PLASTIC SURGERY

VOLUME 5

TUMORS OF THE HEAD & NECK
AND SKIN

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TUMORS OF THE HEAD & NECK AND SKIN

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PLASTIC SURGERY

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Volume 5

Tumors of the Head & Neck and Skin

65

William Y. Hoffman
Daniel C. Baker

Pediatric Tumors of the Head and Neck

The presence of a mass in an infant or child causes great consternation in both physicians and parents. It is important for the surgeon to have a complete understanding of the differential diagnosis in order to reassure the family and to carry out an adequate diagnostic work-up. Although the vast majority of head and neck masses in children are benign (and most of these are inflammatory), malignancies can be devastating, and must be ruled out by appropriate examination and diagnostic studies.

Approximately one-quarter of childhood malignancies are found in the head and neck. The incidence appears to drop slightly with increasing age, being 20 per 100,000 population before the age of 5 years and 10 per 100,000 population in the following ten years.

PREOPERATIVE EVALUATION

Completion of the history taking and physical examination should lead to a diagnosis in most cases. The presence of a mass since birth suggests a benign process, usually of mesodermal origin. A recent respiratory tract infection suggests an inflammatory origin. The rate and duration of growth should also be ascertained.

The age of the patient provides some clues to the diagnosis. Benign mesodermal anomalies—hemangiomas and cystic hygromas—are the most common lesions of the head and neck at birth and in the first year of life. Teratomas and dermoid cysts are also seen in the first year of life, although they are less frequent in the head and neck region than in other anatomic areas. Branchial cleft sinuses may be noted at birth, but cysts of branchial cleft origin, although presumably present at birth, may not present until adolescence. Malignancies are rarely seen in the first year; at this age, leukemias, retinoblastomas, and brain tumors are most common, with lymphomas and sarcomas predominating after age 5 years. Salivary gland tumors are generally seen after age 10 years.

The physical examination also aids in the differentiation of benign and malignant

masses. The findings common to any physical examination should be noted, e.g., size, location, mobility, tenderness, and quality of the overlying skin. The location of a mass is frequently characteristic in certain lesions (these are discussed under the specific lesions below). Transillumination may distinguish between a cystic and a solid mass; the former is almost always benign. Movement of a mass during swallowing may suggest attachment to the thyroid gland or the hyoid bone. Characteristic locations for certain lesions—teratomas in the midline anterior neck, rhabdomyosarcomas in the orbit, ear, or nasopharynx—may suggest a diagnosis.

A careful examination should include direct inspection of the oropharynx, hypopharynx, and nasopharynx. Extremely small caliber endoscopes are available for examination and biopsy of previously inaccessible areas such as the posterior nasopharynx and the larynx. The use of these instruments in smaller children may require general anesthesia. In the presence of lymph node enlargement, axillary and inguinal nodes should also be examined; the abdomen should be examined for masses that might be the primary foci for metastatic nodes.

Biopsy should be considered whenever there is a doubt regarding the possibility of malignancy, as well as in certain benign conditions such as neurofibromatosis or fibrous dysplasia. In addition to standard incisional or excisional biopsies, fine needle aspiration cytology has become established as a minimally invasive technique with high sensitivity and specificity in a variety of lesions. If a malignant tumor is considered a possibility, communication with the pathologist before the biopsy ensures adequate-sized samples and fixation of the specimen (Larson, Robbins, and Butler, 1984). Electron microscopy may be useful in the diagnosis of sarcomas; this study requires glutaraldehyde fixation and should be considered before the time of biopsy (Feldman, 1982).

RADIOGRAPHIC IMAGING

Plain films of the neck and sinuses are particularly useful to visualize the air-filled spaces of the head and neck region. Soft tissue or bony masses may be difficult to interpret on plain films because of the overlying shadows of the craniofacial skeleton. Ultrasound should be considered as a means of differentiating cystic from solid masses. With newer ultrasonographic methods, excellent anatomic definition can also be obtained (Ward-Booth and associates, 1984; Sherman and associates, 1985; Kraus and associates, 1986).

Substantial improvements in the quality of computed tomography (CT) have revolutionized the diagnostic approaches to head and neck masses in children (Russell, 1985). Axial slices can provide information regarding the cystic or solid nature of the mass, the extent of deep involvement or extension, the bony erosion, and the anatomic location of the mass in relation to other structures (Silverman, Korobkin, and Moore, 1983). A high degree of diagnostic accuracy has been reported for CT scanning in cervical infections (Nyberg and associates, 1985), branchial cleft cysts (Salazar, Duke, and Ellis, 1985), salivary gland tumors (Bryan and associates, 1982; Golding, 1982), and other malignancies of the head and neck (Krol and Strong, 1986). Three-dimensional reconstruction of the CT images can be performed; these are quite accurate for examination of skeletal changes, but are less so for soft tissue masses because of the fairly narrow band of CT numbers describing most soft tissue (Fig. 65–1).

