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Research Article

Benign and Malignant Lesions Affecting the Mandibular Condyle

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Abstract

Purpose: A myriad of benign and malignant conditions may affect the mandibular condyle. Often, such conditions remain undiagnosed until they become symptomatic or incidentally discovered on diagnostic images. Radiologic findings in some of these lesions may be unspecific, therefore it is paramount to understand the clinical, radiologic, and histological presentation of such pathologies in order to provide an adequate diagnosis and a treatment plan. The purpose of this paper is to perform a non-systematic review of the literature of both benign and malignant lesions that can affect the mandibular condyle.

Materials and Methods: A review of the English literature was done looking for benign and malignant lesions that can affect the mandibular condyle.

Results: A myriad of benign and malignant conditions can affect the mandibular condyle. Treatment for these conditions is usually surgical.

Conclusion: The mandibular condyle can be affected by a plethora of benign and malignant conditions, which require a proper understanding by the surgical team in order to diagnose and treat them properly.

INTRODUCTION

The skull is composed of the cranial vault, the cranial base, the facial skeleton, the jaws, the acoustic cavity, and the cranial cavity. Six pairs of bones compose the facial skeleton: nasal, lacrimal, palatine, inferior nasal concha, maxilla, and zygomatic; and 2 solitary bones: mandible and vomer. The mandible is the only movable bone of the facial skeleton and it is composed of a body, ramus, and condyle. The condyle articulates with the temporal bone to form the temporomandibular joint (TMJ). Of all the functions of the mandible, movement of the mouth is the most essential. The mandible helps to protect important facial structures, houses the lower teeth, and allows other functions such as speech, yawning and mastication [1].

Development of the mandible takes place from the fifth week post-fertilization. The Meckel cartilage is formed from condensation of mesenchyme and serves as a cartilaginous scaffold for eventual ossification of the mandible. Additional mandibular growth follows, resulting in the accommodation of the muscles of mastication and the different anatomical components including the mandibular symphysis, angle, body, coronoid process and condyle. The distal and proximal portions of the mandible undergo endochondral ossification while the central portion undergoes intramembranous ossification. The mandibular condyle begins its development at approximately the 14th week of embryonic life and it is characterized by a rapid growth of endochondral ossification displacing the condyle upward and laterally while appropriately positioning the lateral pterygoid muscle attachment. At approximately 12 week of development, the condyle elongates towards the temporal bone to form the TMJ [2,3].

The mandibular condyle is innervated by branches of the external carotid artery (ECA), which is divided into the superficial temporal artery and the maxillary artery. Inferior to the TMJ, the maxillary artery is located deep to the condylar neck forming the deep auricular artery, anterior tympanic artery and middle meningeal artery that supplies the retrodiscal tissue of the TMJ. The innervation of the mandibular condyle is carried by the auriculotemporal and masseteric branches of mandibular branch of the trigeminal nerve (V3) [4].

A significant group of benign and malignant lesions can occur in the mandible including the condyle. Radiologic findings in some of these lesions may be non-specific, so the importance of the understanding the clinical presentation and histopathology in order to determine a proper diagnosis and future treatment plan. The purpose of this paper is to provide an overview of both benign and malignant lesions that can affect the mandibular condyle.

MATERIALS AND METHODS

A search of the English literature was performed. Key words for the Medline search included: temporomandibular joint, mandibular condyle, pathology, benign lesions, malignant lesions. Additionally, relevant publications from the reference list of the retrieved papers were also considered. The matches

were evaluated for relevance and analyzed accordingly. Reports dealing with benign and malignant lesions of the mandibular condyle written in English and received until December 15, 2019 were considered.

BENIGN LESIONS AFFECTING THE MANDIBULAR CONDYLE

The most common benign entities affecting the mandibular condyle are myxoma, osteoma, osteoblastoma and osteoid osteoma, chondroma, osteochondroma, chondroblastoma, giant cell tumor, and aneurysmal bone cyst.

