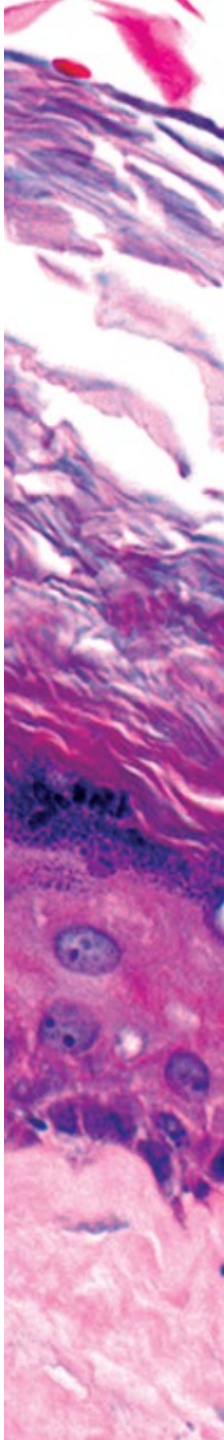
Krasnoyarsk State Medical University Named After Professor V.F. Voino-Yasenetsky
Department of Pathological Anatomy Named After Professor P.G.Podzolkov



Lecture 8.

Contents:

Diseases of the jaw bones and bones of the facial skeleton. Cleft lips and palate. Inflammatory lesions of the jaw bones: periostitis, osteitis, osteomyelitis. Tumor-like, idiopathic and hereditary diseases of the jaw bones. Non-odontogenic cysts of the jaws.

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Candidate of Medical Sciences (PhD),
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Introduction

Congenital malformations can be defined as structural or functional abnormalities (e.g., metabolic disorders) that occur during intrauterine development and can be detected before birth, during birth, or later in life.

Congenital malformations of the maxillofacial region occur in approximately 1 in 700 newborns.

The most severe anomalies in the development of the maxillofacial region occur at 4-8 weeks of embryonic development.

Congenital malformations of the maxillofacial region:

- cleft facial defects
- congenital fistulas and cystic formations;
- defects and deformities of the nose, ears, jaw bones, tongue;
- congenital tumors.

Cleft defects

The cause of the formation of a cleft lip is a violation of the fusion of the medial nasal process with the maxillary process.
Likewise, disruption of the formation and fusion of the palatine protuberances results in a palatal cleft. In about 45% of cases, there is a combination of a cleft lip with a cleft of the hard palate.
An isolated cleft lip occurs in 25%, a cleft palate in 30%. It is necessary to distinguish isolated clefts from clefts, which are an integral part of specific syndromes.

CLASSIFICATION SIGNS OF CLEFT

-cleft lip, hard palate, soft palate, uvula and / or alveolar ridge in various combinations;
-- hidden, complete or incomplete;
-- bilateral, one-sided or median;
-- the degree of decompensation of the closure of the palatopharyngeal valve.

In addition to the options presented, the so-called. a latent cleft of the palate, when only the muscles of the palate are cleaved within the muscular layer and the palatine bone with preserved mucous membrane.

CRANIOFACIAL SYNDROMES

Cruson's Syndrome.
Aper's syndrome.
Pfeifer's Syndrome.
Pierre-Robin syndrome.
Treacher Collins Syndrome.
These syndromes were discussed in detail in previous lectures.

INFLAMMATORY JAW DISEASES

Periostitis of the jaw

- inflammation of the periosteum of the alveolar arch (process), less often the body of the upper or lower jaw, infectious or traumatic genesis

ETIOLOGY OF PERIOSTITIS:

•Odontogenic (chronic periodontitis; alveolitis; inflammation of impacted teeth; periodontitis; festering cyst, etc.)
• Hematogenous (after suffering from tonsillitis, tonsillitis, otitis media, influenza, ARVI, scarlet fever, measles, etc.)
• Lymphogenous (after a sore throat, tonsillitis, otitis media, flu, scarlet fever, measles, etc.)
• Traumatic (after the removal of a complex tooth, surgery, tooth trauma, open fractures of the jaw, infected wounds of the soft tissues of the face, etc.)

Acute periostitis

Acute serous periostitis of the jaw is accompanied by infiltration of the periosteum and the accumulation in the inflammatory focus of a moderate amount of serous exudate.

Acute purulent periostitis of the jaw (flux) proceeds with the formation of a limited subperiosteal abscess, the formation of fistulas through which pus flows out.

Chronic periostitis

Chronic periostitis of the jaw is characterized by a sluggish infectious and inflammatory process in the periosteum, accompanied by the formation of young bone tissue on the surface of the jaw bones.

If, in case of simple periostitis of the jaw, the process of neoplasm of bone tissue is reversible, then with ossifying, ossification and hyperostosis rapidly progress.

By the degree of distribution it's can be limited (in the area of 1 or several teeth) and diffuse (covering almost the entire jaw) purulent periostitis.

Complications:

Osteomyelitis
Phlegmon
Sepsis

JAW OSTEOMYELITIS
several important aspects

The specific local immunological and microbiological aspects determine a major factor in the etiology and pathogenesis of this disease and hence also have a direct impact on its treatment; therefore, to extrapolate from long bone infections to disease of the jaws has limitations

This is reflected by the longstanding recognition of osteomyelitis of jawbones as a clinical entity which differs in many important aspects from that in long bones; hence, a wide variety of classifications, specifically for the jawbones, have been established

Classifications are based on different aspects such as clinical course, pathological–anatomical or radiological features, etiology and pathogenesis

A mixture of these classification systems is often used, leading to confusion, hindering comparative studies and obscuring classification criteria

The Zurich system of osteomyelitis is generally accepted as the most reliable classification for osteomyelitis - Classification is primarily based on clinical course and imaging

Zurich Classification:

The Zurich classification system of osteomyelitis is primarily based on clinical course and imaging

Histopathology is considered a secondary classification criterion because findings are mostly unspecific and inconclusive when considered by themselves; however, tissue examinations of biopsies are irreplaceable to confirm the diagnosis in cases of unclear and atypical clinical and radiological appearance and to exclude possible differential diagnoses

The three major classifications are:

- **Acute Osteomyelitis (AO)**
- **Secondary Chronic Osteomyelitis (SCO)**
- **Primary Chronic Osteomyelitis (PCO)**

Disease	*Clinical criteria*	*Etiology*
Acute osteomyelitis of the mandible	Symptoms of osteomyelitis not exceeding 4 weeks after onset of the disease	Well identified (trauma related, odontogenic, foreign body, transplant/implant induce)
SCO	Presence of pus, fistula and/or sequestration	Well identified (trauma related, odontogenic, foreign body, transplant/implant induce)
PCO	Absence of pus and fistula	Unknown

M. Julien Saint AmandN. SigauxA. GleizalP. BouletreauP.
Breton Chronic osteomyelitis of the mandible: a comparative study of 10 cases with primary chronic osteomyelitis and 12 cases with secondary chronic osteomyelitis (2017), http://dx.doi.org/10.1016/j.jormas.2017.08.006.

most common bacterial species

Bacteria genus	Number of patient from the PCO group	Number of patient from the SCO group
Streptococcus (alpha hemolytic)	8	9
Staphylococcus	7	9
Actinomyces	6	3
Fusobacterium	3	2
Prevotella	2	3
Neisseria	3	1
Aggregatibacter	2	1
Haemophilus	1	2
Eikenella	1	2
Veillonella	1	2
Kocuria	2	0
Parvimonas micra	0	2
Enterobacter	0	2

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primary chronic osteomyelitis

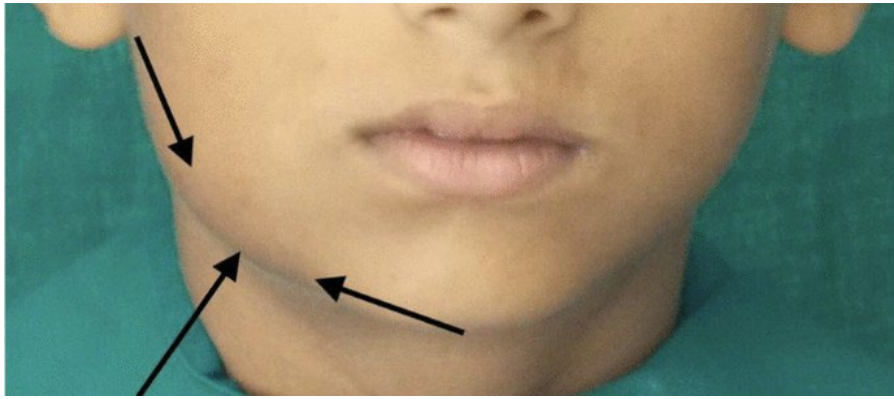
Primary chronic osteomyelitis of the jaw is a rare, non-suppurative, chronic inflammatory disease of unknown aetiology

Age at onset of symptoms demonstrated two incidence peaks.

