

CHAPTER 15

NONDONTOGENIC BENIGN TUMORS OF THE JAWS

Exostoses and Tori

Osteoma

Osteoid Osteoma

Central Hemangioma

Arteriovenous Fistula

Neurogenic Tumors (neurilemmoma, neurofibroma)

Traumatic Neuroma

Chondroma

TUMOR-LIKE AND REACTIVE LESIONS

Central Giant Cell Granuloma

Fibrous Dysplasia

Histiocytosis X

EXOSTOSES AND TORI

Exostoses and tori are localized peripheral overgrowths of bone due to some unknown cause. Although etiology is unknown, a hereditary basis is suspected. They are prevalent in certain populations; the highest incidence is in Eskimos, American Indians, and Asians. They occur on any surface of the jaw bones. When a bony protuberance occurs in the midline of the palate, it is called a torus palatinus; and when it occurs on the lingual surface of the mandible, it is called a torus mandibularis. When multiple small nodular protuberances appear on the buccal surfaces of the alveolar bone, they are called exostoses. The palatal torus occurs in about 25% of the U. S. population and is twice as more prevalent in females than in males. The mandibular torus occurs in about 7% of the U. S. population and has no predilection to a specific sex. Very small exostoses and tori consist entirely of compact bone but when large and nodular, the center consists of cancellous bone which is surrounded by compact bone. They vary in size from a slight elevation to a large protuberance.

Exostoses and tori are neither of pathological nor of clinical significance except that they interfere with normal speech and other functions. They also interfere with the preparation and insertion of prosthetic appliances, in which case surgical removal of the exostosis or torus may be contemplated. On a radiograph, exostoses and tori appear as well-defined round or oval radiopacities superimposed on the roots of teeth. If the radiopacities are at the apical ends of the roots, they may be confused with osteosclerosis. Unlike osteosclerosis, torus mandibularis occurs almost always bilaterally.

Fig. 15-1 Mandibular tori seen bilaterally.

Fig. 15-2 Torus palatinus as seen on a periapical film.

Fig. 15-3 Mandibular torus

OSTEOMA

Osteoma is a benign tumor of bone which occurs most commonly on the skull and jaw bones. It also occurs on long bones as an osteochondroma. It is composed primarily of cortical bone or cancellous bone or a combination of cortical and cancellous bones. The bony-hard discrete mass extends from the cortex and produces asymmetry of the jaw. Osteomas can also occur in maxillary and frontoethmoidal sinuses. Multiple osteomas of the jaws as well as of long bones and skull are a characteristic manifestation of Gardner's syndrome. In Gardner's syndrome, the patient has multiple osteomas, multiple polyps of the large intestine, multiple epidermoid cysts, desmoid (fibrous) tumors of skin, and impacted supernumerary and permanent teeth. On a radiograph, an osteoma presents a radiopaque mass projecting from the surface of bone. When composed of compact bone it is a completely opaque mass, but when composed of cancellous bone it shows evidence of internal trabecular structure.

Fig. 15-4 Osteoma of the compact bone arising from the mandibular cortex. It has to be differentiated from a calcified lymph node and a sialolith because of their similar radiographic appearance.

Fig. 15-5 Osteoma arising from the angle of the mandible.

Fig. 15-6 Osteoma of the mandible which is attached by a pedicle and could be palpated clinically. This osteoma consists of a combination of cortical and cancellous bones. The osteoma should be differentiated from a sialolith.

Fig. 15-7 Osteoma arising from the edentulous alveolar bone. The osteoma is of compact bone.

