Oral pathology

Bone Neoplasms د.بشار

Primary tumors of bone are uncommon lesions in the jaws. They may arise from any of the number of different cells and tissues present in bone including cells (osteoblasts), cartilage, marrow, vascular and fibrous tissues.

No.	Tumor origin	Benign	Malignant
1	**Primary bone tumors**		
	a-of bone origin	**Osteoma** **Osteoid osteoma**	**Osteosarcoma**
	b-of chondroid origin	**Chondroma**	**Chondrosarcoma**
	c-of marrow origin		**Ewings sarcoma** **Lymphoma** **Multiple myeloma** **Leukemia**
	d-of fibrous tissue origin (fibroblastic)	**Desmoblastic fibroma**	
	e-others of vascular origin	**Haemangioma**	

2	**Metastatic tumor**		**Lung, adenocarcinoma, ovary, prostate and renal**

Benign tumors

Osteoma

Osteomas are benign tumors composed of mature compact or cancellous bone. They are essentially restricted to the craniofacial skeleton and rarely if ever, are diagnosed in other bones. The lesion is benign and probably not a true neoplasm. Some cases may represent end stage of other conditions, e.g. fibrous dysplasia or related fibro-osseous lesions. The common palatal and mandibular tori are not considered to represent osteomas, although they are histopathologically identical. The incidence varies, ranging from 0.0029 % to 3 %, the true incidence is unknown.

Clinical and Radiographic Features

Osteomas are most frequently diagnosed in 2nd to 4th decades of life, being uncommon in the 1st decade. Average patient age is from 25 to 35 years. The lesion may arise on the surface of the bone, as a polypoid or sessile mass “periosteal osteoma”, or may be located in the medullary bone “endostealosteoma”. The majority of cases are seen in young adults. It is generally asymptomatic, solitary lesions, or it could be an incidental finding in radiographic evaluation of the jaw for other problems.

In the head and neck region, the most common sites of origin are the paranasal sinuses, inner and outer tables of the cranial bones and the jaw bones. Extra skeletal osteomas occur in the buccal mucosa, tongue and nasal cavity; however, these are not true neoplasms and are termed “choristomas.”

In the gnathic region, the most common locations are the body of the mandible and the condyle. When it is located in the body, it occurs mostly to the premolars on the lingual surface.

Periosteal osteomas appear as slowly growing masses on the surface of the mandible or maxilla. Some types may reach a large size, resulting in facial deformity. Small endostealosteomas are asymptomatic, but large lesions cause a slowly progressive enlargement of the affected area.

An osteoma involving mandibular condyle may cause a slowly progressing shift in the patient's occlusion, with deviation of the midline of the chin toward the unaffected side. Other signs and symptoms include facial swelling, pain, and limited mouth opening.

Symptoms of osteomas in the head and neck region may be quite variable depending on the lesion's location and include, chronic sinusitis, local pain, headache, nasal obstruction, a painful or painless mass, exophthalmus, focal facial asymmetry, difficulty in mouth opening, meningitis, and hearing loss.

Radiologically, osteoma typically appears as a dense, opaque, sharply demarcated mass that is usually broad based and ranges from 1 cm. to 8.5 cm. in diameter. Periosteal osteomas may show a uniform sclerotic pattern or may demonstrate a sclerotic periphery with a central trabecular pattern. Small endostealosteomas are almost impossible to be differentiated from foci of sclerotic bone representing the end stage of an inflammatory process.

Histopathologic Features

Histologically, most osteomas are composed of hard, dense, compact lamellar bone, similar to cortical bone, in which haversian systems are present. These so-called ivory or compact osteomas have little stroma, and that which is present consists of bland fibrous tissue. Osteomas may also be composed predominantly of mature lamellar trabecular bone between which fat and marrow elements are found.

Treatment and Prognosis

Osteomas found incidentally in asymptomatic patients do not need removal, as follow up studies frequently have shown no increase in size over several years duration. For symptomatic lesions, local excision is curative in almost all cases. Recurrence is quite unusual.

Gardner Syndrome

Gardner syndrome is a rare disorder that is inherited as an autosomal dominant trait. The condition represents spectrum of diseases characterized by adenomatous polyps of the large bowel associated with multiple osteomas of the skull and mandible, multiple keratinous cysts of the skin and soft tissue neoplasms especially fibromatosis. Most of the fibromatoses are intra-abdominal and develop following surgical intervention.