Age at onset of symptoms (years)	No. of cases	% of cases
0–10	3	10
11–20	10	33
21–30	3	10
31–40	2	7
41–50	2	7
51–60	3	10
61–70	6	20
71–80	1	3
Total	30	100

Pain, local swelling and limited mouth opening were noticed as recurrent episodes and were the predominant clinical symptoms

Clinical symptoms	No. of cases	% of cases
Swelling	28	93
Pain	26	87
Limited mouth opening	17	57
Myofacial and/or TMJ pain	11	37
Hypoaesthesia (Vincent's symptom)	9	30
Lymphadenopathy	7	23



Mild buccal cortical bone expansion was evident in the right posterior region

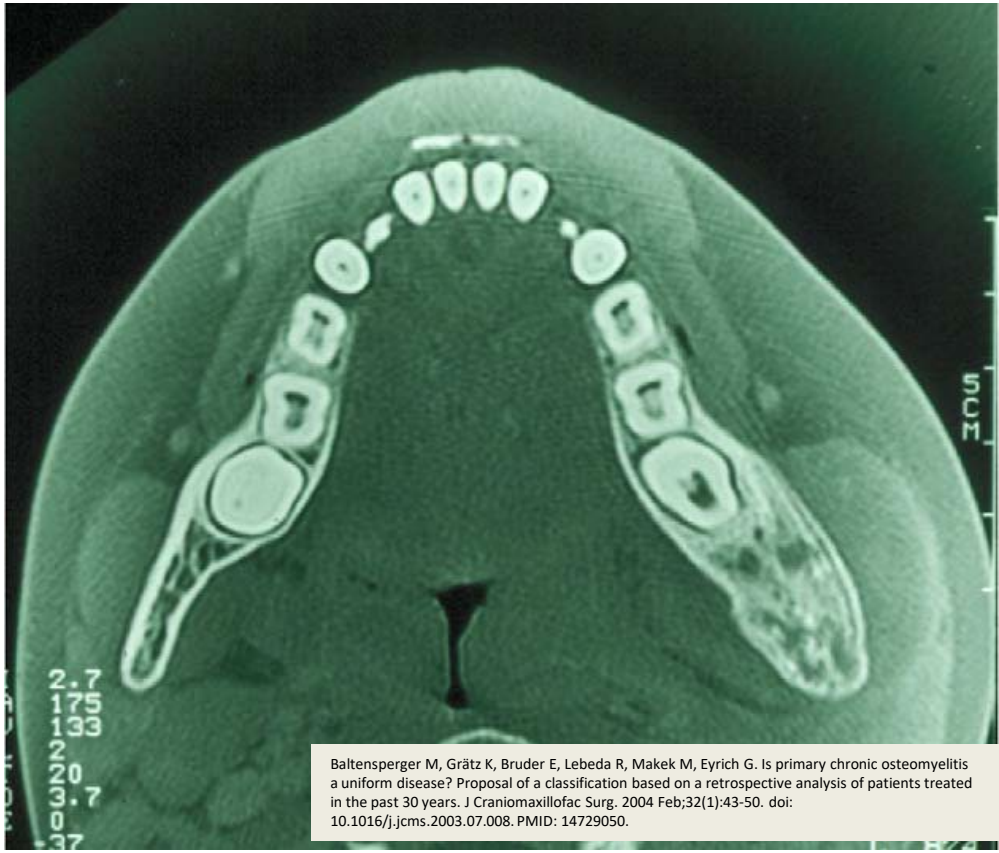
Kavya Priya, T., Hadhimane, A., Rai, K.K. *et al.* Deceptively complex diagnosis of early onset primary chronic osteomyelitis: a case report. *Bull Natl Res Cent* **45,** 64 (2021). https://doi.org/10.1186/s42269-021-00524-y

Baltensperger M, Grätz K, Bruder E, Lebeda R, Makek M, Eyrich G. Is primary chronic osteomyelitis a uniform disease? Proposal of a classification based on a retrospective analysis of patients treated in the past 30 years. J Craniomaxillofac Surg. 2004 Feb;32(1):43-50. doi: 10.1016/j.jcms.2003.07.008. PMID: 14729050.

The distribution of the primary chronic osteomyelitis of the jaws demonstrated an almost exclusive involvement of the mandible (any part).

All patients with primary chronic osteomyelitis underwent extensive imaging during their investigation, including conventional radiographs, and computerized tomographic scans in most cases.

Osteosclerosis was the most striking pattern seen in all patients in variable degrees. However, sclerosis was more predominant in adult cases.



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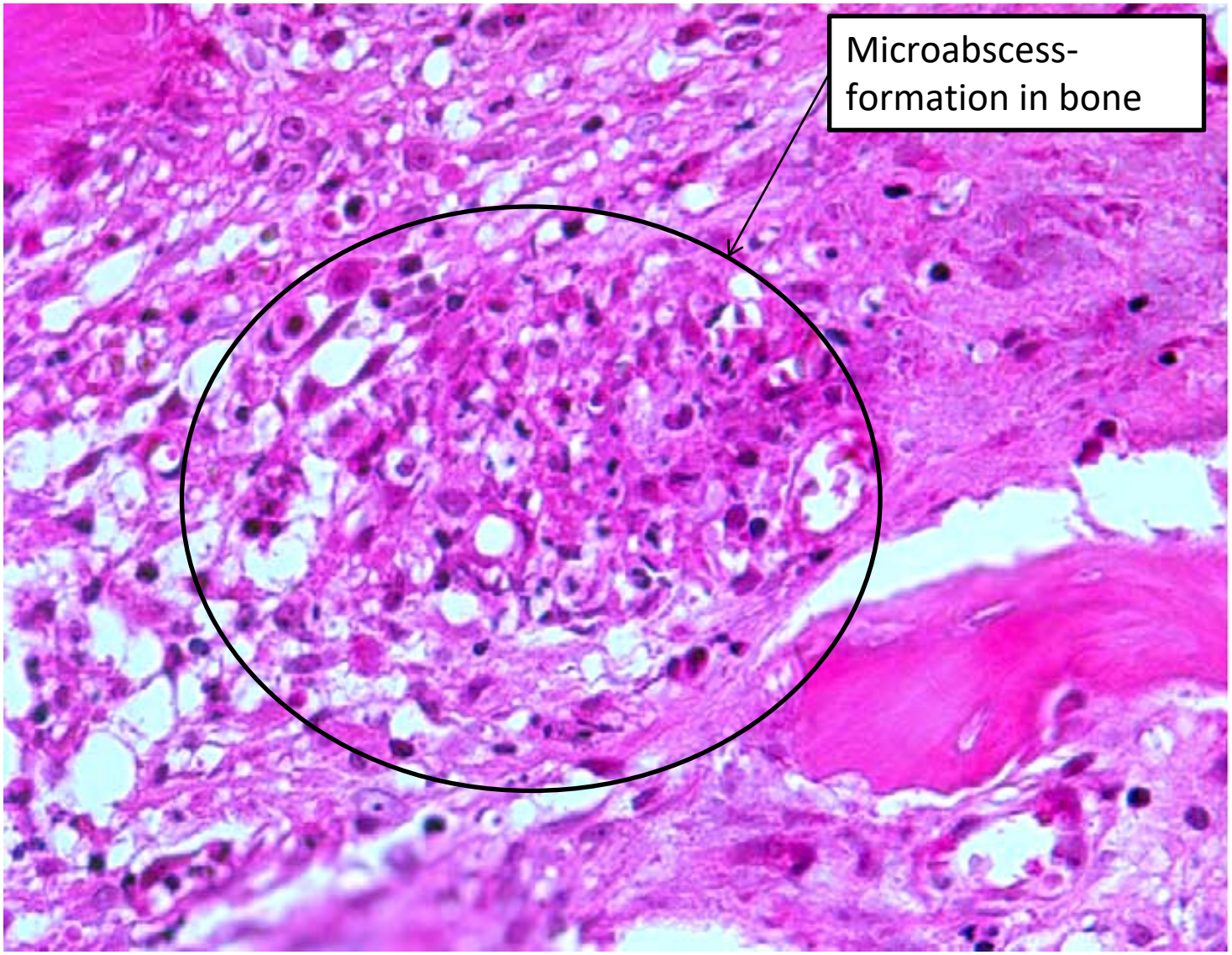
Patients with primary chronic osteomyelitis demonstrated a mild elevation of the erythrocyte sedimentation rate in more than half of the cases.
A similar slight increase of C-reactive protein values was noted.
The leukocyte count was normal in all but 2 patients, where a moderate elevation was noted.