Myxoma

Myxomas are described as benign tumors of connective tissue origin than can be present in hard and soft tissues within the body [4]. Myxoma of the jaw is considered a rare benign odontogenic tumor, most commonly presenting as an asymptomatic expansive, slow growing, non-metastatic, locally aggressive lesion, most frequently found at the center of the mandible. Its occurrence is between the second and third decade of life with female predilection. It comprises about 3-6% of all odontogenic tumors. Odontogenic myxomas are the second most common odontogenic entities after ameloblastomas [5-7]. It appears to originate from the dental papilla, follicle, or periodontal ligament [7]. Radiographically, it presents as a unilocular or multilocular radiolucency with diffuse borders resembling a "soap bubble", "honeycomb" or "tennis racquet strings" lesion that may cause teeth resorption and/or displacement. [8]. Histopathologic features include spindled, stellate-shaped cells in a mucoid-rich intercellular matrix with a ground substance of glycosaminoglycan, hyaluronic acid, and chondroitin sulfate [9].

The treatment for myxoma is surgical. Different therapy options have been suggested, ranging from conservative to aggressive approaches. Conservative options such as curettage and enucleation have a greater incidence of recurrence. The best treatment option seems to be resection with a security margin of 1.5-2.0 cm followed by plate reconstruction. Other factors that may be considered in treatment success are patient follow-up, motivation, and compliance [4].

Osteoma

Osteomas are benign, slow growing osteogenic tumors characterized by proliferation of mature compact or cancellous bone. Three entities are described: central, peripheral, and extra-skeletal. In the maxillofacial region central and peripheral entities are the most frequently found [10]. These affects the mandible in all its anatomical regions. Peripheral cases involving the mandibular condyle are very rare and have been minimally described in the literature [11]. Other locations include paranasal, frontal, ethmoidal and maxillary sinuses [12]. Although of unknown etiology, but some authors believe it is new bone apposition causing its formation as a result of multiple stimuli including inflammation, trauma or infection.

This entity is most commonly seen in the third decade of life with a female predilection. Most cases are solitary and asymptomatic, characterized by slow growth. Radiographically, osteomas are radiopaque and are described as a "mushroom shaped", well-circumscribed sclerotic solid mass. Some can be pedunculated, others show a broad base. Histologic appearance is divided into two presentations: a) compact osteoma (dense, compact bone with few marrow spaces) and b) cancellous osteoma (bony trabeculae with fibro fatty marrow) [13].

Osteomas in the condyle cause a slow, progressive shift in occlusion and deviation of the chin towards the unaltered site, facial asymmetry, malocclusion, pain and mouth opening limitation. An entity that can mimic a mandibular condyle osteoma is condylar hyperplasia. These can be distinguished by the lobular appearance of the osteoma as opposed to the enlarged shape of the hyperplastic condyle [14]. Overall, symptomless osteomas in the mandible do not require treatment, however, when the mandibular condyle is affected and disturbance of the normal function is present, a condylectomy is the treatment of choice. Recurrence rate after excision is low and no report of malignant transformation has been described [15].

There is an inherited autosomal dominant condition that represents a variant of a familial adenomatous polyposis with the name of Gardner's Syndrome. It is characterized by intestinal adenomatous polyps and multiples osteomas that can be found in different areas of the maxillofacial region [16].

Osteoblastoma and Osteoid Osteoma

Osteoblastoma and Osteoid Osteoma are similar entities, both described as being bone tumors arising from osteoblasts. Although osteoblastomas are benign tumors commonly affecting long bones, a little less than 10% of all cases reported, occurred in the maxillofacial region. It has a predilection for the posterior mandible, affecting more males than females and usually presenting before the third decade of life. These are associated with rapid onset of dull and persistent pain and swelling. They present as a well circumscribed, solitary lesion with expansible potential, ranging from 1 to 10cm in diameter. It has osteoblastic origin and is characterized by proliferation of osteoblasts within a high vascular fibro-cellular stroma [17,18].

