor medial to the infraorbital neurovascular bundle. The second is generally made through the inferior meatus. It can be made above the inferior turbinate if the rhinotomy is only for access. The third cut, if the upper limits were not detached previously, is made high between the orbit and the cribriform plate. This is made by placing a sharp chisel in the bone and driving it posteriorly behind the pterygopalatine fossa and the

vertical plate of the palatine bone running parallel to the cribriform plate. A simple incision of mucosa will allow en-bloc removal of the lateral nasal wall.

Variations of the basic operation are possible. For tumors of the roof of the nose such as the esthesioneuroblastoma, a bilateral rhinotomy called the *superior rhinotomy* elegantly combines with anterior craniotomy for the combined en-bloc resection of the cribriform area. In this procedure (Bernard et al, 1989) the soft tissues and the bony attachments of the nose are divided bilaterally across the nasal root hingeing the nose downward on its remaining alar and columellar attachments. The rhinotomy exposure combined with bilateral medial maxillectomies (resection of the lateral nasal wall) allows en-bloc resection of ethmoid neoplasms with or without intracranial exposure.