Histopathology



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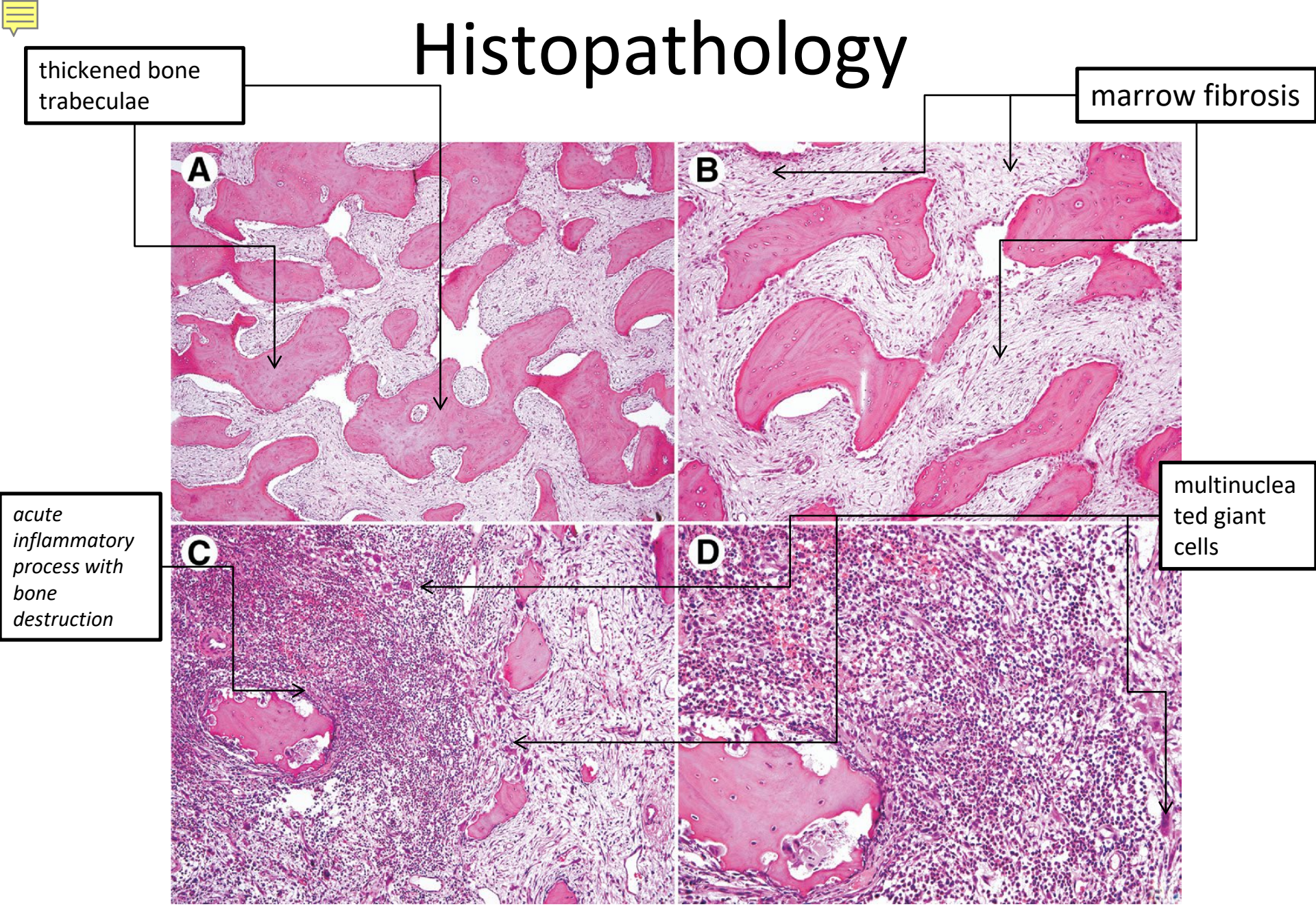
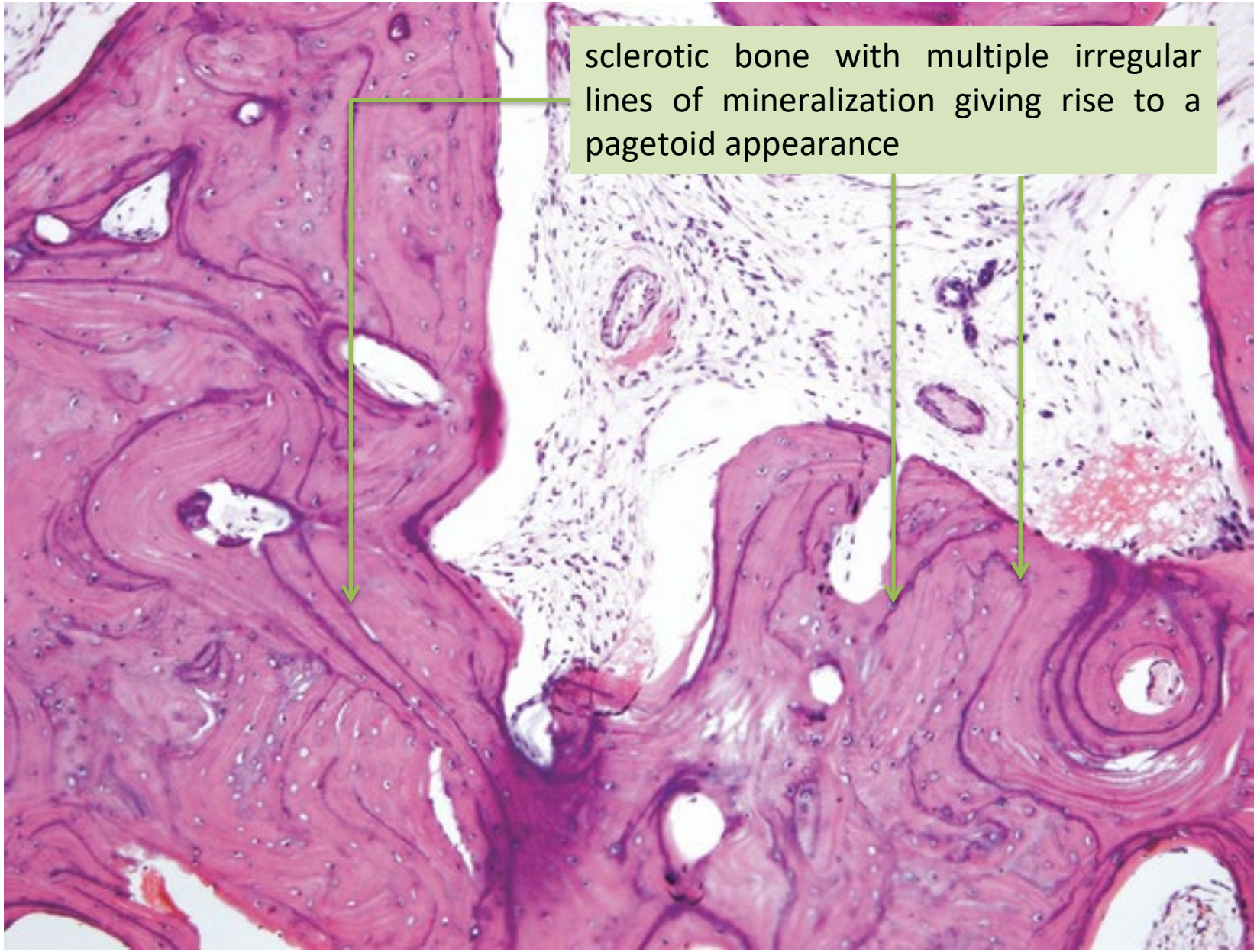
Histopathology



Bevin CR, Inwards CY, Keller EE. Surgical management of primary chronic osteomyelitis: a long-term retrospective analysis. J Oral Maxillofac Surg. 2008 Oct;66(10):2073-85.

Histopathology



Bevin CR, Inwards CY, Keller EE. Surgical management of primary chronic osteomyelitis: a long-term retrospective analysis. J Oral Maxillofac Surg. 2008 Oct;66(10):2073-85.

TREATMENT OPTIONS

1) **nonsurgical:** antibiotics, NSAIDS, hyperbaric oxygen therapy, bisphosphonate treatment, and muscle relaxants;

2) **surgical:** decortication alone, decortication with bone grafting, partial (marginal) resection, and segmental resection.

secondary chronic osteomyelitis (SCO)

chronic suppurative osteomyelitis, is characterized by the presence of pus, fistula and/or sequestration. SCO is usually caused by bacterial invasion from a well-identified etiology, such as trauma or surgery to the mandible.