Radiographic appearance includes a round to oval radiolucency with calcified areas. Most osteoblastomas arise within the medullary bone. However, periosteal or intracortical origin may be possible [19]. Osteoid osteoma is a bone-forming tumor that comprises about 3 % of all primary bone tumors, being most common in the second and third decades of life, showing a male predilection. Its origin can be the result of an inflammatory process or impaired healing process. The clinical appearance is similar to the osteoblastoma with the difference that the pain is relieved with NSAIDS as opposed to the osteoblastoma which is non-reactive to analgesic management [20]. Radiographic findings consist of a demarcated central nidus measuring less than 1 cm in diameter, which might contain localized dense or patchy mineralization with surrounding sclerotic bone. That nidus might include a small radiopaque center resulting in target-like appearance [21]. Both osteoid osteoma and osteoblastoma share histologic features, composed of irregular trabecular or osteoid bone matrix surrounded by osteoblasts and osteoclasts. The osteoblast has abundant cytoplasm and hyper-chromatic nuclei. The loose fibrous stroma contains dilated vessels, but in the osteoid osteoma is more sclerotic at its periphery. The

recommended treatment is surgical excision because it reduces the pain, curing the disease. Some cases can undergo spontaneous remission with no recurrence and no malignant transformation, similar to the osteoblastoma which rarely transforms in osteosarcoma [12].

Chondroma

Chondroma is a benign tumor composed of hyaline cartilage, most commonly affecting hands and feet. Chondromas are classified as follows: a) enchondromas: arising within the medullary bone; b) juxtacortical: arising from the periosteum, and c) periosteal: below the periosteum. These lesions are uncommon in the mandibular condyle. They represent 2.38% of all osteo-cartilaginous tumors [22,23]. In the maxillofacial region, chondromas can develop in the tongue, temporomandibular joint, cheeks, hyoid bone, and the mandibular condyle. It has a female predilection and most commonly occur during the second, third and fourth decades of life. It is characterized by a slow, painless growth. If the tumor is involving the mandibular condyle, it can cause mandibular deviation and mouth opening limitation.

Radiographically, they appear as a well-defined radiolucency with focal opacifications and a small radiopaque area. The two most common presentations are enchondromas and periosteal chondromas. Histopathologic characteristics include mature hyaline cartilage with numerous chondrocytes and small calcified areas. The recommended treatment is complete surgical removal, which results in low recurrence [12,24]. If a condylectomy is performed, condylar reconstruction is necessary to maintain the vertical dimension of the mandible [25].

Osteochondroma

Osteochondromas are the most common benign bone tumors, representing 8-15% of all primary bone tumors, with a 0.6% incidence in the maxillofacial region [26]. It usually affects the condyle, coronoid process, posterior maxilla, zygomatic arch, maxillary sinus, mandibular body and symphysis. It occurs most frequently between the first and third decades of life, showing a male predilection. Clinical hallmarks include non-tender, painless deformity, malocclusion, click joint sound on the affected side and deviation upon opening toward the contralateral site [27]. Radiological characteristics consist of a lobulated growth composed of medullar and cortical bone with a calcified cartilaginous cap which is better evidenced in magnetic resonance imaging (MRI). The histopathologic components include a proliferation of chondrocytes with surrounding periosteum [28,29]. The treatment of choice is surgical resection. No malignant transformation has been reported [30].

Chondroblastoma

Chondroblastoma is a rare benign cartilaginous tumor representing less than 1% of all bone tumors. It is most commonly found in long bones and its involvement in the maxillofacial region has been minimally reported. It can be found in the mandible, temporal bone, the skull and the mandibular condyle. Most often affects individuals between the second and fourth decades of life. Clinical presentation includes facial swelling and limited jaw motion [31]. Radiographic appearance includes resorptive defects or condylar enlargement associated with thinning of the bone cortex. Histopathologic characteristics include proliferative fibrous and cartilaginous tissue with multinucleated giant cells, often with polygonal or spindle-shaped cells and a cellular matrix [32]. The recommended treatment is surgical removal. Enucleation has shown a recurrence rate of approximately 55% [33].

