DENTO-MAXILLARY RADIOLOGY AND IMAGING

Course 7

IMAGING INVESTIGATION OF MAXILLO-FACIAL BENIGN TUMORS

7.1. MORPHOPATHOLOGY

7.1.1. BENIGN TUMORS CHARACTERISTICS

Are non-cancerous masses with benign morphological characteristics also detected by imaging methods:

- **focal lesions;**
- **sharply delineated;**
- **+/- capsule;**
- **round/ovalary shape;**
- **slow growth rate;**
- **produce a "mass effect" that can compress surrounded tissues;**
- **without invasion/destruction.**

Internal structure is almost always homogeneous, typically but not always composed of cells which bear a strong resemblance to a normal cell type in their organ of origin. These tumors are named from the cell or tissue type from which they originate, followed by the suffix "-oma" (but not -carcinoma, -sarcoma, or

blastoma, which are generally cancers). Lipoma is a common benign tumor of fat cells, osteoma is a benign tumor of bone cells…

Evolution is almost always slowly with no locally tissular invasion. They will not metastasize and do not reappear after surgical resection.

Although most benign tumors are not life-threatening, many types of benign tumors have the potential to become cancerous and reappear after surgical resection being surnamed semi malignant.

7.1.2. BENIGN VERSUS MALIGN

Benign tumors are sharply delineated masses with internal structure almost always homogeneous. They produce a "mass effect" that can compress surrounded tissues without invasion/destruction. Also they will not metastasize and do not reappear after surgical resections.

7.1.3. BENIGN VERSUS CYST

Benign tumors are not fluid collection, with homogenous contrast uptake due to benign type of tumoral vascular supply. All imaging methods show no fluid characteristics but tissular type from which they originate: adipose-like, fibrous-like, bone-like, muscle-like…

7.2. RADIOGRAPHIC DIAGNOSIS

Benign tumors are radiographic detected as radioopacity or radiolucensy with benign morphological characteristics:

- **focal lesions;**
- **sharply delineated;+/- capsule;**
- **round/ovalary shape;**
- **slow growth rate;**
- **producing a "mass effect" that can compress regional surrounded tissues;**
- **without invasion/destruction of cortical bone,**
- **usually without periosteal reaction (except: osteoid osteoma and osteoblastoma).**

RADIOGRAPHIC - ASPECTS TYPES

According to lesional process predominance benign tumors may be:

- **PREDOMINANTLY OSTEOLITYC**
- **PREDOMINANTLY OSTEOSCLEROTIC**

PREDOMINANTLY OSTEOLITYC benign tumors are:

- **ameloblastoma,**
- **ameloblasticfibroma,**
- **giant cell granulomas,**
- **hemangiomas,**
- **mixomas,**
- **neurofibroma,**
- **fibroma,**
- **condroma,**
- **childhood melanotic neuroectodermal tumors,**
- **calcifying epithelial odontogenic tumors,**
- **pleomorf adenoma.**

PREDOMINANTLY OSTEOSCLEROTIC benign tumors are:

- **osteoma,**
- **exostoses,**
- **osteocondroma,**
- **osteoidosteoma,**
- **osteoblastoma,**
- **epulis,**
- **tori,**
- **coronoid hyperplasia of mandible.**

7.3. CT - SCAN DIAGNOSIS

Benign tumors appear as hyper or hypodense masses of tissular type density from which they originate: adipose- like, fibrous-like, bone-like, muscle-like with benign morphological characteristics:

- **focal lesions;**
- **sharply delineated;**
- **+/- capsule;**
- **round/ovalary shape;**
- **with slow growth rate;**
- **producing a "mass effect" that can compress surrounded tissues;**
- **without metastasize/invasion/destruction of cortex,**
- **with homogenous contrast uptake due to benign type of tumoral vascular supply;**

- the only internal inhomogeneities being due to calcifications, vascular lakes, secondary necrosis caused by a rapidly increasing volume.

7.4. MRI - EXAM DIAGNOSIS

Benign tumors appear as hyper T2, hypo T1 masses of tissular type signal from which they originate: adipose-like, fibrous-like, bone-like, muscle-like with benign morphological characteristics:

- focal lesions; sharply delineated;
- +/- capsule;
- round/ovalary shape;
- with slow growth rate;
- producing a "mass effect" that can compress surrounded tissues;
- without metastasize/invasion/destruction of cortex,
- with homogenous contrast uptake due to benign type of tumoral vascular supply;
- the only internal inhomogeneities being due to calcifications, vascular lakes, secondary necrosis caused by a rapidly increasing volume.