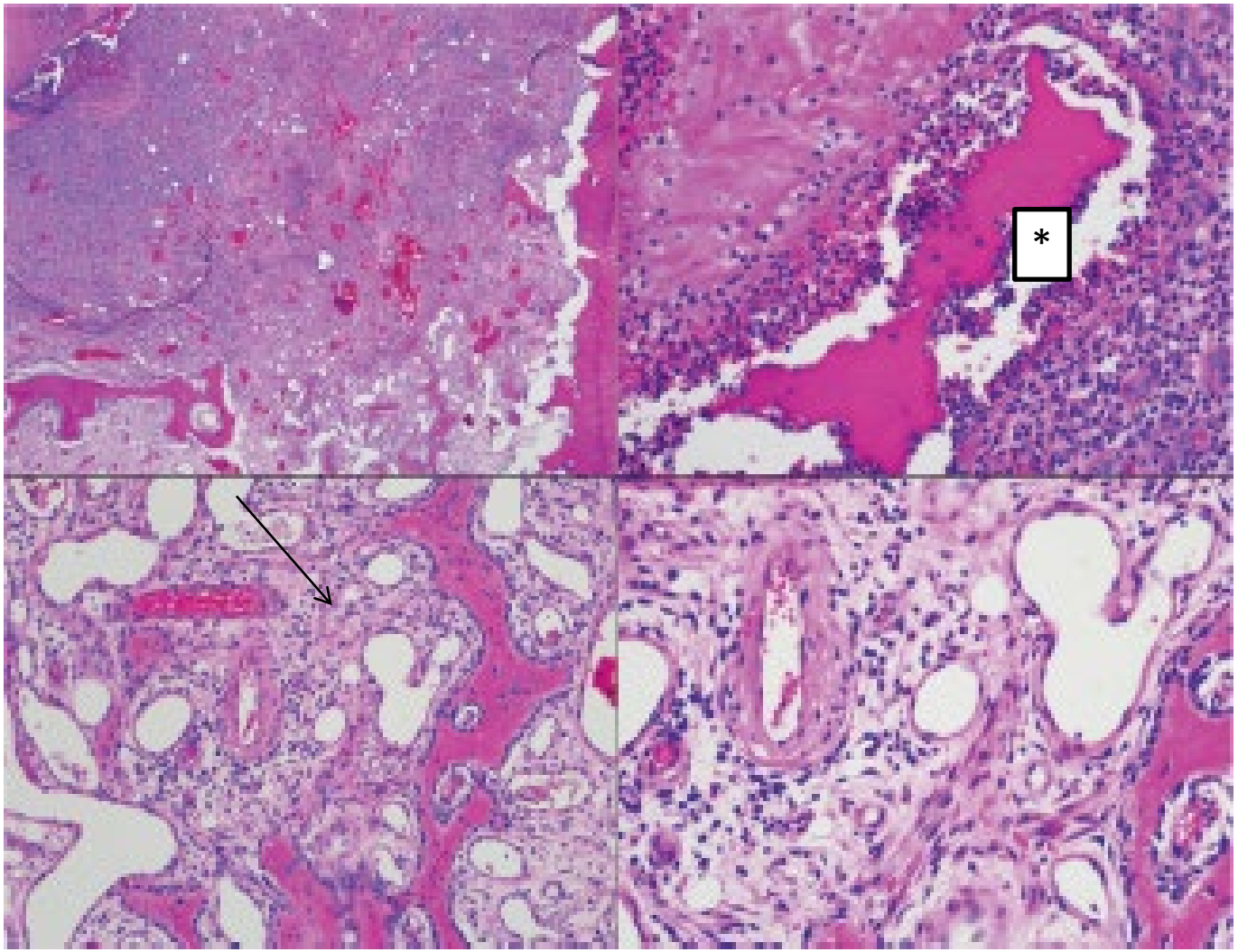
Acute and SCO of the mandible correspond to the same disease at a different stage, sharing the same etiology.

major differences between PCO and SCO

Criteria	*PCO group*		*SCO group*	*p value*
Clinical findings				
- Symptoms lasting longer than 2 years	100% (average 6.8 years)	>>	8.3% (average 0.7 years)	$p < 0.05$
Radiologic findings				
- *Sclerosis*	100%	>>	36.4%	$p < 0.05$
- *Bone hypertrophy*	50%	>>	0%	$p < 0.05$
- *Collection*	0%		27.3%	$p = 0.21$
Histology				
- Bone fibrosis	90%	>	42.9%	$p = 0.10$
Treatment				
- Decortication	100%		25%	$p < 0.05$
- Segmental resection	30%		0%	$p = 0.08$
- NSAIDS and/or steroids treatment	80%		0%	$p < 0.05$
- Persistence of symptoms at the end of follow-up	50%		8.3%	$p = 0.06$

M. Julien Saint AmandN. SigauxA. GleizalP. BouletreauP. Breton Chronic osteomyelitis of the mandible: a comparative study of 10 cases with primarychronic osteomyelitis and 12 cases with secondary chronic osteomyelitis (2017), http://dx.doi.org/10.1016/j.jormas.2017.08.006.

Histologic features of “acute” and “chronic” osteomyelitis exist in the same lesion.



Masters EA, Trombetta RP, de Mesy Bentley KL, Boyce BF, Gill AL, Gill SR, Nishitani K, Ishikawa M, Morita Y, Ito H, Bello-Irizarry SN, Ninomiya M, Brodell JD Jr, Lee CC, Hao SP, Oh I, Xie C, Awad HA, Daiss JL, Owen JR, Kates SL, Schwarz EM, Muthukrishnan G. Evolving concepts in bone infection: redefining "biofilm", "acute vs. chronic osteomyelitis", "the immune proteome" and "local antibiotic therapy". Bone Res. 2019 Jul 15;7:20. doi: 10.1038/s41413-019-0061-z. PMID: 31646012; PMCID: PMC6804538.

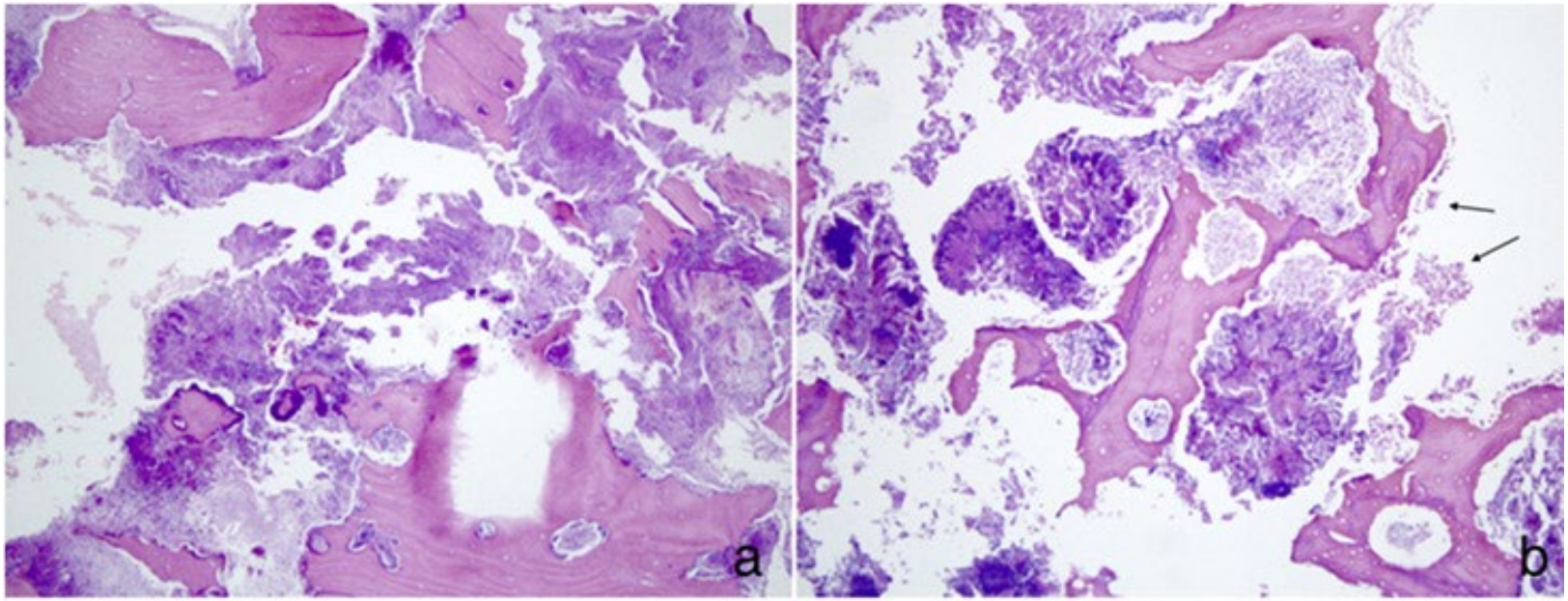
Acute osteomyelitis (AO)

•Early phase of osteomyelitis, usually suppurative (pus forming)

•Exists when an acute inflammatory process moves away from the site of initial infection and spreads through the medullary space of the bone and

•Acute phase may lead to the chronic phase which has been arbitrarily defined as an osseous infection lasting at least 1 month

Histopathologic examination of bone specimens coupled with bone culture is considered the gold standard for the diagnosis of osteomyelitis.