Giant cell tumor

Giant cell tumors (GCT), are primary bone neoplasms, representing 5% of all bone tumors. These mainly originate inside the metaphyseal vicinity of the long bones, specifically within the distal femur, proximal fibula and distal radius. This entity consist of a tumor arising from connective tissue within the bone marrow. These are uncommon lesions in the maxillofacial area, usually reported in the sphenoid, temporal, frontal, parietal, and occipital bones, can be locally aggressive and shows unpredictable behavior [34]. It has a female predilection, and usually occurs between the second and fourth decade of life. Clinical symptoms include pain, limited range of motion, pain in the retromandibular area, hearing difficulties, facial paralysis, and trismus [35]. Radiographic characteristics include a well circumscribed expansive and destructive soft tissue mass. Due to its vague radiological features, histopathological confirmation is essential for diagnosis [35,36]. Histopathologic characteristics demonstrate large multinucleated giant cells within spindle-shaped stromal cells in an abundant eosinophilic cytoplasm [37]. The recommended treatment is surgical excision, which shows less recurrence rate, as opposed to limited resection or curettage, which present a recurrence rate of 40-60%. Depending on its location, GCT might not always be completely excised [35].

Aneurysmal bone cyst

Aneurysmal bone cyst (ABC) is considered an osteolytic benign pseudocyst. Presents as an expansive lesion of variable size with blood-filled spaces surrounded by cellular fibrous connective tissue and reactive bone. It is thought to be a reactive lesion after trauma, local hemodynamic alterations, and/or arteriovenous malformations, increasing venous pressure and expanding the vascular tissue, leading to bone resorption and connective tissue replacement. It commonly affects long bones or vertebrae, presents no sex or age predilection. In the maxillofacial region, usually occurs in the posterior segments of the mandible.

When associated to the mandibular condyle, it appears as a non-tender swelling with mild bony expansion. It can be painless or painful showing limited mouth opening, clicks, and malocclusion. Radiographically, it shows a unilocular or multilocular radiolucency, usually with cortical expansion and thinning. Its borders can present well or poorly defined, resembling a "blow-out" distention of the affected bone. Trabeculae of reactive bone can be observed as a radiopaque area within the radiolucent lesion.

Histopathologic features include multinucleated giant cells and osteoid, woven bone surrounded by blood filled spaced of varying size. Absence of smooth muscle component is very common. Treatment options include curettage or enucleation, arterial embolization, and cryosurgery. *En bloc* resection is reserved for recurrent lesions and the use of bone graft is not indicated since the surgical defect heals approximately within 6

Table 1: Benign and malignant conditions of the mandibular condyle.