7.5. MAXILLARY BENIGN TUMORS CLASSIFICATION

Maxillary benign tumors may be morphopathological classified depending on:

- the tissue type from which they originate, or
- etiopatogenically depending on the odontogenic or non-odontogenic lesional character:

I. BENIGN ODONTOGENIC TUMORS
II. BENIGN NON - ODONTOGENIC TUMORS

I. BENIGN ODONTOGENIC TUMORS:

1. EPITHELIAL TUMORS
2. MESENCHYMAL TUMORS
3. MIXED TUMORS

1. EPITHELIAL TUMORS:

- ameloblastoma,
- squamous odontogenic tumors,
- calcifying epithelial odontogenic tumors,

◆clear cell odontogenic tumors,
◆adenomatoid odontogenic tumors.

2. MESENCHYMAL TUMORS:
◆odontogenic myxoma,
◆central odontogenic fibroma
◆cementifying fibroma,
◆cementoblastoma,
◆periapical cemental dysplasia.

3. MIXED TUMORS :
◆odontomas,
◆ameloblastic fibromaand ameloblastic fibro-odontoma.

II. BENIGN NON - ODONTOGENIC TUMORS:
1. BENIGN OSTEOGENIC TUMORS
2. BENIGN NON - OSTEOGENIC TUMORS

1. BENIGN OSTEOGENIC TUMORS:
◆osteoma
◆osteoid osteoma
◆osteoblastoma
◆exostoses and tori
◆coronoid hyperplasia of mandible.
◆osifying fibroma
◆fibrous dysplasia
◆chondroma
◆osteochondroma
◆benign chondroblastoma

2. BENIGN NON - OSTEOGENIC TUMORS:
◆fibroma (unspeciffying, unosifying, desmoplastic)
◆hemangioma of bone (capillary, cavernous, hemangiopericytoma)
◆mixoma
◆neurogenic tumors (neuroma, neurofibroma, neurinoma)
◆giant cells lesions (central giant cell granulomas, giant cell tumors, cherubism)
◆idiopathic histiocytosis (chronic localized-eosinophilic granuloma, chronic disseminated - Hand-Schüller-Cristian syndrome, acute disseminated - Letterer-Siwe disease)

7.6. BENIGN ODONTOGENIC TUMORS

They are tumors with dental origins derived from different tissular elements epithelial or mesenchymal or both that are part of tooth-forming apparatus, being therefore localized exclusively in the mandible and maxilla (also gingiva on rare occasions).

7.6.1. EPITHELIAL TUMORS

AMELOBLASTOMA (ADAMANTINOMA)

It is a benign tumor with malignant local character originated from the epithelium that is involved in teeth formation.

Sometimes may originated from the epithelial lining of odontogenic cysts, especially the dentigerus cysts.

It Is a lesion of young adult which are deforming the cortical bones, push and disorganized the regional teeth usually with secondary risalysis.

There are known three principals forms:

- cystic,
- solid,
- mixed,

They often reappear after surgical resection and may suffer a malignant transformation often after radiotherapy.

IMAGING DIAGNOSIS

Radiographic may be detected four characteristic lesional types:

- multilocular, most often,
- unilocular in children,
- trabecular septated and
- solid, both less frequent.

Classic radiographic aspect is the multilocular one, with small multiple radiolucencies as soap bubbles.

CT-scan and MRI-exam establish the positive and differential diagnosis and also clearly detect the regional lesional extension.

SQUAMOUS ODONTOGENIC TUMORS

Involve the alveolar process usually appearing in the anterior maxillary region or in posterior mandibular region.

They are considered to derive from epithelial rests of Malassez.

Radiographic appear as a crescent radiolucency with net outline associated to the regional dental roots.

CALCIFYING EPITHELIAL ODONTOGENIC TUMORS

Also named Pindborg tumors, are formed by the fusion of adamantine epithelium with oral epithelium from abnormal proliferation of intermediary layer.

Radiographic appear as radiolucency "honeycomb" lesion with internal calcification radiopaque island.

CLEAR CELL ODONTOGENIC TUMORS

Are aggressive, rarely tumors, appearing in female of over 60 years of ages which frequently metastasize in lungs and in regional lymph nodes.