OSTEIOD OSTEOMA

Osteoid osteoma is a variation of an osteoma and rarely occurs in the jaws. It occurs most commonly in the femur and tibia. This benign tumor is found in young persons under the age of thirty. It occurs in or near the cortex and may produce swelling of overlying soft tissue. There is intense pain which is out of proportion to the size of the lesion. The radiographic appearance is very characteristic and shows a radiopaque nidus of osteoid tissue containing trabeculae. The osteoid tissue is surrounded by a diffuse and irregular radiolucency of vascular connective tissue which is surrounded by a rim of sclerotic bone. The radiographic appearance is sometimes confused with that of chronic suppurative osteomyelitis. Subperiosteal thickening may take place by the formation of new bone under the periosteum. There is no recurrence if the lesion is completely excised.

Osteoid osteoma bears considerable clinical, radiographic, and histologic similarity to a benign tumor called osteoblastoma. Many experts, in fact, regard the two lesions as identical. Classically, the distinction rests primarily on the size of the lesion: osteoid osteoma is under 2 cm, and osteoblastoma is larger than 2 cm.

Fig. 15-8 Osteoid osteoma in the region between the premolar and molar teeth.

CENTRAL HEMANGIOMA

Central hemangioma is a benign vascular neoplasm within bone which produces a proliferation of blood vessels. It occurs more often in the vertebrae and skull but rarely in the jaws. The posterior region of the mandible is the most frequent site of occurrence. The tumor is seen in children and teenagers; and females are affected twice as often as males.

The lesion produces a hard nontender slow-growing swelling. The teeth in the vicinity of the tumor may be loosened and bleeding may occur from the gingiva around the necks of the affected teeth. The teeth have increased hypermobility and may exhibit a pumping action such that, when depressed in an apical direction, the teeth rapidly resume their original position. The lesion may pulsate, and a bruit may be detected on auscultation. Some hemangiomas may be present without any sign or symptom.

The radiographic appearance may take many forms, in most instances it is that of a cyst having a well-defined radiolucency surrounded by a sclerotic border. Sometimes the lesion shows the classic multilocular radiolucency called honeycomb or soap bubble appearance. The roots of the teeth may be resorbed. The cortical region may be thinned and expanded.

The differential diagnosis of a central hemangioma is difficult because of its multiple radiographic appearances. Sometimes radiopaque phleboliths may be visible in the radiolucency. If an angiogram is taken, it will demonstrate the increased vascular supply to the lesion. A central hemangioma is a great surgical risk (including tooth extraction) since bleeding is difficult to control. Therefore, before surgery, the patient must be prepared for emergency blood transfusion and ligation of the carotids. Sometimes partial maxillectomy

and mandibulectomy may be necessary. After adequate removal of the tumor, the prognosis is good because the lesion does not recur and is not malignant.

Fig. 15-9 Central hemangioma involving the entire body of the mandible and showing a coarse trabecular pattern (Courtesy, Dr. A. Wuehrmann and Dr. L. Manson-Hing).

Fig. 15-10 Central hemangioma showing a well-circumscribed cavity.

ARTERIOVENOUS FISTULA (Arteriovenous shunt or malformation)

Arteriovenous fistula is a direct communication between an artery and a vein, and bypassing the intervening capillaries. It can occur in soft tissue or in bone as a result of trauma or a developmental anomaly. There may or may not be the presence of a swelling. Aspiration of the involved site will produce blood. Arteriovenous fistulae may be confused with hemangiomas. On a radiograph, the lesion may be unilocular or multilocular. The most characteristic location for its occurrence is the ramus-retromolar area of the mandible involving the mandibular canal. Sometimes phleboliths may occur in the lesion. An angiogram may be necessary to show the vascular nature of the lesion.

Fig. 15-11 Arteriovenous fistula involving the whole palate (Courtesy, Dr. John Coryn).

NEUROGENIC TUMORS

Neurogenic tumors are rare benign intraosseous nerve tumors which occur mostly in the body and ramus of the mandible. This is explained by the fact that the mandibular canal conveys a large neurovascular bundle for a long distance than does any other bony canal. These tumors arise from the nerve sheaths as well as from the nerve fibers in combination with their supporting tissues.

