

Soft tissue tumors

LIPOMA

The **lipoma** is a benign tumor of fat. The pathogenesis of lipomas is uncertain, but they appear to be more common in obese people.

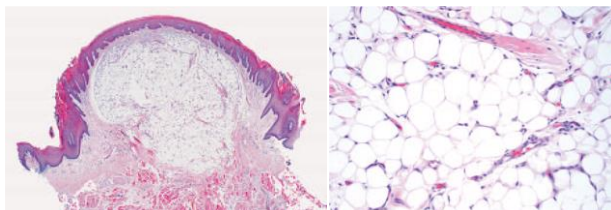
CLINICAL FEATURES

Oral lipomas are usually soft, smooth-surfaced nodular masses that can be sessile or pedunculated. Typically, the tumor is asymptomatic and often has been noted for many months or years before diagnosis. Most are less than 3 cm in size, but occasional lesions can become much larger. Although a subtle or more obvious yellow hue often is detected clinically, deeper examples may appear pink. The buccal mucosa and buccal vestibule are the most common intraoral sites and account for 50% of all cases. Most patients are 40 years of age or older; lipomas are uncommon in children.



HISTOPATHOLOGIC FEATURES

Most oral lipomas are composed of mature fat cells that differ little in microscopic appearance from the surrounding normal fat. The tumor is usually well circumscribed and may demonstrate a thin fibrous capsule. A distinct lobular arrangement of the cells often is seen.



TREATMENT

Lipomas are treated by conservative local excision, and recurrence is rare.

LYMPHANGIOMA

Lymphangiomas are benign, hamartomatous tumors of lymphatic vessels. It is doubtful that they are true neoplasms; instead, they most likely represent developmental malformations

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that arise from sequestrations of lymphatic tissue that do not communicate normally with the rest of the lymphatic system. There are three types of lymphangioma:

1. **Lymphangioma simplex (capillary lymphangioma)**, which consists of small, capillary-sized vessels
2. **Cavernous lymphangioma**, which is composed of larger, dilated lymphatic vessels
3. **Cystic lymphangioma (cystic hygroma)**, which exhibits large, macroscopic cystic spaces

CLINICAL FEATURES

Lymphangiomas have a marked predilection for the head and neck, which accounts for 50% to 75% of all cases . Oral lymphangiomas may occur at various sites but are most frequent on the anterior two thirds of the tongue, where they often result in macroglossia . Usually, the tumor is superficial in location and demonstrates a pebbly surface that resembles a cluster of translucent vesicles. The surface has been likened to the appearance of frog eggs.

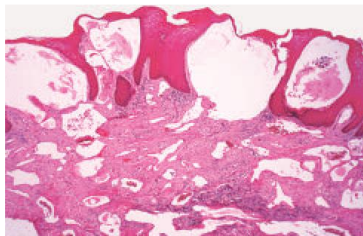


Secondary hemorrhage into the lymphatic spaces may cause some of these “vesicles” to become purple

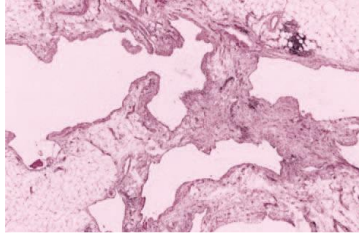


HISTOPATHOLOGIC FEATURES

Lymphangiomas are composed of lymphatic vessels that may show marked dilatation (cavernous lymphangioma)



or macroscopic cyst like structures (cystic hygroma)



The vessels often diffusely infiltrate the adjacent soft tissues and may demonstrate lymphoid aggregates in their walls. The lining endothelium is typically thin. In intraoral tumors, the lymphatic vessels are characteristically located just beneath the epithelial surface and often replace the connective tissue papillae. This superficial location results in the translucent, vesicle-like clinical appearance.

TREATMENT

The treatment of lymphangiomas usually consists of surgical excision, Recurrence is common, especially for cavernous lymphangiomas of the oral cavity, because of their infiltrative nature.