Radiographic appear as a diffuse outlined radioopacity with local aggressive potential.

ADENOMATOID ODONTOGENIC TUMORS

They are considered to be an ameloblastoma subtype named due to intralesional presence of ductal and glandular structures.

Radiographic appear as a unilocular net outlined radiolucency around the crown of an impacted tooth.

They may also contain disseminated focal radioopacities due to the presence of some intratumoral enamel islands.

7.6.2. MESENCHYMAL TUMORS

ODONTOGENIC MYXOMA

It is a mesenchymal proliferation imitating dental pulp structure or follicular conjunctive tissue.

Although considered a benign tumor it has infiltrative and aggressive capacities and may also regional recur.

Radiographic is identified as a multilocular radiolucency, often "honeycomb" lesion, which secondary may determine cortical expansions and radicular displacement.

CENTRAL ODONTOGENIC FIBROMA

It is the central variant of peripheral odontogenic fibroma, radiographic detected as a multilocular radiolucency which secondary determine cortical expansions.

CEMENTIFYING FIBROMA

It seems impossible to differentiate cementifying fibroma from ossifying fibroma being probably a central variant of fibroma which contains calcifying material microscopically detected as cement.

Radiographic is revealed as a net outlined radiolucency with internal focal radioopacities or the lesions may appear diffuse calcificated.

CEMENTOBLASTOMA

Or true cementoma is a rarely benign tumor with cementoblast origin appearing in young adults, in closed relation with a vital tooth root.

Radiographic appear as a radioopacity that replace the root of the tooth usually surrounded by a radiolucent ring.

PERIAPICAL CEMENTAL DYSPLASIA

Also named cementoma appear as an abnormal reaction of periapical bone and of the cementum to commune factors like traumas and infections.

Radiographic appear as a periapical radiolucency of a vital tooth which in time become a solid radiopaque mass.

A rare condition described as florid osseous dysplasia appears to be an exuberant form of periapical cemental dysplasia radiographic appearing as diffuse radiopaque mass throughout the jaw.

7.6.3. MIXED TUMORS

ODONTOMAS

Are known as mixed odontogenic tumors compound from tissue with both epithelial and mesenchymal origin, their development lead to enamel deposition done by ameloblasts and dentin deposition done by odontoblasts.

May appear as:

- **a compound form with multiple miniature or rudimentary teeth,**
- **a complex form with amorphous conglomeration of hard tissue.**

- the mixed form, the association of the two forms with an peripheral compound odontoma and a central dense core.

Radiographic the lesional radioopacity is surrounded by a marginal radiolucent halo.

AMELOBLASTIC FIBROMA AND AMELOBLASTIC FIBRO-ODONTOMA

Appear as benign proliferations of odontogenic origin with similar clinico-biological characters, radiographic are uni or multilocular, net outlined with a sclerotic margin.

Ameloblastic fibro-odontoma is detected as a radiopaque odontoma surrounded by a radiolucent ring. Ameloblastic fibroma appear only as a radiolucent lesion.

Any form may be associated with the crown of an impacted tooth.

7.7. BENIGN NON - ODONTOGENIC TUMORS

They are benign space occupying masses which have not the origin in tooth-forming apparatus. These have different tissular etiopathogenies, specific for anatomic component elements of maxillary region: bone tissue, muscular tissue, fibrous tissue, neurovascular structures, etc.

Benign non - odontogenic tumors are also divided in:

1. **BENIGN OSTEOGENIC TUMORS**
2. **BENIGN NON - OSTEOGENIC TUMORS**

7.7.1. BENIGN OSTEOGENIC TUMORS

Are characterized by primordial presence of ossifying component, calcifyied, of neoformation tissue, being imaging detected by benign model of tumoral osteogenesis:

- focal osteolysis or osteosclerosis,
- net outlined,
- +/– marginal osteosclerosis,
- homogenous contrast uptake,
- slow progressive evolution with deformations and thinning of the regional cortical bone.

There are identified the following histological types:

- **OSTEOMA**
- **OSTEOID OSTEOMA**
- **OSTEOBLASTOMA**

- EXOSTOSES AND TORI
- CORONOID HYPERPLASIA OF MANDIBLE
- OSIFYING FIBROMA
- FIBROUSDYSPLASIA
- CHONDROMA
- OSTEOCHONDROMA
- BENIGN CHONDROBLASTOMA

OSTEOMA

It's a benign tumor with slow growth appearing usually at the viscerocranium level, compound from mature bone tissue:

- cancellous – cancellous osteoma or

- compact – compact osteoma.