Magnetic resonance imaging (MRI) is a new modality that is particularly useful in the definition of soft tissue masses. It does not image bone well, largely because of the decreased blood flow through bone. The indications for MRI as opposed to CT scans are still being defined at this time.

INFLAMMATORY MASSES

Lymphadenopathy

Neck masses of lymphoid origin are a frequent occurrence following upper respiratory tract infections. Although these most commonly result from *Staphylococcus aureus* or beta-hemolytic *Streptococcus* species (80 to 90 per cent), numerous other etiologic agents have been described, including a full spectrum of bacterial and viral organisms (Bedros and Mann, 1981). Most bacterial adenopathy presents with tenderness on palpation but without evidence of overlying skin involvement. If a patient does not respond rapidly to penicillin or first-generation cephalosporin therapy, additional antibiotics should be

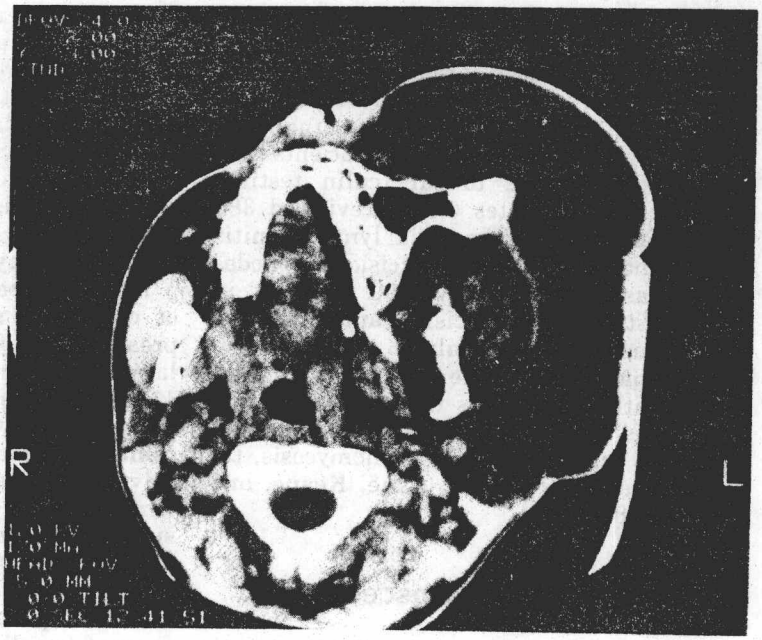

Figure 65–1. Axial CT scan demonstrating an extensive neurofibroma with displacement and hypertrophy of the craniofacial skeleton.

added to treat gram-negative and anaerobic organisms.

Needle aspiration may provide culture material and may also be curative for small abscesses. Ultrasound and computed tomography have both been used to delineate the origin and extent of lymphoid masses, as well as to localize abscess cavities when present.

Excisional biopsy of an enlarged lymph node should be performed when the usual diagnostic maneuvers have been carried out without yield. The reported results of cervical lymph node biopsies in children show that approximately 80 per cent are hyperplastic or granulomatous, 18 per cent represent a lymphoma or other lymphoreticular malignancy, and only 2 per cent are involved with metastatic or primary head and neck disease.

Chronic Cervical Lymphadenitis

Mycobacteria, both tuberculous and atypical, are the most common etiologic agents for chronic suppuration in cervical lymph nodes. Hippocrates mentioned drainage of suppurative neck nodes, and in the Middle Ages scrofula was known as the "King's evil" after several reigning sovereigns claimed to effect cures by touching the patients. In recent years there has been an increased incidence of this relatively unusual disease, with the influx of immigrants from endemic areas in Asia and South America. Chronic granulomatous disease in the neck typically presents in children under 6 years old, with painless progressive enlargement of nodes that ultimately form "cold" abscesses, which may mimic neoplasms (Levin-Epstein and Lucente, 1982).

Tuberculous cervical adenitis (scrofula) occurs in approximately 5 per cent of all cases of tuberculosis, but in up to two-thirds of cases of extrapulmonary disease (Cantrell, Jensen, and Reid, 1975). Pathologic study reveals a range of findings from simple hyperplasia to caseating necrosis, and organisms are seen in most cases. Approximately one-half of patients with tuberculous neck nodes have systemic symptoms, although the chest radiograph and sputum smear may be negative. Skin tests are diagnostic in virtually all cases. Treatment should consist of surgical excision of the involved nodes, as simple incision and drainage frequently results in draining fistulas; patients should concomitantly receive long-term antituberculous chemotherapy (Appling and Miller, 1981; Castro, Hoover, and Zuckerbraun, 1985).