These show coexistence of somatic and germline mutations of the APC gene (adenomatous polyposis coli gene) on chromosome 5q21, suggesting that inactivation of both alleles of this gene is involved in their development. Because of the variable degree of penetrance, only one of

the extracolonic manifestations may be present such as fibromatosis. In this association, the osteomas tend to be multiple and most frequently arise in the mandible, especially in the mandibular angle, and the maxilla. Osteomas may be the 1st manifestation of these syndromes and occur up to 10 years prior to the discovery of the intestinal polyps that ultimately transform into **adenocarcinoma.**

Clinical Features

The prevalence is variable from 1:8300 to 1:16000 live births. The colonic polyps typically develop during the second decade. In addition, detection of extracolonic polyps is not rare in small intestine or stomach.

About 90% of patients demonstrate skeletal abnormalities, the most common of which are osteomas. Although any part of the skeleton may be affected, the most common sites are the skull, paranasal sinuses, and the mandible, mostly at the mandibular angle, with prominent facial deformity.

The osteomas are usually seen during puberty and precede the development of, or any symptoms from, the bowel polyps. Most patients demonstrate between 3-6 osseous lesions. Dental abnormalities an increased prevalence of odontomas, supernumerary teeth, and impacted teeth. Most patients show one or several epidermoid cysts of the skin. To a lesser extent, an increased risk for thyroid carcinoma.

Histopathology

The same as osteoma.

Treatment and Prognosis

The major problem is the high rate of malignant transformation of bowel polyps into invasive adenocarcinoma. Prophylactic colectomy is usually recommended.

Osteoid osteoma and Osteoblastoma

Osteoid osteoma is a benign bone neoplasm that is found more frequently in patients between 10 and 30 years of age, and exhibit 2:1 male female ratio. Intense pain is the most prominent symptom, this is often sharply localized and accompanied by clinical or lab. Evidence of infection.

Oseoidosteoma has been reported in every bone but occurs more frequently in femur, tibia, humerus, bones of the hands and feet, vertebrae, and fibula. The tumor is very rare in the jaw bone. In the head and neck area, the cervical spine is the most common site.

Radiographically

The typical finding is a radiolucent central nidus that is seldom larger than 1.5 cm. and that may, or may not, contain a dense center. This nidus is surrounded by a peripheral sclerotic reaction that may extend for several centimeters.

Microscopically

The sharply delineated central nidus is composed of more or less calcified osteoid lined by plump osteoblasts and growing within highly vascularized connective tissue, without evidence of inflammation. Surrounding the nidus, there is a variably thick layer of dense bone.

The pain associated with this tumor is characteristically more intense at night, relieved by nonsteroidal anti-inflammatory drugs such as aspirin,

and eliminated by excision of the lesion. The pain has been attributed to be the effect on nerves and vessels of osteoblast- produced prostaglandin E2, which is typically present in large amounts in those lesions. Another suggestion is that pain is due to the presence of entrapped and proliferating nerves within and particularly around the nidus.

Osteoblasoma "Giant osteoid Osteoma"is a tumor closely related to osteoid osteoma both microscopically and ultrastructurally. It is distinguished from the osteoid osteoma by the larger size of the nidus, the absence or inconspicuousness of a surrounding area of reactive bone formation. Most cases arise in the medulla of the spine or major bones of the lower extremity, although cortical and subperiosteal forms also occur.

Because of the significant similarities between oseoblastoma and cementoblastoma some consider them to be identical, with one primary difference, which is fusion of the lesion to a tooth or not.

Clinically

Rarely affect the jaw bone, with slight mandibular predilection, mostly in the posterior regions. A slight male predominance is noted, and about 85 % occur before age 30. Most of the lesions are between 2 to 4 cm. but may be as large as 10 cm. Pain is a common presenting feature. Unlike osteoid osteoma, the pain is not relieved with aspirin.

In some cases it is difficult to distinguish between aggressive osteoblastoma and low grade osteosarcoma.

Treatment

Complete en block resection, is curative if not possible marginal resection, or curettage must be used with 10-20% recurrence rate. Prognosis is good. The lesion rarely recurs or transform into osteosarcoma.