Usually appear solitary as a single lesion - or may occur associated with Gardner syndrome, malformation due to a chromosome abnormality at the long arm of the fifth pair characterized by: multiple osteomas, intestinal polyposis, skin fibromas, epidermal cysts, odontomas and impacted permanent and supernumerary teeth.

As topographic variants are described the following types of localizations:

- endosteal or

- periosteal,

Radiographic is detected as a radiopaque focal mass, net outlined, of cancellous or compact bone, with or without marginal sclerosis.

OSTEOID OSTEOMA

It's a lesion related with osteoblastoma, some authors believing that exist a direct relation between a trauma and a regional inflammation in antecedents.

Clinico-radiologic aspect is often like chronic sclerozing osteomyelitis, being formed by a central region compound of osteoid tissue with different degrees of calcification surrounded peripherally by a osteocondensing osseous tissue.

Radiographic appear as a focal, ovalary, milimetric, radiolucency surrounded by a marginal osteosclerotic ring of about 2 cm diameter. Central tumoral "nidus"may have different degrees of calcifications till the aspect of a central compact radioopacity.

Commented [s1]:

OSTEOBLASTOMA

A benign tumor of the child and young adult, often described as a giant osteoid osteoma.

It's also often considered to be an osteoid osteoma with more than 2 cm in diameter to which is missing the peripheral mature compact tissue.

Radiographic is detected as a peripheral radiolucency which surrounds a osseous tissue mass with different degrees of calcifications. Marginal osteosclerotic ring is missing, in rare cases, the presence of a malign type periosteal reaction in ''divergent sunray'' delimit aggressive forms of disease.

EXOSTOSES AND TORI

They are nodular protuberances compound from mature bone appearing as net outlined islands of osseous tissue with maxillary localizations.

Exostoses appear as a dense cortical type radioopacity attached to the alveolar process of maxilla usually localized at the level of maxillary tuberosity.

Torus palatines is a sessile, nodular mass of bone that presents along the midline of the hard palate, radiographic detected as diffuse radiopaque bilateral nodular lesions symmetric localized along the midline.

Torus mandibularis are bony exophytic growths that present along the lingual aspect of the mandible, superior to the mylohyoid ridge limiting the tongue movement and making impossible prosthetic device mandibular adaptation to edentate.

CORONOID HYPERPLASIA OF MANDIBLE

With different etiopathogenies usually appear bilateral, as a regional hyperplasia, X-ray exam identifying a coronoid process ''in mushroom'' shape or unspecific deformed, with secondary mandible movement limitation.

OSIFYING FIBROMA

It's a fibrous osteoma form identified as a benign fibro-osseous lesion of periodontal ligament with clinico-radiologic aspect of cementifying fibroma due to simultaneously intratumoral presence of osseous, fibrous and cementum elements.

Radiographic is detected as a uni or multilocular radiolucency within time different calcificated deposits which may have a progressive evolution leading to an inverted lesional aspect of central radiopaque mass surrounded by a radiolucent ring.

FIBROUS DYSPLASIA

It's an idiopathic lesion characterized by medullary proliferation of abnormally fibrous tissue containing various quantity of osteoid or osseous material resulted by regional metaplasia which:

- **invades cancellous osseous tissue**
- **deforming and thinning of regional cortical bone**
- **with no limits to the regional healthy osseous tissue.**

May affects one or more osseous segments being described as monoostotic fibrous dysplasia or polyostotic fibrous dysplasia.

FIBROUS DYSPLASIA – types

Polyostotic fibrous dysplasia form associated with pigmented skin lesions define Jaffe-Lichtenstein syndrome.

McCune-Albright syndrome associates: polyostotic fibrous dysplasia, pigmented skin melanotic type lesions – café au lait – and endocrine abnormality as girl precocious puberty, hyperthyroidism, acromegaly and hyperprolactinemia.

Lesional maxillary localization with polyostotic extension at the facial massive level known as cranio-facial dysplasia with alveolar secondary involvement will lead to dental displacement, malocclusion and abnormal dental mobility.