Atypical mycobacteria may present more diagnostic difficulty, because the chest radio-

graph is often normal and the involved nodes usually unilateral. The most common organisms isolated in large series are *M. scrofulaceum* and *M. avium-intracellulare* (Saitz, 1981). Specific skin tests for these organisms exist but are difficult to obtain; patients may be weakly positive to tuberculin testing. Schaad and associates (1979) reviewed 380 cases of atypical mycobacterial lymphadenitis and found that surgical excision of nodes alone was curative in over 90 per cent, in contradistinction to incision and drainage, which was effective in only 16 per cent. Medical management alone effected no cures. Other pathologic states that have been described as causing similar presentations include cat scratch disease, actinomycosis, tularemia, and sarcoidosis (Lane, Keane, and Potsic, 1980).

Lateral Pharyngeal Abscesses

Suppuration of an untreated deep lymph node or extension of a peritonsillar abscess may lead to the development of a lateral pharyngeal abscess, which may represent a surgical emergency. The abscesses present as fullness in the lateral cervical region, with tenderness and dysphagia as early signs. Progression of the infectious process may lead to trismus, hoarseness, cough, and dyspnea; in the advanced stage, there may be progression to sepsis, airway obstruction, and possible carotid artery erosion.

A plain lateral radiograph of the neck may demonstrate soft tissue swelling in the posterior pharyngeal space. As noted above, ultrasound and computed tomography further delineate the abscess cavity, and may guide percutaneous drainage if the cavity is accessible. Wide surgical drainage is mandatory if the later complications of sepsis or airway obstruction have occurred; tracheotomy may also be required.

Sialadenitis

Inflammation accounts for more than one-third of the pathologic conditions of the salivary glands in the pediatric patient. Regardless of whether the submaxillary or parotid gland is affected, the clinical presentation is similar, with a tender, swollen gland. Acute parotitis is seen most commonly in premature infants and children with systemic illness; it is frequently associated with dehydration, fever, or immunosuppression. Postoperative parotitis is rarely seen in children, although it is probably the most common form of salivary gland inflammation seen in adults. Possible causes include inspissation of mucus and retrograde infection, as well as congenital sialoangiectasis. *S. aureus* is the most common organism found. Therapy consists of antibiotics and hydration.

Recurrent parotitis is primarily a disease of childhood, seen most frequently in the preadolescent years. The etiology may be related to sialoangiectasis, which was found in 11 of 16 patients in one series (David and O'Connel, 1970). *Streptococcus* is the most common offending organism. Most cases resolve spontaneously in the teenage years; hence, therapy is generally symptomatic, consisting of hydration, antibiotics, and occasionally duct dilation and drainage. Parotidectomy is rarely required.

Chronic parotitis is similar to the recurrent form of the disease in its periodic presentation, but does not display clinical signs of infection. Autoimmune disease and allergy have been implicated as etiologic factors. Symptomatic therapy consists of local heat and massage, as well as control of any underlying systemic disease.

Unlike inflammation of the parotid, acute submaxillary gland infection is associated with stones or congenital strictures. Generally, removal of the calculi is inadequate and resection of the involved gland is required (Kaban, Mulliken, and Murray, 1978).

DEVELOPMENTAL ANOMALIES

Branchial Cleft Anomalies

Branchial cleft anomalies include branchiogenic sinuses, cartilaginous rests, fistulas, and cervical cysts. Between the third and sixth weeks of embryonic development, the neck develops four clefts and four corresponding pharyngeal pouches, which are separated by a membrane, the branchial plate (see Chaps. 46 and 62). Fistulas form when the branchial clefts fail to close completely, with rupture of the branchial plate (Fig. 65–2). A complete fistulous tract may form, one end may close with the formation of a blind sinus tract, or both ends may close with a cyst resulting from the preservation of a central cellular rest. The sinuses and cysts that re-