HEMANGIOMA AND VASCULAR MALFORMATIONS

hemangiomas are considered to be benign tumors of infancy that display a rapid growth phase with endothelial cell proliferation, followed by gradual involution. Most hemangiomas cannot be recognized at birth, but arise subsequently during the first 8 weeks of life.

On the other hand, **vascular malformations** are structural anomalies of blood vessels without endothelial proliferation. By definition, vascular malformations are present at birth and persist throughout life.

CLINICAL AND RADIOGRAPHIC FEATURES

HEMANGIOMA

Hemangiomas are the most common tumors of infancy, occurring in 5% to 10% of 1-year-old children. They are much more common in females than in males (ratio: 3:1 to 5:1), The most common location is the head and neck, which accounts for 60% of all cases. Superficial tumors of the skin appear raised and bosselated with a bright-red color ("strawberry" hemangioma)



They are firm and rubbery to palpation, and the blood cannot be evacuated by applying pressure. About half of all hemangiomas will show complete resolution by 5 years of age, with 90% resolving by age 9.

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VASCULAR MALFORMATIONS

In contrast to hemangiomas, vascular malformations are present at birth and persist throughout life. Port wine stains are relatively common capillary malformations that occur in 0.3% to 1.0% of newborns.

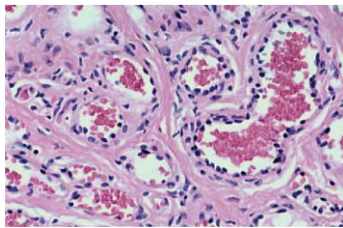


They are most common on the face, particularly along the distribution of the trigeminal nerve. Port wine stains are typically pink or purple macular lesions that grow commensurately with the patient. As the patient gets older, the lesion often darkens and becomes nodular because of vascular ectasia. Typically, venous malformations are blue and are easily compressible



HISTOPATHOLOGIC FEATURES

Congenital hemangiomas are composed of abundant capillary spaces lined by endothelium without muscular support. Congenital vascular malformation may consist not only of capillaries, but also of venous, arteriolar and lymphatic channels.



TREATMENT

- Congenital hemangioma → spontaneous involution during early childhood if not → surgery, arterial embolization, and sclerosant therapy and laser therapy.
- Congenital vascular malformation → the same → difficult to eradicate.

TRAUMATIC NEUROMA (AMPUTATION NEUROMA)

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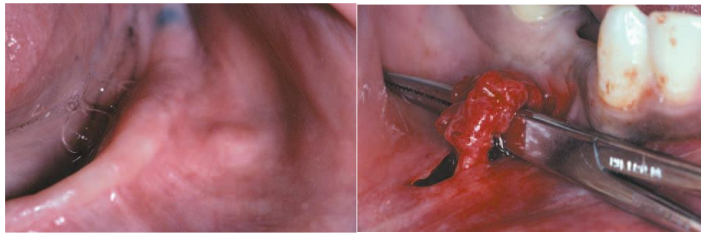
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The **traumatic neuroma** is not a true neoplasm but a reactive proliferation of neural tissue after transection or other damage of a nerve bundle. After a nerve has been damaged or severed, the proximal portion attempts to regenerate and reestablish innervation of the distal segment by the growth of axons through tubes of proliferating Schwann cells. If these regenerating elements encounter scar tissue or otherwise cannot reestablish innervation, then a tumor like mass may develop at the site of injury.

CLINICAL AND RADIOGRAPHIC FEATURES

Traumatic neuromas of the oral mucosa are typically smooth-surfaced, nonulcerated nodules. They can develop at any location but are most common in the mental foramen area, tongue, and lower lip. A history of trauma often can be elicited; some lesions arise subsequent to tooth extraction or other surgical procedures. Intraosseous traumatic neuromas may demonstrate a radiolucent defect on oral radiographs.