FIBROUS DYS PLASIA - radiographic aspect

Radiographic are described more lesional steps grouped in three distinct syndromes:

- **uni or multilocular radiolucent syndrome,**
- **mixed syndrome where the lesional radiotransparency are filled with multiple trabecular radioopacities resulting a ''ground glass'' aspect.**
- **mottled radiolucent syndrome associated with diffuse radioopacities similar to that noted in Paget disease.**

CHONDROMA

It's a benign tumor characterized by the replacement of osseous tissue with mature hyaline cartilaginous tissue.

May develop:

- peripheral – perichondroma or
- central – enchondroma, radiographically appearing as an irregular radiotransparency with internal multiple calcifictions.

Has a highly malignization potential, the malignant transformation moment being difficult to identify by imaging.

OSTEOCHONDROMA

Is formed by osseous and cartilaginous tissue being radiographic detected as a usually irregular radioopacity with internal radiotransparencies determined by cartilaginous tissue islands.

BENIGN CHONDROBLASTOMA

It's characterized by chondroid transformation of affected osseous segment associated with multiple osteoid or osseous elements and multiple internal calcifications.

Radiographic detected as a mixed image of a radiolucent lesion with multiple internal calcifications.

7.7.2. BENIGN NON - OSTEOGENIC TUMORS:

There were identified the following histological types:

- **fibroma (unspeciffying, unosifying, desmoplastic)**
- **hemangioma of bone (capillary, cavernous, hemangiopericytoma)**
- **mixoma**
- **neurogenic tumors (neuroma, neurofibroma, neurinoma)**
- **giant cells lesions (central giant cell granulomas, giant cell tumors, cherubism)**
- **idiopathic histiocytosis(chronic localized: eosinophilic granuloma, chronic disseminated: Hand - Schüller - Cristian syndrome, acute disseminated: Letterer - Siwe disease)**

FIBROMA (unspeciffying, unosifying, desmoplastic)

Represent a fibrous tissue proliferation at the level of osseous matrix, anatomo-pathologic distinguishing 3 different types:

- unspeciffying,
- unosifying,

- desmoplastic.

UNSPECIFFYING FIBROMA

It seems to be an intermediary form with frequent localization to the angle of mandible, often affecting young women.

Radiographic appear as a uni or multilocular radiolucency surrounded by a marginal osteosclerosis ring. The lesion resorbs regional teeth and lead to osteolysis by resorbtion of the involved maxillary osseous tissue.

UNOSIFYING FIBROMA

Has periosteal origin and cortical extends appearing as typical form of fibrous cortical defect. Usually found in children and young male adults, radiographically having the same aspect.

DESMOPLASTIC FIBROMA

It has also periosteal origin and produce important regional osteolysis having an important aggressive potential, usually without sex or age involvement predilection.

Radiographic has the same aspect but with an unusually expansive potential that may lead to cortical destruction and the involvement of regional muscle tissue.

HEMANGIOMA OF BONE
(capillary, cavernous, hemangiopericytoma)

They are considered rather vascular malformations than true tumors anatomo-pathologic being described 3 forms with distinct radio-clinic aspect:

- **capillary hemangioma,**
- **cavernous hemangioma,**
- **hemangiopericytoma.**

CAPILLARY HEMANGIOMA

Appear at the level of any maxilla-facial segment being characterized by muriform capillary crowding.

Radiographic is detected as osteolysis type of radiotransparency with reticulate specific aspect.

CAVERNOUS HEMANGIOMA

Appear as intraosseous endogen vascular structures proliferation with resulting blood filled vascular cavities with a lining vascular endothelium.

Radiographic appear as a complex osteolytic lesion radio transparent with internal radiopaque septum and ''grill'' aspect.

HEMANGIOPERICYTOMA

Derived by tumor proliferation of highly specialized cells from the vascular walls named pericytes has important aggressive potential, being described recurrences after surgical resection and regional or distant metastasize.

Radiographic appear as focal radiotransparency with expansion characters.

MIXOMA

It's a benign aggressive tumor with invasive character that replaces the osseous regional tissue with a gelatinous mucoid mass, deforming and thinning the osseous cortical.

In association with fibroma appear a mixed tumor named fibromixoma.

Radiographic appear as a focal, solitary, ovalary shape radiotransparency with multiple internal septum and a ''tennis racket'' aspect.

NEUROGENIC TUMORS
(NEUROMA, NEUROFIBROMA, NEURINOMA)

Appear by abnormally tissular proliferation detected on a peripheral nerve trajectory.