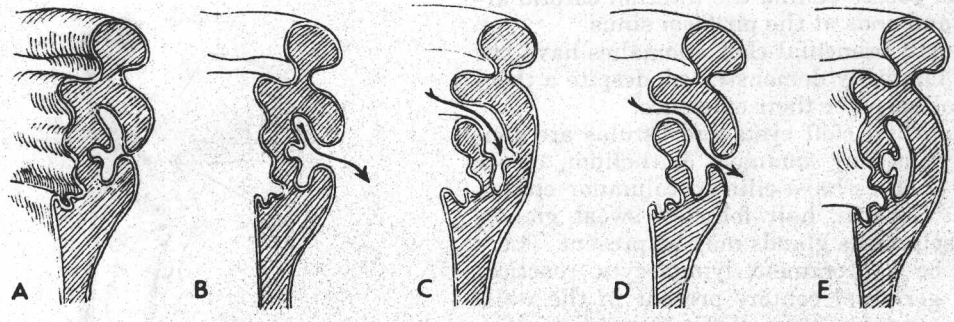

Figure 65–2. Series of diagrams to show how several kinds of tracts and cysts may arise through faulty development of the pharyngeal wall. *A*, Normal pharynx showing closure of the cervical sinus. *B*, Incomplete closure of the cervical sinus forming the basis for a tract opening externally upon the surface of the neck. *C*, Rupture of the closing membrane leaving a permanent opening into the position of the second pouch. *D*, Branchial fistula resulting from a combination of the conditions in *B* and *C*. *E*, Cystic remnant of the cervical sinus. (Adapted with permission from Ward, G. E., and Hendrick, J. W.: Diagnosis and Treatment of Tumors of the Head and Neck. Copyright 1950. The Williams & Wilkins Company, Baltimore.)

main in communication with the pharynx are particularly prone to infection and may present with cellulitis and abscess formation.

Branchial cleft cysts typically present as a smooth, nontender mass, lying along the anterior border of the sternocleidomastoid muscle between the external auditory canal and the clavicle. There is frequently a history of waxing and waning size associated with upper respiratory infections. Fistulas, in contrast, are usually palpable as fibrous cords; the external opening may be extremely small and may be associated with skin tags or cartilaginous remnants. The cutaneous orifice of the fistula retracts with swallowing because of the connection to the pharyngeal wall.

First branchial cleft anomalies are uncommon. They are always above the level of the hyoid bone; the external orifice is usually found in the vicinity of the auricle or beneath the mandibular ramus. The fistulas may traverse the parotid gland and are variably situated in relationship to the branches of the facial nerve (Olsen, Maragos, and Weiland, 1980; Liston, 1982; Al Fallouji and Butler, 1983).

Second branchial cleft anomalies are the most common (Fig. 65–3), and are usually found near the junction of the middle and lower thirds of the sternocleidomastoid muscle. Fistulas characteristically follow the carotid sheath, crossing the hypoglossal nerve and passing between the internal and external carotid arteries (near the bifurcation of the common carotid artery) to reach the tonsillar fossa (Salazar, Duke, and Ellis, 1985).

Third branchial cleft anomalies are rare. The external orifice may be located in a similar manner to the second branchial cleft fistulas, along the anterior border of the lower half of the sternocleidomastoid muscle; the

Figure 65–3. Branchial cleft (second) cyst. Note the external orifice along the anterior aspect of the sternocleidomastoid muscle.

fistula passes behind the internal carotid artery and ends at the piriform sinus.

Fourth branchial cleft anomalies have not been clinically demonstrated, despite a theoretical basis for their existence.

Branchial cleft cysts and fistulas are generally lined by squamous epithelium; 10 per cent of cases have ciliated columnar epithelium. Keratin, hair follicles, sweat glands, and sebaceous glands may be present. There may be a prominent lymphocytic reaction, with germinal centers present in the walls that react to upper respiratory tract infections. Fine needle aspiration cytology has been reported as a means of diagnosis of branchial cleft cysts (Ramos-Gabatin and Watzinger, 1984). Ultrasound and CT scanning may also be of assistance (Byrd and associates, 1983; Salazar, Duke, and Ellis, 1985).

The variable age at onset mandates a thorough work-up of presumed branchial cleft anomalies. Cinberg and associates (1982) reported that four of 18 adult patients, ranging in age from 37 to 74 years and diagnosed as having simple cysts, proved to have metastatic carcinoma with unknown primaries. A few cases of branchial cleft carcinoma have been reported (Jablokow, Kathuria, and Wang, 1982; Shreedhar and Tooley, 1984), since Martin's initial report (Martin, Morfit, and Ehrlich, 1950) and the establishment of criteria for the diagnosis of branchiogenic carcinoma.

Therapy consists of total surgical extirpation of the entire fistulous tract and any coexistent cyst. Superficial skin tags and cartilaginous rests can be removed easily under local anesthesia in the older child. Removal of a branchial cleft cyst and/or fistula requires general anesthesia, with preparation for significant dissection of the facial nerve in first branchial cleft anomalies, and of the neck vessels in second and third branchial cleft anomalies.

Thyroglossal Duct Remnants

The thyroid gland develops from the thyroglossal duct, extending from the foramen cecum in the posterior midline of the tongue through the hyoid bone to the midline of the lower neck (Fig. 65–4). Persistence of the thyroglossal duct may result in cysts, fistulas, and thyroid gland remnants along its course.