Microscopic (histologic) description

Neutrophils (may persist for weeks), lymphocytes and plasma cells with bone necrosis and reactive new bone formation

Capillary proliferation and fibrosis foamy macrophages)

Bone marrow space replaced by inflammatory tissue

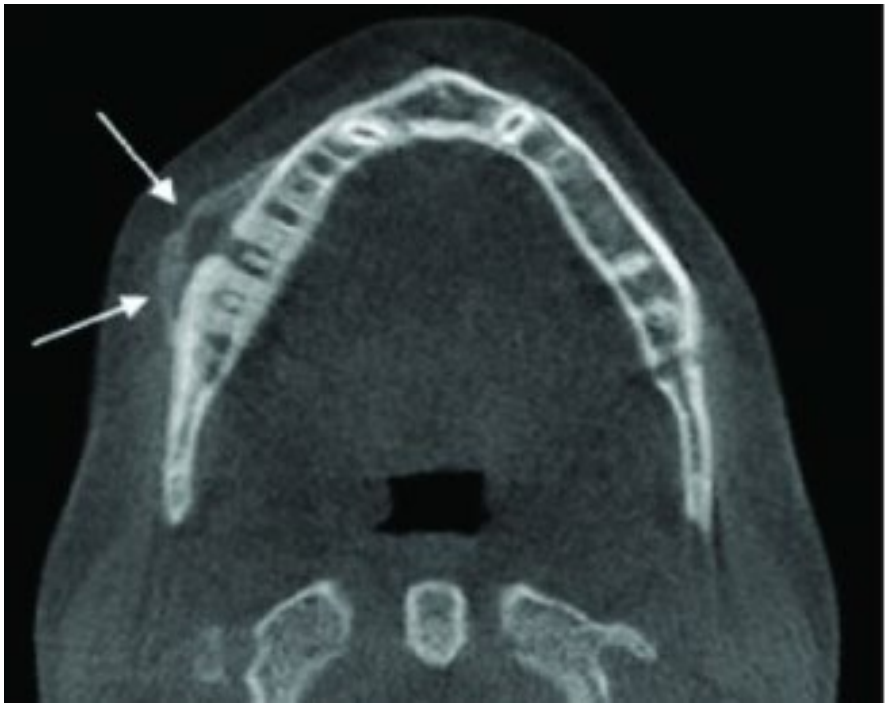
Subtypes include xanthogranulomatous osteomyelitis

Salmonella infection may produce tuberculoid granules with variable central necrosis

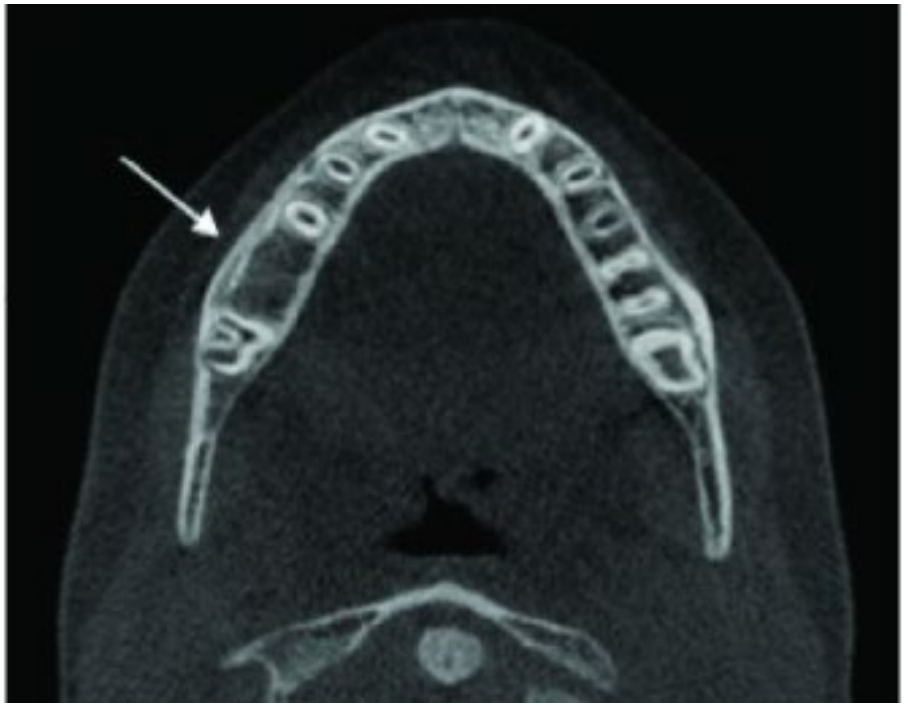
Shuster A, et al. Comparison of the histopathological characteristics of osteomyelitis, medication-related osteonecrosis of the jaw, and osteoradionecrosis, Int J Oral Maxillofac Surg (2018), https://doi.org/10.1016/j.ijom.2018.07.002

Garre's osteomyelitis

Garre's osteomyelitis is a localized periosteal thickening caused by mild irritation or infection



Axial and cross sections in CBCT showing new bone formation and a tunnel-like defect in the vestibule cortical surface of the inflamed bone starting from the apical region of tooth number 46



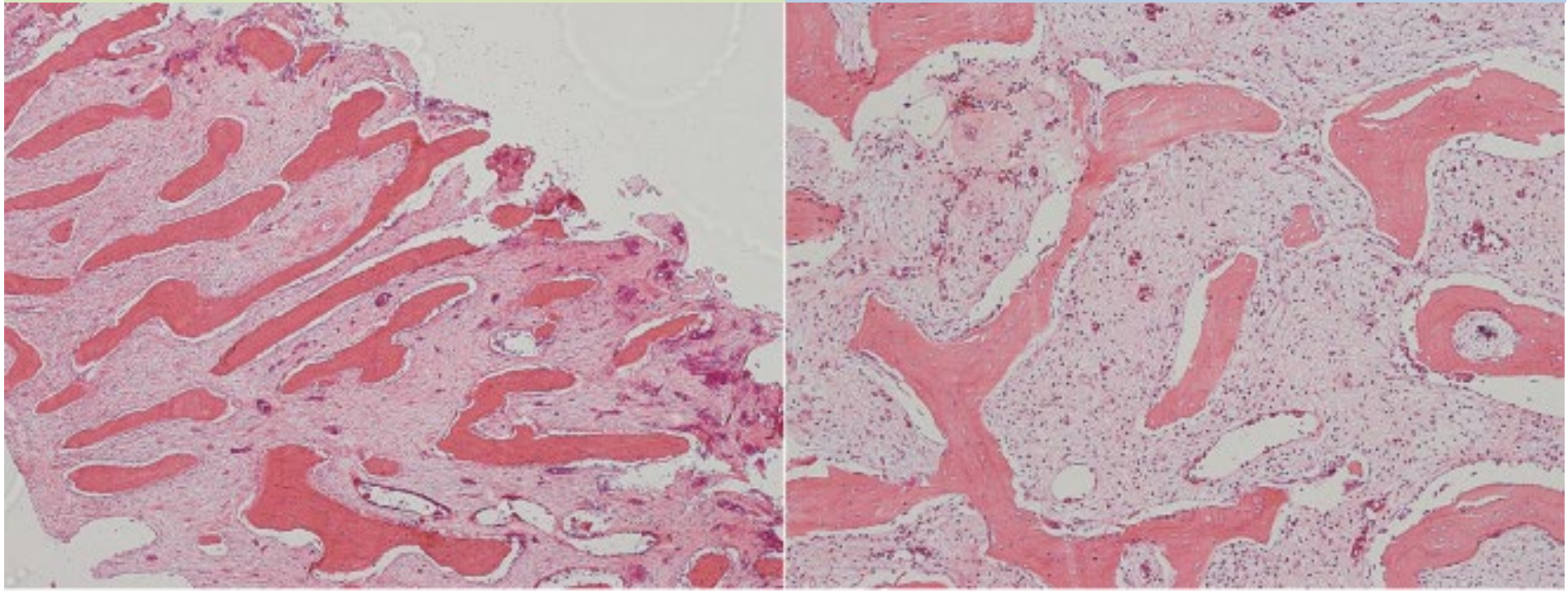
CBCT image showing decreased cortical bone thickness and the presence of the original cortex within the enlarged portion of the jaw in the postoperative control

Liu D, Zhang J, Li T, et al. Chronic osteomyelitis with proliferative periostitis of the mandibular body: report of a case and review of the literature. Annals of the Royal College of Surgeons of England. 2019 May;101(5):328-332. DOI: 10.1308/rcsann.2019.0021. PMID: 30855166; PMCID: PMC6513367.

Garre's osteomyelitis

Bony trabeculae of the reactive bone arranged perpendicular to the bone surface

Dense fibrous stroma with chronic inflammation in the intertrabecular space



Histological examination showed new bone formation with periosteal reactivity.