Lesion	Behavior	Origin	Presentation	Radiographic	Histology	Treatment
Myxoma	Benign	Dental papilla, follicle, periodontal ligament	Asymptomatic, expansible and slow growing. Locally aggressive. No metastatic	Unilocular or multilocular radiolucency with diffuse borders. Soap bubble, honeycomb and tennis racquet strings appearance	Spindle, Stellate shaped cells in a mucoid rich intercellular matrix with ground substance of glycodasminglycans, hyaluronic acid, chondroid culfate	Resection with 1.5-2 cm margins. Enucleation and current age yield high recurrence rate
Osteoma	Benign	Unknow. New bone apposition due to multiple stimuli (inflammation,trauma or infection)	Shift in occlusion, chin deviation towards unaltered side, facial asymmetry, pain and mouth opening limitation	Mushroom shaped, circumscribed sclerotic solid mass radiopaque lesion. Pedunculated Broad base	1) compact osteomas: dense compact bone with few marrow spaces. 2) Cancellous osteomas: bony trabeculae with fibrofatty marrow	Condylectomy if symptomatic
Osteoblastoma	Benign	Osteoblasts	Well circumscribed, solitary lesion with expansile potential, ranging from 1-10 cm in dimension	Round to oval radiolucency with calcified areas	Proliferation of osteoblasts within a high vascular fibro cellular stroma	Surgical excision. Some cases can undergo spontaneous remission
Osteoid osteoma	Benign	Inflammatory process or impaired healing process	Similar to osteoblastoma. However, pain can decrease with NSAIDs	Demarcated central nidus which may contain localized dense or patchy mineralization with surrounding sclerotic bone. The nidus might contain small radiopaque center resulting in target like appearance	Irregular trabecular or osteoid bone matrix surrounded by osteoblastoma and osteoclasts. Osteoblast with abundant cytoplasm and hyper chromatic nuclei. The loose fibrous stroma contains dilated vessels, but the osteoid osteoma is more sclerotic at the periphery	Surgical excision. Some cases can undergo spontaneous remission
Chondroma	Benign	Hyaline cartilage	Sow growth, painless, mandibular deviation and mouth opening limitations	Well defined radiolucency with focal opacifications and small radiopaque area inside	Mature hyaline cartilage with numerous chondrocytes a small calcified areas	Complete surgical removal
Osteochondroma	Benign	Unknown developmental aberration	Non-tender painless deformity, malocclusion, click joint sound	Lobulated growth composed of medullar and cortical bone with a cartilaginous cap	Proliferation of chondrocytes with periosteum	Resection of the affected condyle
Chondroblastoma	Benign	Chondroblast	Facial swelling and diminished jaw movement	Resorptive defect or condylar enlargement with thinning of the cortex	Proliferation fibrous and cartilaginous tissue with multinucleated giant cells often with polygonal or spindle-shaped and cellular matrix surrounded by cartilaginous matrix	Removal of the attached condyle. Enucleation have recurrence rate of 55%

Giant cell tumor	Benign	Connective tissue within the bone marrow	Pain in retromolar area limited ranger of motion, hearing difficulties, facial paralysis, trismus	Well circumscribed expansive and destructive soft tissue mass	Large multinucleated giant cells disposed within spindle-shaped stromatolites cells in abundant eosinophilia cytoplasms	Surgical excision. Limited resection or current age represents recurrence rate of 40-60%
Aneurysmal bone cyst	Benign	Reactive lesion followed by trauma, locally hemodynamics alteration, AVM, leading to bone resorption	Non-tender swelling with low bony expansion. Limited mouth opening, click and malocclusion	Unilocular or multilocular radiolucency, usually with cortical expansion and thinning. Radiopaque trabecular reactive bone. Resembling blow out lesion	Multinucleated giant cells with osteoid and woven bone surrounded by blood filled spaced of varying size	Curettage or enucleation, arterial embolization, cryosurgery. En bloc resection for recurrences
Osteosarcoma	Malignant	Osteogenic mesenchymal matrix forming osteoid or immature bone	Swelling,pain, parenthesis, mouth opening limitation, nasal obstruction	Radiopaque or mixed radiolucency-radiopaque, unilocular bone destructive lesion with irregular margins that might appear sclerotic or mixed with sunburst appearance and Codman triangle sign	Formation of osteoid by malignant mesenchymal cells. Chondroid and fibrous connective tissue. Cells might be uniform, round or spindle-shaped	Surgical resection is the goal standard. Radiotherapy and chemotherapy is going to be dependent on prognosis, size, and surgical margins invasion. Radical neck dissection if lymph nodes are involved. 5 year survival rate 60-80%
Chondrosarcoma	Malignant	Cartilage	Painless swelling, trismus, lateral deviation of the mandible	Ill-defined radiolucency with radiopaque foci. Penetration of the cortex demonstrate sunburst appearance	Atypical chondrocytes organized in hyaline matrix with different degree of maturation	Surgical resection and curretage with cryosurgery. Radiotherapy and chemotherapy only for Hugh grade presentation 5 years survival rate of 87.2%
Fibrosarcoma	Malignant	Fibroblastic producing collagen and elastin	Pain, swelling, rapid clinical growth. Symptoms can be consumed with TMD	Lytic lesion with destructive pattern, cortex is disrupted, soft tissue invasion can be present	Spindle-shaped cells arrange in a fascicular growth pattern with collagen production	Surgical excision. Curettage or local excision shows high recurrence
Metastatic tumor	Malignant	Most common organs from which carcinoma leads are breast, lung, thyroid, prostate, kidney and pancreas	Pin, swelling, paresthesia, trismus, tooth mobility, preauticular swelling, parotid swelling	Moth-eaten radiolucency. Some tumors might induced bone formation producing a mixed radiolucency	Exhibits characteristics based on primary tumor. They are all poorly differentiated	Goal of treatment depends on the underlying tumor, pain relief and prevention of pathological fractures. Bisphosphonates can help slow progression and prevent fracture. 4 year survival rate of 10%