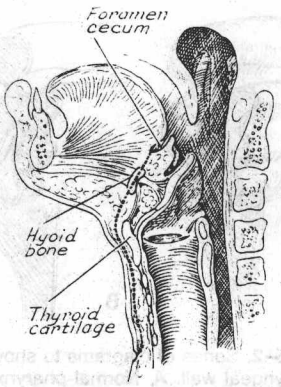

Figure 65–4. Thyroglossal duct sinus and/or fistula. The dotted line traces the pathway from the foramen cecum at the tongue through the hyoid bone, in front of the thyroid cartilage, and onto the skin in the midline of the neck. (Adapted from Marcus, E., and Zimmerman, L. M.: Principles of Surgical Practice. Copyright © 1960 by McGraw-Hill Book Company, Inc.)

Clinical presentation usually consists of a round cystic mass just below the hyoid bone in the midline of the neck. The lesions are usually asymptomatic despite their fairly large sizes. The cysts move with swallowing. Small cysts without infection may be observed; larger cysts, which cause cosmetic deformity or chronic drainage, should be excised. Extirpation includes the cyst, the body of the hyoid bone, and the fistulous tract up to the foramen cecum (Fig. 65–5).

Laryngocele

A laryngocele is an air-filled cyst arising from the laryngeal ventricle between the true and false vocal cords. If confined to the larynx, it is known as an internal laryngocele and may cause stridor and a weakened voice; an external laryngocele extends through the thyrohyoid membrane to present as a nontender, compressible mass that increases in size with a Valsalva maneuver. An air-filled sac may be demonstrated on lateral radiography or CT scan. A laryngocele presenting near the sternocleidomastoid muscle may be confused with a branchial cleft cyst, but the association with the thyrohyoid membrane and the absence of fistulous tract confirm the diagnosis. Surgical excision of the lesion is the only therapy for larger, symptomatic lesions; a lateral cervical approach is recommended. Smaller lesions may be observed (Baker, Baker, and McClatchey, 1982).

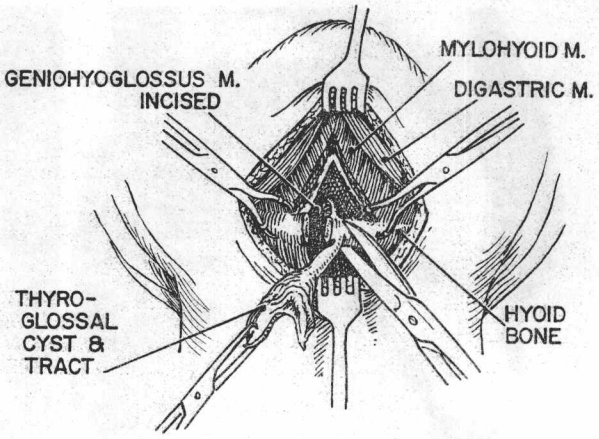

Figure 65–5. Dissection of the thyroglossal cyst and tract.

Teratomas and Dermoids

Teratomas are rare congenital tumors that occur in one in 4000 births. There is a male-to-female ratio of 1:6. The tumors are most commonly in the sacrococcygeal area and the ovaries. Less than 10 per cent are found in the head and neck region; one review found only 14 head and neck teratomas out of a total series of 245 (Tapper and Lack, 1983).

Teratomas contain elements from all three germ cell layers, often in different stages of maturation. Rudimentary organ formation may be seen. Head and neck teratomas (Fig. 65–6) have a preponderance of neurogenic tissue (Batsakis, 1984). Although these are generally tumors of newborns, teratomas may also occur in older patients and are associated with a higher incidence of malignancy with increasing age. In one series, six of nine patients over 15 years of age with teratomas showed evidence of malignant degeneration (Watanatittan, Othersen, and Hughson, 1981). Survival is directly related to the ability to extirpate the tumor in the first six months of life (Stone, Henderson, and Guidio, 1967; Tapper and Lack, 1983).

In the neck, teratomas often arise within or adjacent to the thyroid gland and cause compression of neck structures. Polyhydramnios may be noted, owing to the inability of the fetus to swallow amniotic fluid; this finding is associated with an increased incidence of stillbirth and premature births (Hajdu and associates, 1966). Neck teratomas frequently present with respiratory distress at birth; the mortality rate in unoperated cases may be as high as 80 per cent (Stone, Henderson, and Guidio, 1967). Early surgery has reduced the mortality rate to 15 per cent (Abemayor and associates, 1984; Gundry and associates, 1983). Other head and neck sites include the orbit, the midline of the nose, and the sinuses. Teratoid tumors, or hairy polyps, are most commonly found in the nasopharynx as polyps with a long, relatively avascular stalk; they are easily snared for diagnosis and cure.

Dermoid cysts contain epithelium and adnexal structures. They are typically found in the lateral brow and in the midline of the nose (Fig. 65–7) or neck. Large lesions with intraorbital extension may cause exophthalmos. Midline nasal dermoids may present as a small pit near the radix but may extend into the septum, cribriform plate, and dura, and should therefore be investigated with CT scanning before surgery. Dermoid cysts of the neck have presented with large masses causing respiratory obstruction soon after birth, requiring early surgical intervention. Extirpation of the lesions may require a craniofacial approach in order to remove the skeletal extension of the cysts.