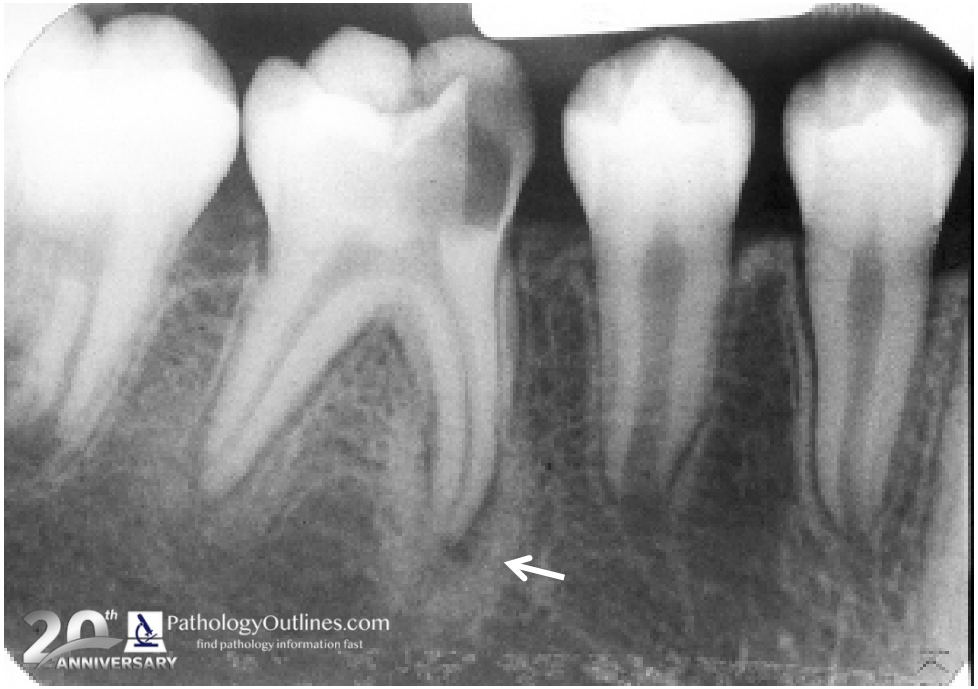
Liu D, Zhang J, Li T, et al. Chronic osteomyelitis with proliferative periostitis of the mandibular body: report of a case and review of the literature. Annals of the Royal College of Surgeons of England. 2019 May;101(5):328-332. DOI: 10.1308/rcsann.2019.0021. PMID: 30855166; PMCID: PMC6513367.

Condensing osteitis

Synonyms:
Focal sclerosing osteitis
Focal sclerosing osteomyelitis

Definition / general
Localized areas of radiographic bone sclerosis associated with apices of inflamed, dead or dying teeth (pulpitis or pulpal necrosis)
The association with an area of inflammation, usually the apex of an associated tooth, is critical, because these lesions can resemble several other intrabony processes that produce a somewhat similar radiographic pattern

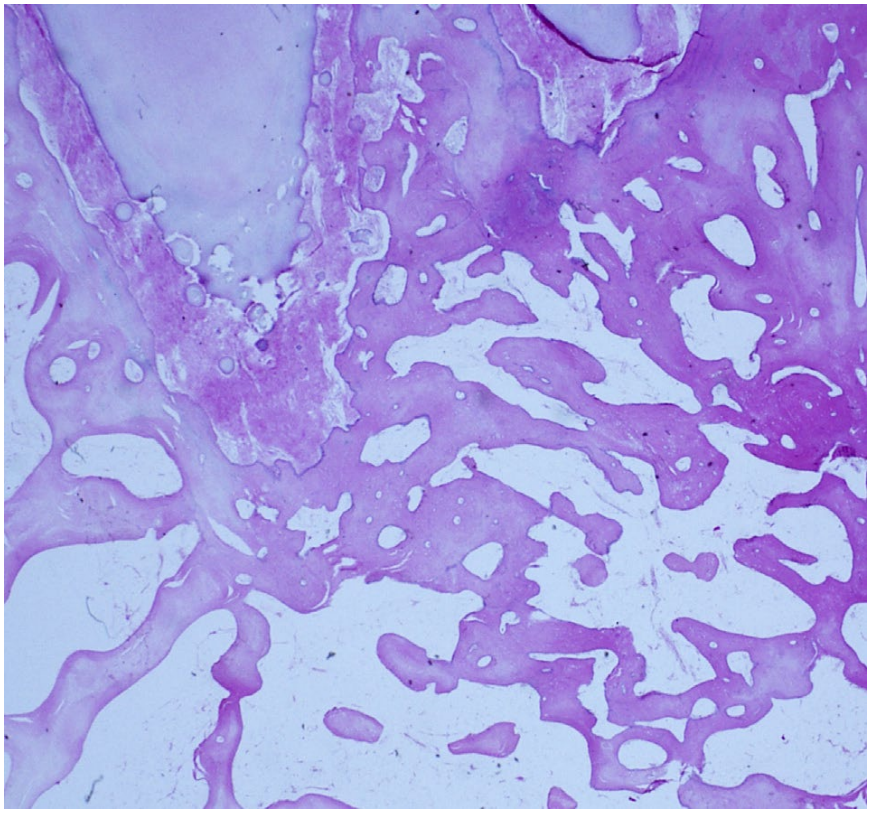
Pathophysiology / etiology
- osteoblastic response causing secondary sclerosis in response to a low grade inflammatory stimulus from an inflamed dental pulp



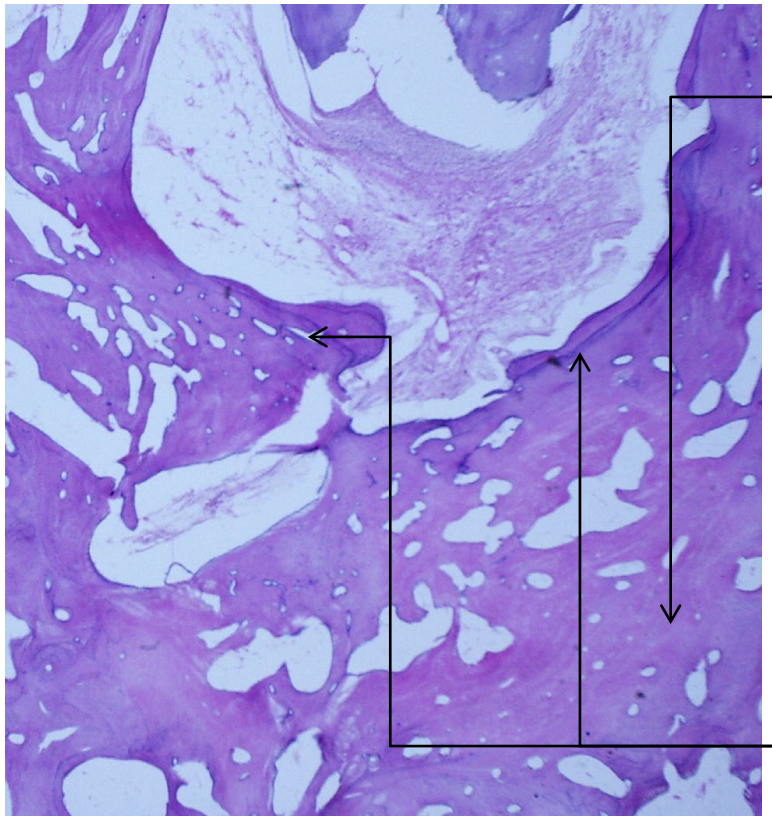
Radiology description
localized, usually uniform zone of increased radiodensity adjacent to the apex of a tooth

Morrison A. Condensing osteitis. PathologyOutlines.com website. https://www.pathologyoutlines.com/topic/mandiblemaxillacondensingosteitis.html. Accessed January 15th, 2022.

Condensing osteitis



Normal periapical region histologically.
There is a predominance of medullary bone, with the spaces being occupied by fatty marrow



Condensing osteitis. There is an increase in bone relative to the normal periapical findings. The bone shows a definitive incremental pattern

Green TL, Walton RE, Clark JM, Maixner D. Histologic examination of condensing osteitis in cadaver specimens. J Endod. 2013 Aug;39(8):977-9. doi: 10.1016/j.joen.2013.02.002. Epub 2013 Apr 25. PMID: 23880262.

Alveolar osteitis

Synonyms:
dry socket
ibrinolytic alveolitis

is inflammation of the alveolar bone (i.e., the alveolar process of the maxilla or mandible). Classically, this occurs as a postoperative complication of tooth extraction.