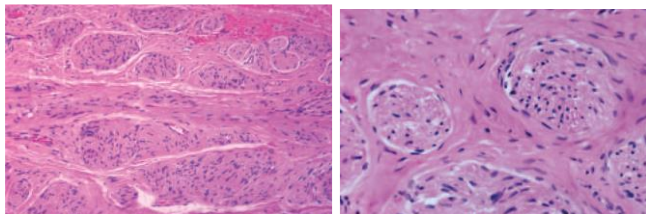
Traumatic neuromas can occur at any age, but they are diagnosed most often in middle-aged adults. They appear to be slightly more common in women. Many traumatic neuromas are associated with altered nerve sensations that can range from anesthesia to dysesthesia to overt pain. pain has been traditionally considered a hallmark of this lesion.



HISTOPATHOLOGIC FEATURES

Microscopic examination of traumatic neuromas shows a haphazard proliferation of mature, myelinated and unmyelinated nerve bundles within a fibrous connective tissue stroma that ranges from densely collagenized to myxomatous in nature.

An associated mild chronic inflammatory cell infiltrate may be present.



TREATMENT

Surgical excision, including a small portion of the involved nerve bundle.

NEURILEMOMA (SCHWANNOMA)

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The **neurilemoma** is a benign neural neoplasm of Schwann cell origin. It is relatively uncommon, although 25% to 48% of all cases occur in the head and neck region.

CLINICAL AND RADIOGRAPHIC FEATURES

The solitary neurilemoma is a slow-growing, encapsulated tumor that typically arises in association with a nerve trunk. As it grows, it pushes the nerve aside. Usually, the mass is asymptomatic. The lesion is most common in young and middle-aged adults and can range from a few millimeters to several centimeters in size. The tongue is the most common location for oral neurilemmomas. On occasion, the tumor may produce bony expansion. Intraosseous examples are most common in the posterior mandible and usually appear as either unilocular or multilocular radiolucencies on radiographs.



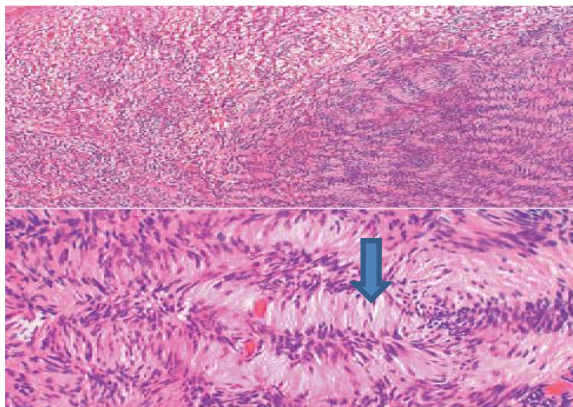
HISTOPATHOLOGIC FEATURES

The neurilemoma is usually an encapsulated tumor that demonstrates two microscopic patterns in varying amounts:

(1) **Antoni A**

(2) **Antoni B.**

Streaming fascicles of spindle-shaped Schwann cells characterize Antoni A tissue. These cells often form a palisaded arrangement around central a cellular, eosinophilic areas known as **Verocay bodies**. These Verocay bodies consist of reduplicated basement membrane and cytoplasmic processes. Antoni B tissue is less cellular and less organized; the spindle cells are randomly arranged within a loose, myxomatous stroma.



TREATMENT

By surgical excision, and the lesion should not recur.

NEUROFIBROMA

The **neurofibroma** is the most common type of peripheral nerve neoplasm. It arises from a mixture of cell types, including Schwann cells and perineural fibroblasts.

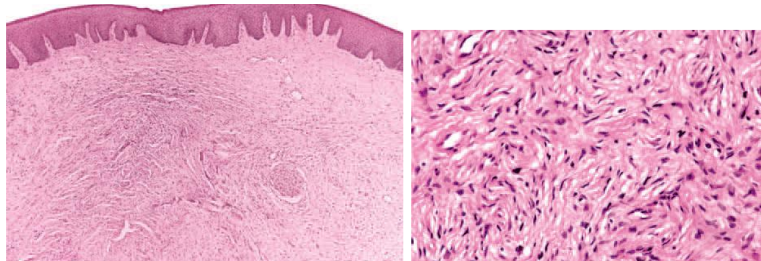
CLINICAL AND RADIOGRAPHIC FEATURES

Most common in young adults and present as slow-growing, soft, painless lesions that vary in size from small nodules to larger masses. The tongue and buccal mucosa are the most common intraoral sites. On rare occasions, the tumor can arise centrally within bone, where it may produce a well-demarcated or poorly defined unilocular or multilocular radiolucency .