Ewing sarcoma	Malignant	Neural crest. Mesenchymal cells with potential for limited neural crest differentiation	Painful local mass, swelling, increase ESR and leukocytes	Non-specific, diffuse irregular radiolucency or a mi ex's radiolucent-radiopaque with courtical expansion and periosteum reaction (onion-skin) like appearance	Large sheets of small round cells with well-delineated nuclear outlines and ill-defined boarded. Tumors cells are separated by fibrovascular septa creating a lobular pattern	Resection with chemotherapy showing a 5 year survival rate of 70%
Multiple myeloma	Malignant	Plasma cell, characterized by proliferation of abnormal immunoglobulin-secreting plasma cells	Pain, bone swelling and function impairment	Multiple radiolucent lesions characterized as a punched out lesion	Massive diffuse stromatolites infiltration by monoclonal binucleate and trinucleate plasmacytes with basophilic cytoplasms, hypodense chromatin and nucleus with multiple nucleoli	Surgery, radiation and chemotherapy. 5 year survival rate if 55%
Malignant fibrous histiocytoma	Malignant	Arise from soft tissue, tendons, bones and joint	Solid mass	Lobulated mass with muscle density and peripheral solid portions, with calcification in some instances	Lesion filled with fibroblast and histocyte-like cells in different distribution with spindle and round cell	Local excision and radiotherapy 5 year survival rate is 46%

months after enucleation. It presents a wide range of recurrence rate, from 8-70% usually attributed to incomplete removal. Management depends on the age of the patient, aggressiveness of the lesion, size, location and the lesion behavior [12,38-40].

MALIGNANT LESIONS AFFECTING THE MANDIBULAR CONDYLE

Osteosarcoma

Osteosarcoma is one of the most common malignant bone tumors, representing about 30% of all primary malignant tumors. It originates from osteogenic mesenchymal matrix, forming osteoid or immature bone. It commonly arises from long bones and their incidences in the jaw have been reported to range from 5 to 10%. Although of unknown etiology, hormonal factors play a major role in the development of these lesions. Others factors that can be associated are radiation exposure, Paget's disease, Li-Fraumeni syndrome and Rothmund-Thompson syndrome [12,41,42].

It usually occurs between the third and fourth decades of life showing a male predilection [43]. It is most commonly seen in the mandibular body, followed by the angle, symphysis, ramus and condyle. Maxillary lesions arise predominately from the inferior portion (alveolar ridge, sinus floor, palate).

Clinical presentation may involve swelling, pain, paresthesia, mouth opening limitation and nasal obstruction, depending on tumor extension. Radiologic features include a radiopaque or mixed radiolucent-radiopaque, unilocular bone destructive lesion with irregular margins that might appear sclerotic or mixed with a sunburst appearance and Codman triangle sign. A computed tomography (CT), scan is necessary for detection of pulmonary metastasis [12,42]. The main histopathologic feature is the formation of osteoid by malignant mesenchymal cells. Also, chondroid and fibrous connective tissue might be found. The cells may be uniform, round or spindle-shaped and these are classified depending on the amount of osteoid, collagen or cartilage content within the lesion, as osteoblastic, chondroblastic or fibroblastic. Surgical resection with 2 cm margins is the gold standard of treatment and the use of chemotherapy and radiotherapy for these tumors is going to be dependent upon its prognosis, size and surgical margins invasion at time of resection. A radical neck dissection is indicated if lymph nodes are associated. Osteosarcomas of the jaws usually do not metastasize. Patients with localized disease at diagnosis exhibit a 5 years survival rate of approximately 60-80% [12,41].