BENIGN TUMORS

Neurofibromas and Related Tumors

Neurofibromas are benign growths of multiple cellular elements of peripheral nerves. They may occur in isolation in the head and neck, or as multiple lesions, as in von Recklinghausen's disease (neurofibromatosis). The latter is seen more commonly in children; isolated tumors occur more often in adults.

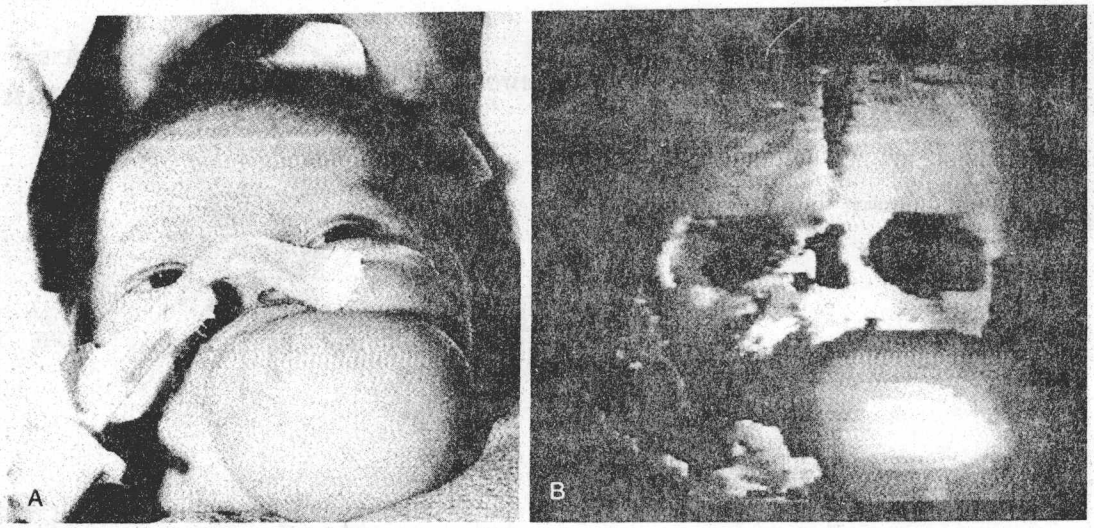

Figure 65–6. Teratoma arising from the palate (extracranial) in a newborn. *A*, Preoperative view. *B*, Three-dimensional CT scan showing that the lesion is extracranial. (Patient of Dr. Joseph G. McCarthy, New York University.)

Von Recklinghausen's (1882) disease is an autosomal dominant disorder of unknown etiology characterized by multiple neural sheath tumors (Fig. 65–8), café au lait spots, and bone lesions. Griffith and associates (1972) reviewed 50 cases of neurofibromatosis and found that 92 per cent of patients had tumors, 68 per cent café au lait spots, and 46 per cent a positive family history. Thirty-three per cent had bony abnormalities, including 25 per cent with scoliosis; 42 per cent had intracranial anomalies, and 16 per cent were mentally retarded. Skull and facial bone deformities may be seen on plain films or on CT scan, with osteolytic lesions observed in the mandible and skull even in the absence of tumor. An associated anomaly of the facial skeleton is aplasia of the greater wing of the sphenoid bone (Fig. 65–9), leading to pulsatile exophthalmos; this may occur in the absence of tumor (Gupta and associates, 1979).

Neurofibromatosis follows a characteristic course of steady growth in childhood, with the appearance of multiple subcutaneous and cutaneous nodules; Maceri and Saxon (1984) found that somewhat more than one-third of patients had head and neck involvement. Plexiform lesions are locally invasive in soft tissue and may cause significant deformity (Crikelair and Cosman, 1968). Malignant degeneration of tumors occurs in 5 to 15 per cent of patients, manifested by symptoms of sudden increase in tumor size and severe localized pain.

Treatment consists of surgical resection, either total or subtotal, and depends on the age of the patient, the location and growth rate of the tumor, and the degree of functional or cosmetic deformity. Multiple staged debulking procedures are commonly performed, usually with limited success. The tumors tend to be unencapsulated, and dissection can be hampered by an intimate association of the

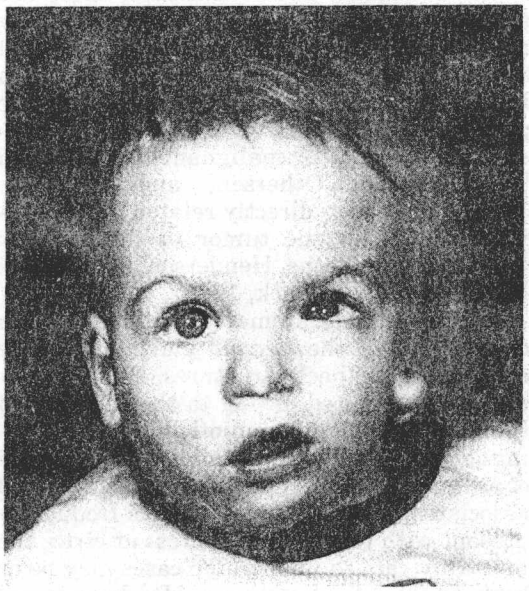

Figure 65–7. Dermoid of the midline of the nasal tip. (Patient of Dr. Joseph G. McCarthy, New York University.)