Desmoplastic Fibroma

Desmoplastic fibroma is a benign, locally aggressive lesion of bone that can be considered the bony counterpart of fibromatosis. The tumor appears usually in long bones and the pelvis but may occasionally affect the jaws. The cause of desmoplastic fibroma is unknown. The lesion usually exhibits locally aggressive clinical behavior, suggesting a neoplastic process. The potential role of genetic, endocrine, and traumatic factors in the pathogenesis of the lesion has led to speculation that it might represent an exuberant reactive proliferation.

Clinical Features

Most cases of desmoplastic fibroma of the jaws have occurred in patients under the age of 30 years, with a mean age of 14 years. There appears to be no gender predilection. The mandible, usually the body ramus region, is affected more often than the maxilla. The lesions are slowly progressive and asymptomatic, eventually causing swelling of the jaw.

Radiographically

desmoplastic fibroma may be unilocular or multilocular. The radiographic margins may be either well demarcated or poorly defined. Cortical perforation and root resorption may be seen.

Histopathology

The lesion consists of interlacing bundles and whorled aggregates of densely collagenous tissue that contains uniform spindled and elongated fibroblasts. Some areas may exhibit hypercellularity with plumper fibroblast nuclei. However, cytologicatypia and mitotic figures are not found. Bone is not produced by lesional tissue.

Differential Diagnosis

Differential radiographic diagnostic considerations include odontogenic cysts, odontogenic tumors, and nonodontogenic lesions that typically occur in this age group. The presence of aggressive features, such as cortical perforation, or local symptoms might suggest the possibility of a malignancy. In some cases histopathologic distinction between desmoplastic fibroma and well-differentiated fibrosarcoma may be difficult. The latter would exhibit greater cellularity, mitotic figures, and nuclear pleomorphism. Some similarities are noted histologically with central odontogenic fibroma, a nonaggressive lesion that contains odontogenic rests.

Treatment

Surgical resection of the lesion is generally reported as the treatment of choice. Curettage alone has been associated with a significant recurrence rate.

Hemangioma of Bone

Hemangiomas of bone are rare intraosseous vascular malformations that, when seen in the jaws, can mimic both odontogenic and nonodontogenic lesions. Difficult to control hemorrhage is a notable complication of surgical intervention.

Clinical Features

More than half of the central hemangiomas of the jaws occur in the mandible, especially the posterior region. The lesion occurs approximately twice as often in females as in males. The peak age of discovery is the second decade of life.

A firm, slow-growing, asymmetric expansion of the mandible or maxilla is the most common patient complaint. Spontaneous gingival bleeding around teeth in the area of the hemangioma may also be noted. Paresthesia or pain, as well as vertical mobility of involved teeth, is occasionally evident. Bruits or pulsation of large lesions may be detected with careful auscultation or palpation of the thinned cortical plates.

Trophic effects of the hemangioma on adjacent hard and soft tissues are also common. Significantly, hemangiomas may be present without any signs or symptoms.

Radiographic findings: more than half of jaw hemangiomas occur as multilocular radiolucencies that have a characteristic soap bubble appearance.

A second form of these lesions consists of a rounded, radiolucent lesion in which bony trabeculae radiate from the center of the lesion, producing angular loculations. Less commonly, hemangiomas appear as cyst-like

radiolucencies. The lesions may produce resorption of the roots of teeth in the area.

Histopathology

Hemangiomas of bone represent a proliferation of blood vessels. Most intrabonyhemangiomas are of the cavernous type (large-caliber vessels); fewer are of the capillary type (smallcaliber vessels). Separation of hemangiomas into one of these two microscopic subtypes is, however, academic, since there is no difference in biologic behavior.

Differential Diagnosis

The differential diagnosis of multilocularhemangioma of bone includes ameloblastoma, odontogenicmyxoma, odontogenickeratocyst, CGCG, and aneurysmal bone cyst. A unilocular lesion may be easily confused with other cystic processes that occur within the jaws. Angiography often provides useful information in establishing the diagnosis of hemangioma.

Treatment and Prognosis

The most significant feature of hemangiomas of bone is that these lesions may prove life threatening if improperly managed. Extraction of teeth in an area involved by a central vascular lesion may result in potentially fatal bleeding. It is imperative to perform needle aspiration of any central lesion that may be of vascular origin before performing a biopsy.

Methods used in the treatment of hemangioma of bone include surgery, radiation therapy, sclerosing agents, cryotherapy, and presurgical embolization techniques. The vascular supply of a given lesion, as well as its size and location, must be evaluated before the selection of a given treatment method.