1) Neurilemmoma (Schwannoma)

Although neurilemmoma is a rare tumor, it is the most common of the neurogenic tumors. It arises from the Schwann's cells which make up the inner layer covering the peripheral nerves. This slow growing encapsulated benign tumor occurs at any age and may cause expansion and perforation of the cortical plates with subsequent soft tissue swelling. The soft tissue nodular mass is usually painless although pain may occur in rare instances. The tongue is the favored location, although lesions have been described in the palate, floor of the mouth, buccal mucosa, gingiva, lips, vestibule, and jaws. Bony lesions show a radiographic finding of a radiolucent area of bone distal to the mental foramen, and surrounded by a well-defined radiopaque border similar to that of a cyst. Sometimes the lesion may be multilocular. The lesion has an intimate relationship with the mandibular canal. Like all nerve tumors, the neurilemmoma is not responsive to x-radiation therapy. Therefore, surgical excision is the treatment of choice, especially since it is encapsulated (unlike neurofibroma which is non-encapsulated). For extensive bony lesions, curettage or resection should be performed.

2) Neurofibroma

Neurofibroma is a benign slow growing tumor composed of components of the peripheral nerves (including the axons) and from the connective tissue of the sheath of Schwann. Although the tumor is not common, it is by no means a clinical rarity. It can occur in soft tissue and/or in bone. Neurofibromas may appear as solitary lesions or as multiple lesions as part of von Recklinghausen's disease (syndrome neurofibromatosis). Neurofibroma occurs at any age but is usually found in young patients and has a high potential for malignant change. The solitary neurofibroma is asymptomatic and occurs on the tongue, buccal mucosa, and vestibule. The swelling is firm on palpation and may perforate the cortex. The central (bony) lesion associated with a mandibular nerve is most likely to produce pain and paresthesia. On a radiograph, a central neurofibroma of the inferior dental nerve appears as a fusiform enlargement of the canal. The margins of the lesion may or may not be sharply defined. The tumor is not sensitive to x-radiation therapy. The syndrome of von Recklinghausen's neurofibromatosis includes multiple neurofibromas on the skin, cutaneous café-au-lait spots (especially diagnostic when located in the axilla), bone deformities and neurologic abnormalities.

Fig. 15-12 Neurilemmoma (schwannoma) involving the mandibular canal and the mental foramen. The lesion has a well-defined radiopaque border (Courtesy, Dr. Doug Damm).

Fig. 15-13 Neurofibroma of the inferior dental nerve demonstrating a multilocular form.

TRAUMATIC NEUROMA (Amputation neuroma, neuroma)

Traumatic neuroma is not a true tumor but is a proliferation produced by a damaged or severed nerve trunk. The trauma to the nerve is caused by tooth extraction, local anesthetic injection, jaw fracture, soft tissue trauma, ill-fitting denture or accident. The traumatized nerve segments proliferate in an attempt to regenerate and re-establish innervation. When these nerve elements get entangled and trapped in the developing scar tissue, they form a composite mass of fibrous tissue and disorganized nerve at the site of injury. This tumor of neural elements and scar tissue is non-encapsulated. Unlike neurilemmoma and neurofibroma, neuralgic pain is experienced locally and in distant parts. The type of pain varies from one patient to another and ranges from occasional tenderness to constant severe pain.

In the oral cavity, the mental foramen is the most common site of occurrence; presumably arising following tooth extraction with damage to the mental nerve. Other common sites are alveolar ridge, lips and tongue where the small nodules of traumatic neuroma are visible. In the soft tissues, the nodule of traumatic neuroma is of normal color, firm and sessile. Traumatic neuroma should be included in a clinical differential diagnosis of any small mass that is spontaneously painful or painful when compressed. The bony lesion shows a discrete radiolucency. The lesion should be surgically excised along with the obstructing agent. Some cases may experience continued pain despite the absence of a lesion.

Fig. 15-14 Traumatic neuroma associated with a fracture of the ramus. Note the fracture extending from the posterior border of the ramus.