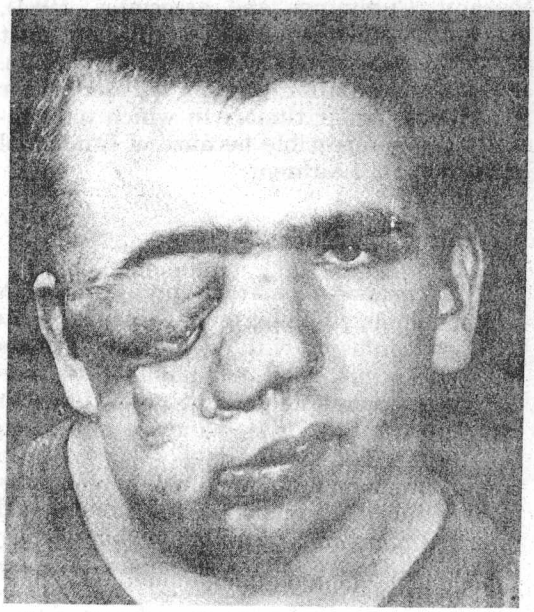

Figure 65–8. Von Recklinghausen's disease in an adolescent male characterized by multiple neural sheath tumors and pachydermatous involvement of the orbital, nasal, and cheek structures. He also has café au lait spots.

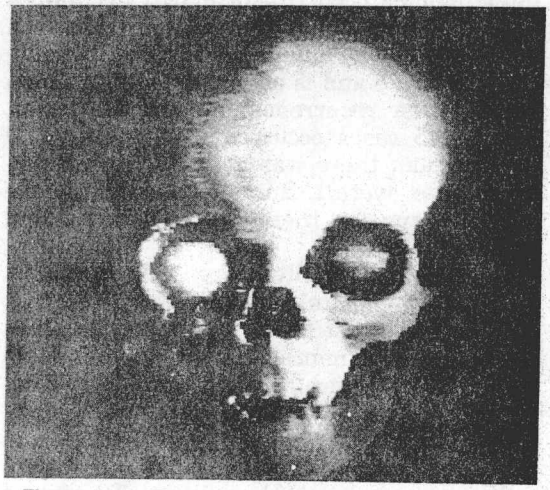

Figure 65–9. Three-dimensional CT scan demonstrating aplasia of the greater wing of the sphenoid bone with exophthalmos and displacement of the globe in a patient with neurofibromatosis. (Courtesy of Dr. Joseph G. McCarthy, New York University.)

tumor with the facial nerves and muscles. In severe deformities, consideration may be given to radical resection of all involved skin and subcutaneous tissue, and resurfacing with a skin graft or thin cutaneous free tissue transfer (Adekeye, Abiose, and Ord, 1984). The typical orbital deformity is treated by a craniofacial surgical approach, with a subtotal tumor resection and bone grafting of the roof of the orbit, and expansion of the orbital volume to accommodate the tumor mass (Marchac, 1984) (see Chap. 33).

Benign Lipomatous Tumors

Infantile Lipoblastomatosis (Chung and Enzinger, 1973). Lipoblastomatosis refers to tumors made up of immature or embryonal fat cells. The myxoid stroma seen histologically may be confused with myxoid or well-differentiated liposarcoma; the benign tumor has few mitoses and no evidence of atypia. Most are well-circumscribed growths resem-