Chondrosarcoma

Chondrosarcomas are rare malignant mesenchymal tumors originating from cartilage. It is the third most common primary malignant neoplasm of the bone, as they comprise 10% -12% of all malignant mesenchymal tumors. It is most commonly seen in femur, humerus, and pelvis, but in the head and neck region it has been reported with an incidence of 1 - 12%. The maxilla is most commonly affected, followed by the nasal septum, ethmoid, condyle and the coronoid process. It is classified as two different entities, depending on their developmental origin. Primary, derived from normal cartilage, and secondary, from pre-existing benign lesions. They occur between the second and third decades of life with a male predilection. Clinical manifestations included painless swelling, trismus and lateral deviation of mandible. Maxillary lesions can lead to nasal obstruction, congestion and visual loss.

Radiographic appearance shows an ill-defined radiolucency with radiopaque foci. Penetration to the cortex can demonstrate a sunburst appearance similar to the one seen in osteosarcomas. Histopathologic features include atypical chondrocytes organized in a hyaline matrix with different degree of maturation. These tumors can be classified depending on their aggressiveness. Low grade tumors resemble normal cartilage, but with the increase of tumor grading, less cartilaginous matrix and an increase in cellularity, nuclear size, pleomorphism, mitotic activity, and necrosis are found.

Uncommon variants of chondrosarcoma include, clear cell chondrosarcoma, dedifferentiated chondrosarcoma, myxoid chondrosarcoma, and mesenchymal chondrosarcoma. Treatment options include surgical resection and curettage with cryosurgery, in stage I chondrosarcomas. Radiation and chemotherapy are not very efficient, and therefore are reserved for high-grade presentation. They present low metastatic potential, and their prognosis will depend on their clinical and histopathological staging and the treatment selected. These tumors presents a 5 years survival rate of 87.2%. [12,44-47].

Fibrosarcoma

Fibrosarcomas are malignant mesenchymal tumors of fibroblastic origin producing collagen and elastin. They are most commonly seen in long bones and represent 5 - 6% of all adult soft tissue sarcomas. They constitute about 10 % of the sarcomas occurring in the maxillofacial region [48]. They can arise within soft tissue or bone. The mandible is more likely involved than the maxilla, affecting primarily the ramus and condyle. It has a male predilection, with an average incidence between the second and third decades of life. Clinical findings include pain, swelling, rapid clinical growth and in some cases, its symptoms can be confused with temporomandibular disorder (TMD). Radiographic characteristics show a lytic lesion with a destructive pattern, the cortex is disrupted, and soft tissue invasion can be present. Histopathologic features include a population of spindle-shaped cells arranged in a fascicular growth pattern with collagen production. The treatment of choice is surgical excision. Due to his aggressiveness and ability to infiltrate adjacent structures, curettage or local excision have shown high recurrence rate [12,49,50].

Metastatic carcinoma

Metastatic carcinoma is the most common malignant bone tumor. The most common organs from which primary carcinoma leads to bony metastasis are breast, lung, thyroid, prostate, kidneys and pancreas. Metastatic disease to the oral cavity is not very common, representing about 1 - 8% of all oral malignancies. Their incidence in the mandibular condyle is even less common [51]. The etiology for condyle involvement has been reported to be due to hematogenous spread, because the condyle presents poor local blood supply, lack of bone marrow and lymphatic system. It usually occurs between the fourth and seventh decade of life.

Clinical findings are pain, swelling, paresthesia, trismus, tooth mobility, preauricular swelling, and parotid swelling. Radiographic appearance includes a “moth-eaten” radiolucency, but some tumors may induce bone formation producing a mixed radiolucency (commonly seen associated to breast and prostate cancer metastasis). Histopathology varies from one individual to another. It exhibits characteristics based on the primary tumor. However, they are all poorly differentiated. In some cases, diagnosis of a metastasis bone tumor in the maxillofacial region can indicate the location of a primary occult tumor. The goal of treatment depends on the underlying tumor, pain managment, and the prevention of infection or pathologic fracture. Bisphosphonates therapy can help to slow progression and prevent fractures. Bone metastasis is categorized as stage IV disease; with very poor prognosis, showing a 4-year survival rate of 10% with most of the patients surviving less than 1 year [12,51-53].