Fig. 15-15 Traumatic neuroma associated with the extraction site of the mandibular first molar. Note the bulbous enlargement of the mandibular canal due to increased growth.

CHONDROMA

It is a central benign cartilaginous tumor. Although it is common in other parts of the body, it is exceedingly rare in the jaws. The anterior portion of the maxilla, condyle and coronoid process are the most commonly involved. A chondroma is a painless, slow growing tumor producing destruction and exfoliation of teeth. Tumors that involve condyle and coronoid process may affect mandibular function. On a radiograph, the tumor appears as cyst-like radiolucencies; some are sclerotic. The borders are usually ill-defined. Irregular calcifications may occur with the radiolucencies and then it is called an osteochondroma. Roots of the involved teeth may be resorbed. A chondrosarcoma can develop from a pre-existing chondroma.

CENTRAL GIANT CELL GRANULOMA

Central giant cell granuloma is a fairly common lesion which occurs almost exclusively in the jawbones, the mandible is more frequently affected than the maxilla (3:1 ratio). The lesion occurs in the first three decades of life prior to 30 years of age. Its etiology is unknown. Investigators regard the lesion as either a reparative response or a benign nonodontogenic tumor or a developmental anomaly related closely to the aneurysmal bone cyst. The lesion occurs exclusively in the tooth-bearing areas of the jaws anterior to the molar teeth. When located near the anterior teeth, it is one of the few intrabony jaw lesions to cross the midline. A small lesion is detected incidentally on radiographic examination for an unrelated purpose. A large lesion is detected in the investigation of an asymptomatic localized expansion of a jawbone producing facial asymmetry.

Radiographic appearance is classically that of a multilocular radiolucency with discrete scalloped borders. Of the different types of multilocular lesions, the central giant cell granuloma is the single most common multilocular lesion. A few central giant cell granulomas appear as unilocular radiolucencies. The margins of the lesion may be either well-defined or poorly defined. Teeth may be displaced in cases of jaw expansion. In some cases, the central type of giant cell granuloma may be associated with the peripheral type. Since the histologic features of the giant cell granuloma are indistinguishable from those of a brown tumor of hyperparathyroidism, it is prudent to evaluate any patient with a central giant cell granuloma for the possibility of hyperparathyroidism. Treatment consists of conservative surgical removal by curettage. The prognosis is excellent with only rare recurrences.

Fig. 15-16 Central giant cell granuloma exhibiting a multilocular radiolucency with a localized expansion of the jawbone.

Fig. 15-17 Central giant cell granuloma shows jaw expansion on an occlusal projection. The multilocular lesion crosses the midline of the mandible

Fig. 15-18 Central giant cell granuloma in the midline region of the mandible.

Fig. 15-19 Central giant cell granuloma exhibiting a unilocular radiolucency with well-defined margins in the mandibular premolar region.

FIBROUS DYSPLASIA

Fibrous dysplasia is a benign fibro-osseous lesion in which normal medullary bone is gradually replaced by an abnormal fibrous connective tissue proliferation that contains foci of irregularly shaped trabeculae of immature bone. Microscopic examination shows these trabeculae to resemble Chinese characters. Fibrous dysplasia has some features of a metabolic condition and some features of a neoplastic condition. The lesion begins before puberty but is discovered later on. Fibrous dysplasia presents three clinical forms: monostotic (occurs in a single bone), polyostotic (occurs in many bones), and Albright's syndrome. Albright's syndrome consists of polyostotic fibrous dysplasia, café-au-lait spots on skin (skin pigmentation), and precocious sexual development. Fibrous dysplasia may affect any bone in the skeleton but most often affects the skull and facial bones, or ribs. Monostotic fibrous dysplasia is much more common than the polyostotic form. A painless, slow enlargement of the affected bone produces facial asymmetry. The swelling is bony hard to palpation. The maxilla is more often involved than the mandible.