HISTOPATHOLOGIC FEATURES

The tumor is composed of interlacing bundles of spindle-shaped cells that often exhibit wavy nuclei . These cells are associated with delicate collagen bundles and variable amounts of myxoid matrix. Mast cells tend to be numerous and can be a helpful diagnostic feature.



TREATMENT

Local surgical excision, and recurrence is rare.

LEIOMYOMA

Leiomyomas are benign tumors of smooth muscle . Leiomyomas of the oral cavity are rare. The three types are as follows:

1. Solid leiomyomas
2. Vascular leiomyomas (angiomyomas or angioleiomyomas)
3. Epithelioid leiomyomas (leiomyoblastomas)

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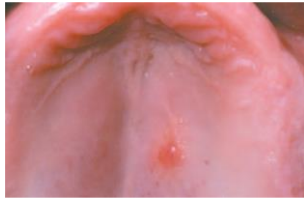
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Almost all oral leiomyomas are either solid or vascular in type; angiomyomas account for nearly 75% of all oral cases.

CLINICAL AND RADIOGRAPHIC FEATURES

The oral leiomyoma can occur at any age and is usually a slow-growing, firm, mucosal nodule. Most lesions are asymptomatic, although occasional tumors can be painful. Solid leiomyomas are typically normal in color, although angiomyomas may exhibit a bluish hue. The most common sites are the lips, tongue, palate, and cheek, which together account for 80% of cases.

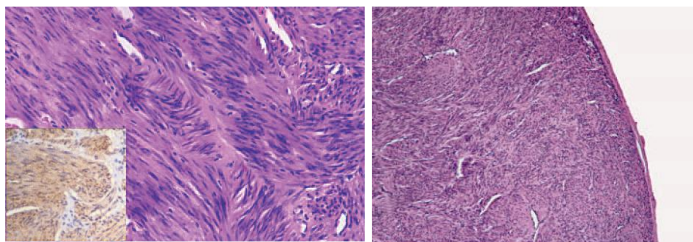


HISTOPATHOLOGIC FEATURES

Solid leiomyomas are well-circumscribed tumors that consist of interlacing bundles of spindle-shaped smooth muscle cells. The nuclei are elongated, pale staining, and blunt ended.

Angiomyomas also are well circumscribed lesions that demonstrate multiple tortuous blood vessels with thickened walls caused by hyperplasia of their smooth muscle coats.

As its name implies, the **epithelioid leiomyoma** is composed primarily of epithelioid cells rather than spindle cells.



TREATMENT AND PROGNOSIS

Local surgical excision. The lesion should not recur.

RHABDOMYOMA

Benign neoplasms of skeletal muscle are called **rhabdomyomas**. Rhabdomyomas of the head and neck can be subclassified into two major categories:

- (1) adult rhabdomyomas
- (2) fetal rhabdomyomas.

CLINICAL FEATURES

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ADULT RHABDOMYOMA

Occur primarily in middle-aged and older patients, with about 70% of cases found in men. The most frequent sites are the pharynx, oral cavity, and larynx; intraoral lesions are most common in the floor of the mouth, soft palate, and base of tongue. The tumor appears as a nodule or mass that can grow to many centimeters before discovery .



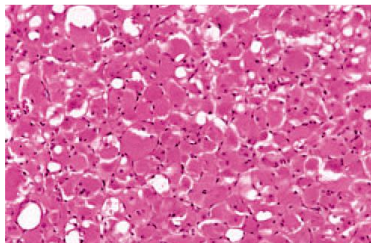
FETAL RHABDOMYOMAS

Usually occur in young children, although some also develop in adults. A similar male predilection is noted. The most common locations are the face and periauricular region.