Ewing sarcoma

Ewing sarcoma is a rare malignant bone neoplasm arising from small and undifferentiated round cells. It is most commonly seen in pelvis and long bones. It accounts for 1 % of all childhood malignancies. In the maxillofacial region the reported rate is less than 1% of all Ewing sarcomas. The mandibular ramus is the most common site followed by the condyle. Although of unknown etiology, possible etiologies are reported to be from the neural crest, and from mesenchymal cells with potential for limited neural crest differentiation. Occurring primarily in adolescents with a mean age of 15 with and showing a male predilection, clinical findings include a painful local mass, swelling, increase erythrocytes sedimentation rate, with leukocytosis commonly seen in advanced disease.

Radiologic features are non-specific, showing a diffuse irregular radiolucency or a mixed radiolucent and radiopaque lesion, with cortical expansion and a periosteal reaction that can be interpreted an “onion-skin” like appearance. Histopathologic findings include large sheets of small round cells with well-delineated nuclear outlines and ill-defined borders, and the tumors cells are separated by fibrovascular septa, creating a lobular pattern. The treatment goal is resection in combination with chemotherapy, showing a 5-year survival rate of 70%. Radiotherapy is not indicated because it has proved to carry the potential for the occurrence of a secondary malignant neoplasm. Also, radiation might interfere with facial growth. The presence of metastasis is considered the most important prognosis factor [54,55].

Another two entities have been reported in the literature to be remarkably rare in the mandibular condyle. These are multiple myeloma and malignant fibrous histiocytoma [56]. Multiple Myeloma is a malignant neoplasm of plasma cells, characterized by a proliferation of abnormal immunoglobulin-secreting plasma cells. Patients who have not been diagnosed yet with multiple myeloma in some cases might develop jaw lesions as the first sign of the disease. Plasmacytoma of bone from multiple myeloma is seen in radiographs as multiple radiolucent lesions in the jaw characterized as a “punch-out” lesion. Usually present with pain, bone swelling and function impairment. Histopathologic characteristics include massive diffuse stromal infiltration by monoclonal binucleate and trinucleate plasmacytes with basophilic cytoplasm, hypodense chromatin, and nucleus with multiple nucleoli. Long-term prognosis and low recurrence rates are the result of a multidisciplinary treatment involving

surgery, radiation, and chemotherapy. Early diagnosis is key. In some instances, a plasmacytoma can be the first sign of multiple myeloma. The 5-year survival rate of a solitary plasmacytoma is about 60-55% for multiple myeloma [57,58].

Malignant fibrous histiocytoma most commonly arises from soft tissue, tendons, bones and joints in upper and lower extremities of adults. It has a male predilection with higher incidence in the sixth decade of life. Only a few are reported in the literature. It is characterized by the presence of histocytes, fibroblast, and myofibroblast. Clinically, it resembles a solid mass. Radiographically, it appears as a lobulated mass with muscle density and peripheral solid portions, with calcification in some instances. Histopathologic features include a lesion filled with fibroblast and hystocite-like cells in different distribution, with spindle and round cells. It is classified into four subtypes, depending on their cellular component: storiform-pleomorphic (50%-60%), myxoid (25%), giant cell (5-10%), and inflammatory (5%). The treatment of choice is local excision and radiotherapy. Chemotherapy is not indicated. The overall 5-year survival rate is about 46% [59,60].

CONCLUSIONS

The mandibular condyle can be affected by multiple benign and malignant conditions. Most of the times, these conditions can presents with similar clinical manifestations. Therefore, understanding of its histologic and radiographic features is a key in their diagnosis which requires a proper knowledge by the surgical team in order to treat them properly.

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