On a radiograph, the lesion is seldom well-defined, it tends to blend imperceptibly into adjacent normal bone. The radiographic appearance varies, depending on its stage of maturity. In the early (osteolytic) stage, the lesion appears as a radiolucency; in the intermediate stage, it appears as a "ground glass" or "orange peel" appearance; in the final (mature) stage, it appears as a radiopacity. Conservative osseous contouring for cosmetic correction should be delayed until after the cessation of growth of the skeleton and of the lesion.

Fig. 15-20 Fibrous dysplasia presenting a "ground glass" appearance and an unilateral expansion of the mandible.

Fig. 15-21 Fibrous dysplasia presenting a "ground glass" appearance.

Fig. 15-22 Fibrous dysplasia in the mature stage presenting a radiopaque image. The affected region is expanded.

Fig. 15-23 Fibrous dysplasia of the maxilla showing a mixed appearance of radiopacities and radiolucencies.

HISTIOCYTOSIS X (Reticuloendotheliosis, Langerhans' cell granulomatosis)

Histiocytosis X is a group of reticuloendothelial diseases that are not well understood; the etiology and pathogenesis are unknown. It acts like a metabolic and a neoplastic disease. The disease is characterized by a proliferation of differentiated histiocyte-like cells. The three entities making up this group are: Letterer-Siwe disease, Hand-Schüller-Christian disease, and eosinophilic granuloma. Histiocytosis X affects the reticulo-endothelium organs such as the spleen, liver (hepatosplenomegaly), lymph nodes, bone marrow, and may infiltrate mucosa, skin or viscera. The jaws and oral soft tissues are involved in 20% of the cases and may be the only sites of the disease. The posterior region of the mandible is the most commonly involved site and may sometimes be associated with mild, dull pain. The radiographic appearance of teeth is that of "floating in air".

1. Letterer-Siwe disease occurs in the first three years of life. The disease often starts with a skin rash, persistent low-grade fever, malaise, and irritability. Other findings include hepatosplenomegaly, lymphadenopathy and anemia. In the oral cavity ulcerations may be present, as well as loss of alveolar bone. Severe cases usually result in death.
2. Hand-Schüller-Christian disease occurs in childhood between ages 3 to 10 years. In 10% of reported cases, the three classic signs are observed: bone lesions, diabetes insipidus (due to involvement of pituitary gland), and exophthalmos. The one or more punched-out radiolucent bone lesions of the skull are similar to the land and sea areas seen on a map, and hence called a geographic skull. In the oral cavity

there is severe loss of alveolar bone, which may resemble a common periodontal disease. Also, the oral soft-tissues may be involved.

3. Eosinophilic granuloma is believed to be a variant of Hand-Schüller-Christian disease and occurs in adolescents and young adults. It is a localized form of histiocytosis X. The skull and the jaws are both affected. The jaw lesions may be solitary or multiple radiolucencies and characteristically destroy the periodontal bone support of one or more teeth, especially in the posterior areas, resulting in "floating teeth."

Letterer-Siwe and Hand-Schüller-Christian diseases are treated by aggressive chemotherapy and radiotherapy similar to that used in leukemia. The prognosis of these two diseases is poor and often is fatal. Eosinophilic granuloma is usually treated by surgical curettage although radiotherapy is also effective. The prognosis of eosinophilic granuloma is good because there is only local destruction of tissue.

Fig. 15-24 Eosinophilic granuloma (histiocytosis X) destroying the periodontal bone around the roots of the maxillary first molar and giving the appearance of a "floating tooth".

Fig. 15-25 Eosinophilic granuloma destroying the bone around the mandibular left premolars and canine.

Fig. 15-26 Eosinophilic granuloma in the apical region of the mandibular premolars and first molar.

Fig. 15-27 Hand-Schüller-Christian disease (histiocytosis X) showing multiple radiolucent skull lesions giving the appearance of a geographic skull.