HISTOPATHOLOGIC FEATURES

ADULT RHABDOMYOMAS

The adult rhabdomyoma is composed of well-circumscribed lobules of large, polygonal cells, which exhibit abundant granular, eosinophilic cytoplasm . These cells often demonstrate peripheral vacuolization that results in a “spider web” appearance of the cytoplasm.



FETAL RHABDOMYOMAS

Has a less mature appearance and consists of a haphazard arrangement of spindle-shaped muscle cells that sometimes are found within a myxoid stroma.

TREATMENT

Surgical excision. Recurrence is uncommon.

Soft Tissue Sarcomas

LIPOSARCOMA

The **liposarcoma** is a malignant neoplasm of fatty origin. It currently is considered to be the most common soft tissue sarcoma and accounts for 20% of all soft tissue malignancies in adults.

CLINICAL FEATURES

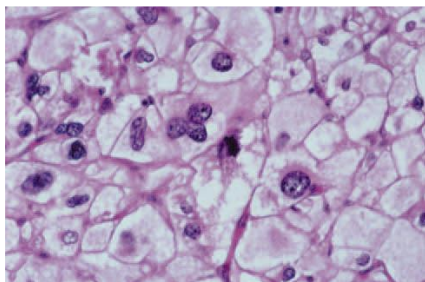
Liposarcomas are primarily seen in adults, with peak prevalence between the ages of 40 and 60. The tumor is typically a soft, slow-growing, ill-defined mass that may appear normal in color or yellow. The most frequent oral locations are the tongue and cheek.

HISTOPATHOLOGIC FEATURES

Most liposarcomas can be divided into three major categories:

1. Well-differentiated liposarcoma/atypical lipomatous tumor
2. Myxoid/round cell liposarcoma
3. Pleomorphic liposarcoma

The most common of these variants in the oral cavity is the **well-differentiated liposarcoma**, which accounts for 55% to 90% of all cases. These tumors resemble benign lipomas but demonstrate scattered lipoblasts and atypical, hyperchromatic stromal cells .



TREATMENT

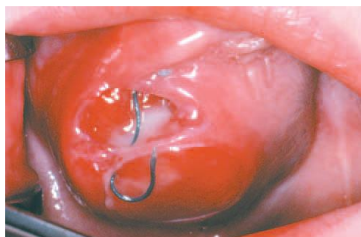
Radical excision is the treatment of choice.

FIBROSARCOMA

The **fibrosarcoma** is a malignant tumor of fibroblasts. The tumor is most common in the extremities; only 10% occur in the head and neck region.

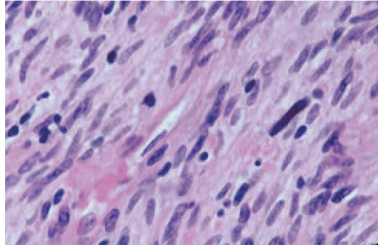
CLINICAL FEATURES

Fibrosarcomas most often present as slow-growing masses that may reach considerable size before they produce pain . They can occur anywhere in the head and neck region. They can occur at any age but are most common in young adults and children.



HISTOPATHOLOGIC FEATURES

Well-differentiated fibrosarcomas consist of fascicles of spindle-shaped cells that classically form a “herringbone” pattern .



The cells often show little variation in size and shape, although variable numbers of mitotic figures can usually be identified.

TREATMENT

Surgical excision, including a wide margin of adjacent normal tissue.

LEIOMYOSARCOMA

The **leiomyosarcoma** is a malignant neoplasm of smooth muscle differentiation, which accounts for 5% to 10% of all soft tissue sarcomas.