Hysto-pathologic were described 3 forms:

- **amputation neuroma,**
- **neurofibroma**
- **neurinoma**

AMPUTATION NEUROMA

Appear accidental after dental extractions, traumas or tumor ablation. It is not a true tumor rather a repairing proliferation of regional axonal cells.

Appear as a nodular form, +/- capsule, with slowly growth, rarely over one cm in diameter. Radiographic is detected as a net outlined radiolucent lesion with regional specific localization.

NEUROFIBROMA

Caused by neurofibromatous proliferation localized on a peripheral nerve trajectory, in cases of intraosseous localizations determined some dilatations productions:

- fusiform or
- lacunar

round – ovalary, net outlined and with intact cortical bone.

NEURINOMA

It's a tumor proliferation of Schwann cells always appearing on a peripheral nerve trajectory, without sex or age predilected involvement, very often found in patients with neurofibromatosis.

Radiographic is detected as a net outlined radiolucent lesion localizated along a peripheral nerve trajectory.

GIANT CELLS LESIONS (CENTRAL GIANT CELL GRANULOMAS, GIANT CELL TUMORS, CHERUBISM)

Known as myeloplax lesions, characterized a group of morbid entities with various degrees o aggressivity, from granuloma to benign tumor, highly invasive and recurrence.

CENTRAL GIANT CELL GRANULOMAS

Appear after repeated chronic microtraumas on a capillary vascular deficiency background, etiopathogenic an endocrine cause was identify, young women being predilected involved.

Radiographic is detected similar lesional aspect as in ameloblastoma with uni or multilocular radiolucency, in time transformed in "honeycomb" aspect due to internal reticulated radiopaque deposits material.

GIANT CELL TUMORS

Or myeloplax tumors are true tumoral proliferation with mesenchymal macrophage related cells, being so really differentiated from granulomas which are reactive proliferation.

Radiographic are detected uni or multilocular radiolucency with regional intraosseous localization which produce displacement and resorbtion of adjacent dental segments.

They must be clinico-radiographic and histologic differentiated from other pathogenic entities compound from giant cell: brown tumors from hyperparathyroidism, osteolytic lesions from cherubism and aneurysmal osseous cyst, Paget disease.

IDIOPATHIC HISTIOCYTOSIS (CHRONIC LOCALISED: EOSINOPHILIC GRANULOMA, CHRONIC DISEMINATED: HAND - SCHÜLLER - CRISTIAN SYNDROME, ACUTE DISEMINATED: LETTERER -SIWE DISEASE)

Are non - lipid reticuloendothelyosis characterized by a normal circulant cholesterol phagocytosis by endothelial cells.

There were described 3 syndromes:

- **chronic localized: eosinophilic granuloma,**
- **chronic disseminated: Hand - Schüller - Cristian disease,**
- **acute disseminated: Letterer - Siwe disease,**

considered as different clinic expressions of the same morbid entity.

IDIOPATHIC CHRONIC LOCALISED HISTIOCYTOSIS

Eosinophilic granuloma is the mildest form of the disease without systemic manifestations and predominantly involvement of bone and soft tissues: gingiva, oral mucosa, skin and lung inphiltrates.

Radiographic are detected as a solitary polylobulated irregular expansible radiolucency, which over a dimensional threshold are similar to ameloblastoma radiographic aspect.

IDIOPATHIC CHRONIC DISEMINATED HISTIOCYTOSIS

Hand - Schüller - Cristian disease, is a systemic dissemination of initial eosinophilic granulomas type. Clinic the classic triad is present: bone lesions, exofthalmous, insipid diabetes.

Radiographic appear an increase of periodontal space with the image of suspended teeth, the initial radiolucency being extended under a lacunar form beyond the perforated osseous cortical. Due to secondary osteolysis calvaria appear as “geographic map“.

IDIOPATHIC ACUTE DISEMINATED HISTIOCYTOSIS

Letterer-Siwe disease is the worst form with malignant proliferation aspect also named Langerhans cells disease.

Characterized by cranio-facial pluryfocal osteolysis, with regional lymphadenopathies, hepatosplenomegaly, fever, anemia, rash, cachexia, gastroenteritis, lung inflammations, medullary tumors that compress hematogenous medulla with secondary pancytopenia.

Radiographic it is hard to detect due to rapidly exitus evolution. Typical images are similar with those from osteomyelitis characterized by diffuse radiolucency with cancellous bone deleting and periapical localizations.