Incidence

range of 0.5 to 5% for routine dental extractions and up to 37% after removal of mandibular third molars

Pathophysiology

- After extraction of a tooth, a blood clot is formed at the extraction site with eventual organization of this clot by granulation tissue and gradual replacement by bone
- Destruction of the initial clot is thought to delay the aforementioned additional series of steps required for uneventful extraction site healing and leads to a clinical condition known as alveolar osteitis
- Clot is lost secondary to transformation of plasminogen to plasmin, with subsequent lysis of fibrin and formation of kinins which are potent pain mediators

Radiology description

Plain films may be suggested in the work-up of a dry socket to excluding the presence of radiographically detectable material retained in the socket, help exclude additional problems with neighboring teeth and to exclude detectable jaw fractures

Microscopic (histologic) description

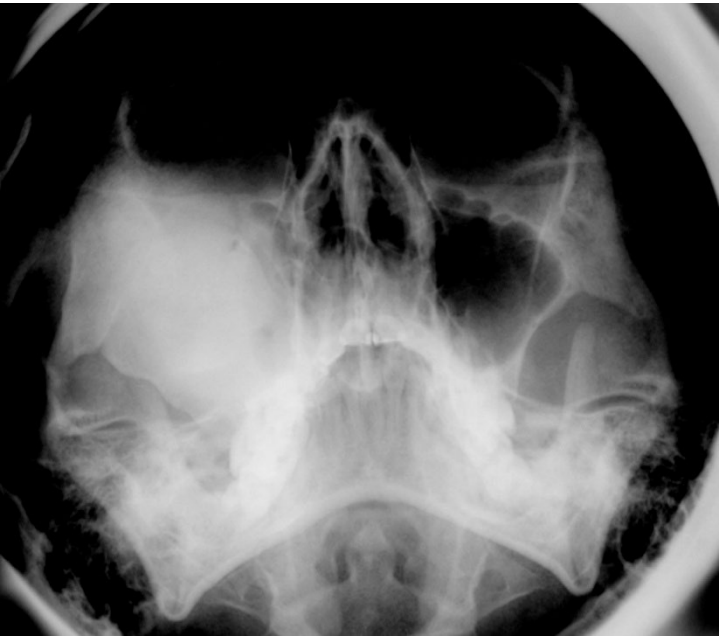
Uncommon to have a surgical specimen, because the procedure is debridement or removal of the residual clot
If material is submitted, may be composed of:

- Inflammatory cellular infiltrate with numerous phagocytes and giant cells in the remaining clot
- Viable and non-viable bone fragments

Fibrous dysplasia

Synonyms:
Craniofacial fibrous dysplasia

Fibrous dysplasia (FD) is a skeletal anomaly in which normal bone is replaced and distorted by poorly organized and inadequately mineralized immature bone and fibrous tissue.
It may involve a single bone (**monostotic** FD) or multiple bones (**polyostotic** FD)



fibrous dysplasia involving the right maxilla, showing ground-glass appearance, with indistinct borders.

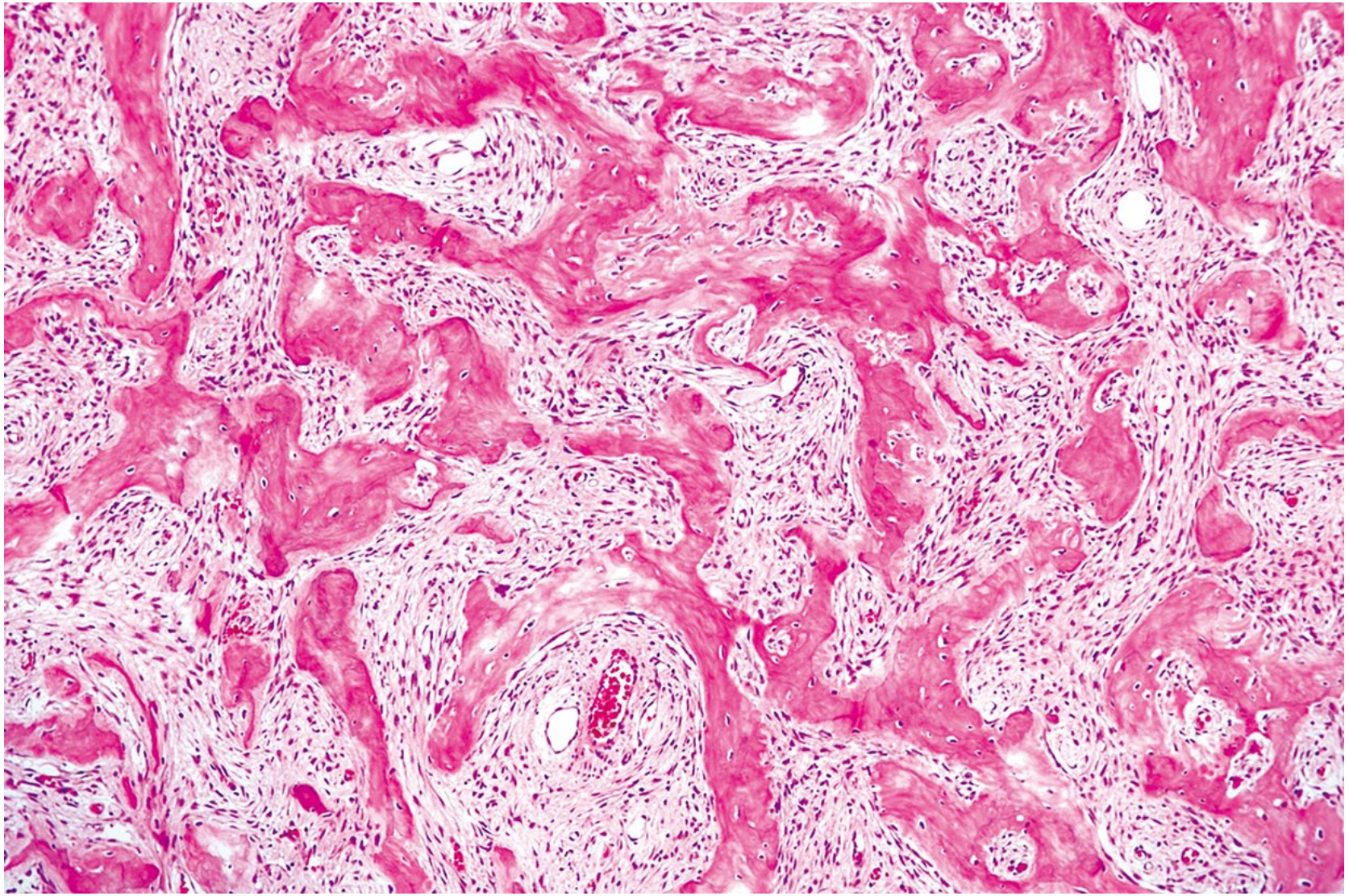
Localization: The cranio-facial bones and the femur are the two most common sites of both monostotic and polyostotic FDs

Epidemiology: 7% of all benign bone tumours

Etiology: postzygotic activating missense mutations in the GNAS gene

Prognosis and prediction
In most cases, the lesions seem to stabilize with skeletal maturation; therefore, surgical intervention in younger patients should be delayed for as long as possible

FD: Irregularly shaped trabeculae of woven bone, without osteoblastic rimming in afibroblastic cell-rich stroma.



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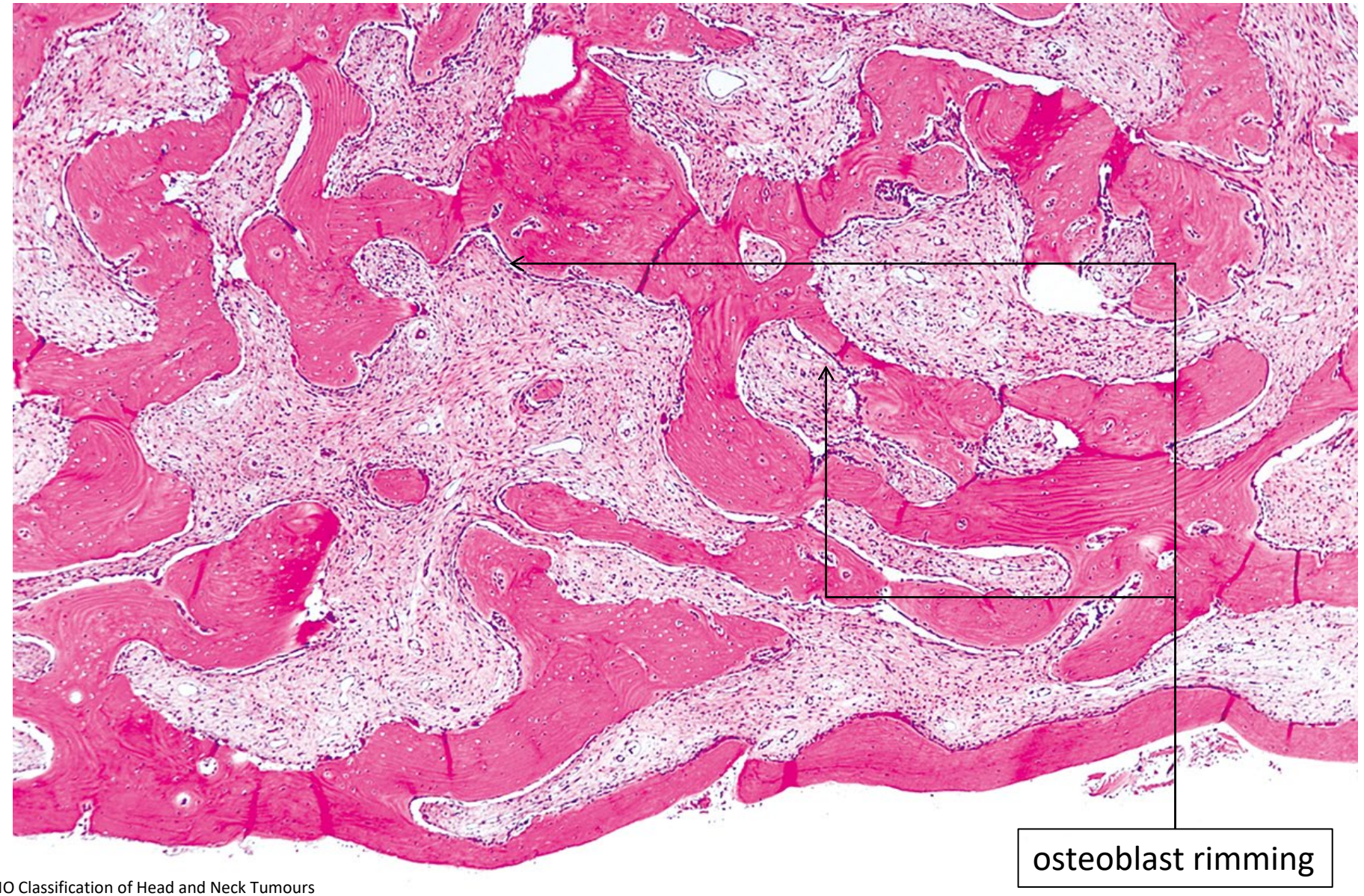
Progressive maturation to lamellar bone in a longstanding maxillary lesion of 30 years' duration; lamellar bone with osteoblast rimming is present.