CLINICAL FEATURES

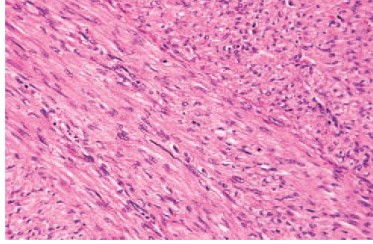
Tumors in the oral and maxillofacial region occur over a wide age range without a predilection for any age group. They have been reported at various sites, but half of all oral cases occur in the jawbones. The clinical appearance is nonspecific; there is usually an enlarging mass that may or may not be painful . Secondary ulceration of the mucosal surface may occur.



HISTOPATHOLOGIC FEATURES

The microscopic examination of a leiomyosarcoma shows fascicles of spindle-shaped cells with abundant eosinophilic cytoplasm and blunt-ended, cigar-shaped nuclei .

The degree of pleomorphism varies from one tumor to the next.



TREATMENT

Radical surgical excision, sometimes with adjunctive chemotherapy or radiation therapy.

RHABDOMYOSARCOMA

Rhabdomyosarcoma is a malignant neoplasm that is characterized by skeletal muscle differentiation. These tumors are much more common in young children, accounting for 60% of soft tissue sarcomas in childhood.

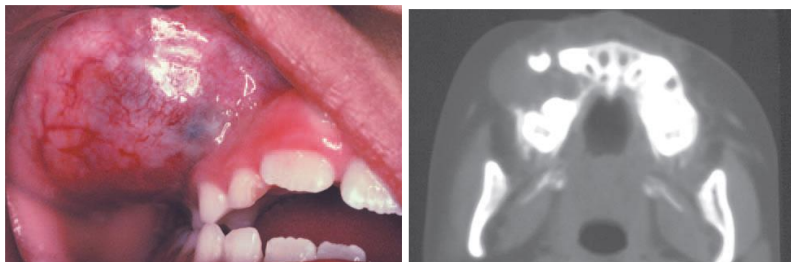
CLINICAL FEATURES

Rhabdomyosarcoma primarily occurs during the first decade of life but also may occur in teenagers and young adults. It is rare in people older than 45 years, and approximately 60% of all cases occur in males.

Major Types

- Embryonal rhabdomyosarcoma
- Alveolar rhabdomyosarcoma
- Undifferentiated sarcoma
- Anaplastic rhabdomyosarcoma

Most head and neck lesions are embryonal or alveolar types. The tumor is most often a painless, infiltrative mass that may grow rapidly. The palate is the most frequent intraoral site, and some lesions may appear to arise in the maxillary sinus and break through into the oral cavity.



HISTOPATHOLOGIC FEATURES

EMBRYONAL TYPE

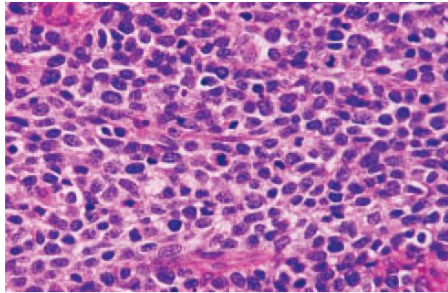
The embryonal rhabdomyosarcoma resembles various stages in the embryogenesis of skeletal muscle. Poorly differentiated examples may be difficult to diagnose and consist of small round or oval cells with hyperchromatic nuclei. Alternating hypercellular and myxoid zones

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may be seen. Better-differentiated lesions show round to ovoid rhabdomyoblasts with distinctly eosinophilic cytoplasm and fibrillar material around the nucleus. Some tumors show better-differentiated, elongated, strap-shaped rhabdomyoblasts.



ALVEOLAR TYPE

Both classic and solid variants of alveolar rhabdomyosarcoma are recognized. The classic pattern is characterized by aggregates of poorly differentiated round to oval cells separated by fibrous septa. These cells demonstrate a central loss of cohesiveness, which results in an alveolar pattern. In contrast, solid alveolar rhabdomyosarcoma demonstrates cellular fields of small round basophilic cells without fibrovascular septa.

TREATMENT

Treatment typically consists of local surgical excision followed by chemotherapy.