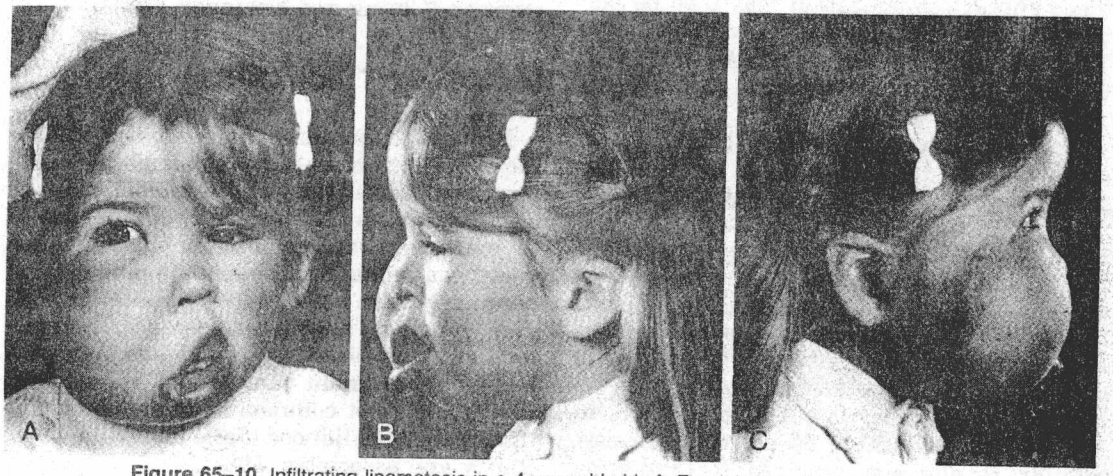

Figure 65–10. Infiltrating lipomatosis in a 4 year old girl. *A*, Frontal view. *B*, *C*, lateral views.

bling lipomas, occurring primarily in children under 10 years of age. The diffuse form, seen in approximately one-third of patients, is locally invasive and is associated with a somewhat higher recurrence rate. In one series, four of 35 cases occurred in the head and neck region; there was a 14 per cent recurrence rate overall. Surgical excision is the only therapy for these lesions (Chung and Enzinger, 1973).

Congenital Infiltrating Lipomatosis. This is a rare clinicopathologic entity (Fig. 65–10) that is distinct from common lipomata in that it is unencapsulated and tends to infiltrate local tissue. Histologic study demonstrates mature lipocytes that do not display the proliferative changes, pleomorphism, and mitoses of liposarcoma. They may mimic neurofibromatosis or lymphangiomas. Treatment consists of aggressive early surgical excision; as with neurofibromas, subtotal resection may be the only alternative in the attempt to balance preservation of function and cosmesis (Mattel and Persky, 1983; Slavin and associates, 1983).

Angiofibromas

Angiofibromas are benign vascular tumors that most commonly present in the nasopharynx in adolescent males. Three-quarters of patients present with severe or recurrent epistaxis; two-thirds have nasal obstruction. The tumors tend to invade locally, growing into the pterygomaxillary fossa and the paranasal sinuses. CT scanning is useful in defining the extent of the lesion. Arteriography confirms the diagnosis and identifies the blood supply; embolization may also be performed to reduce intraoperative blood loss. There have been case reports of malignant transformation to fibrosarcoma (Witt, Shah, and Sternberg, 1983).

Surgical resection is the treatment of choice, since natural regression of these neoplasms has not been demonstrated. A lateral rhinotomy or transpalatine approach has been recommended. Twenty per cent of the tumors have intracranial extensions and require a combined neurosurgical and craniofacial approach for resection. Operative blood loss may be considerable, averaging over 2500 ml in one series (Witt, Shah, and Sternberg, 1983). Surgery is effective in eradicating the tumor in 80 to 85 per cent of cases. Similar results are reported for radiation therapy (Cummings and associates, 1984), but the possible sequelae of radiation must be considered, especially in younger patients. In general, radiation therapy is reserved for recurrences or for tumors in which total resection is not possible because of functional or cosmetic impairment.

Nodular Fasciitis

This pseudosarcomatous proliferation of soft tissue is of importance because of its frequent confusion with fibrosarcoma. In the head and neck region, nodular fasciitis most commonly occurs deep in the soft tissue, in close proximity to the mandible and along the sternocleidomastoid muscle. It may represent an anomalous reparative process after injury. Surgical excision of the tumors with minimal margins and without other therapy has been curative in most cases (Dahl and Jarlstedt, 1980).

Infantile Myofibromatosis

This form of myofibromatosis occurs almost exclusively in children under 2 years of age; 60 per cent of cases are noted in the first month. Its name derives from the histologic findings of cells that have an appearance between fibroblasts and smooth muscle. Three-quarters are solitary tumors, and 69 per cent occur in males; a multicentric type is also described, which may involve bone and viscera as well as soft tissue. The latter carries a 20 per cent mortality rate from visceral (pulmonary) involvement; the solitary type generally does well after only limited surgical excision (Chung and Enzinger, 1981).

MALIGNANT TUMORS

Malignant tumors are rare in children. The incidence of various tumors changes with the age of the population. The first year of life is associated with a particularly low incidence of malignant neoplasms. In the first five years, leukemia, retinoblastoma, and central nervous system tumors predominate. As children progress toward adolescence, lymphomas and soft tissue sarcomas are the more common malignant tumors and, in fact, represent the most common head and neck malignancies in childhood (Jaffe and Jaffe, 1973; Raney and associates, 1981).