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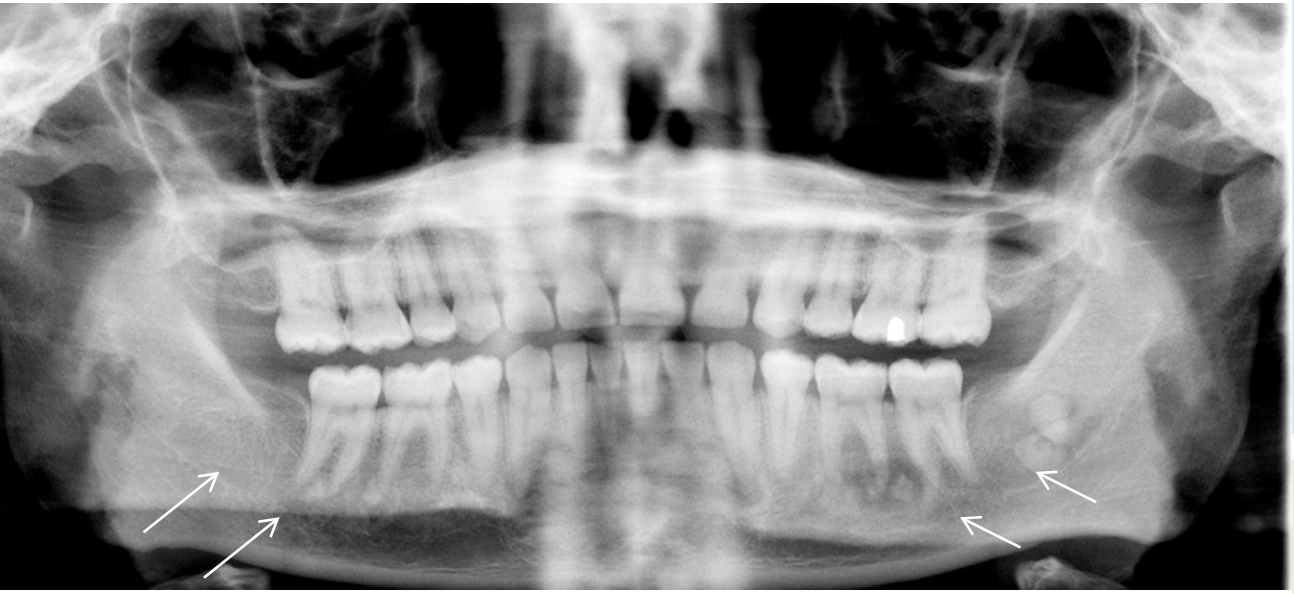
Cemento-osseous dysplasia (COD)

Synonyms:
osseous dysplasia;
cemental dysplasia;
cementoma

Cemento-osseous dysplasia (COD) is a non-neoplastic fibro-osseous lesion of the tooth-bearing regions of the gnathic bones.

Localization

COD occurs exclusively in the tooth-bearing regions of the jaws.



Radiography shows lesions of mixed radiolucent and radiopaque florid cemento-osseous dysplasia in both quadrants of the mandible.

COD

1. **periapical** COD is associated with the apical areas of mandibular anterior teeth;

2. **focal** COD is associated with a single tooth;

3. **florid** COD has multifocal (multiquadrant) involvement.

Epidemiology

-most common benign fibro-osseous lesion of the jaws.
-middle-aged Black women

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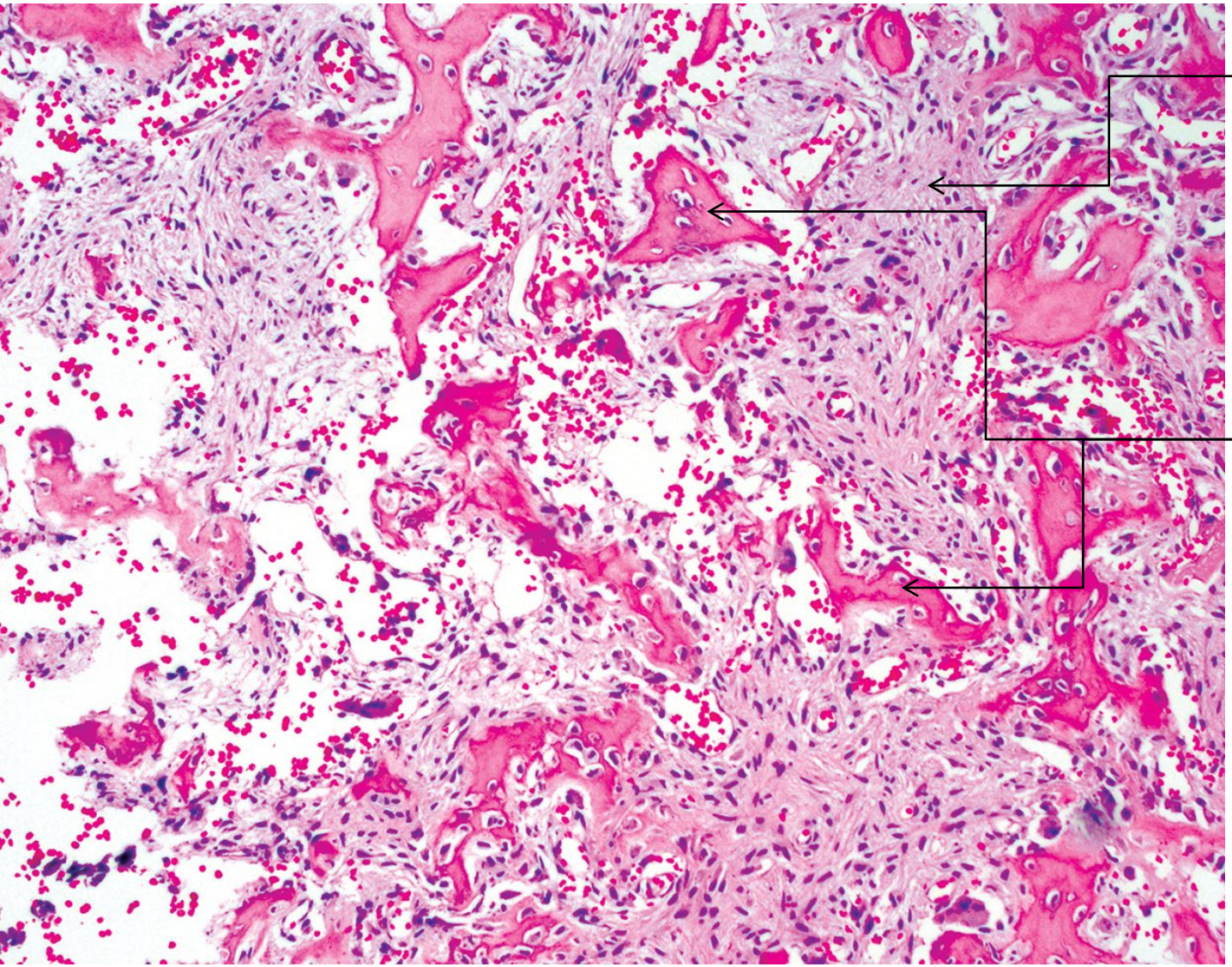

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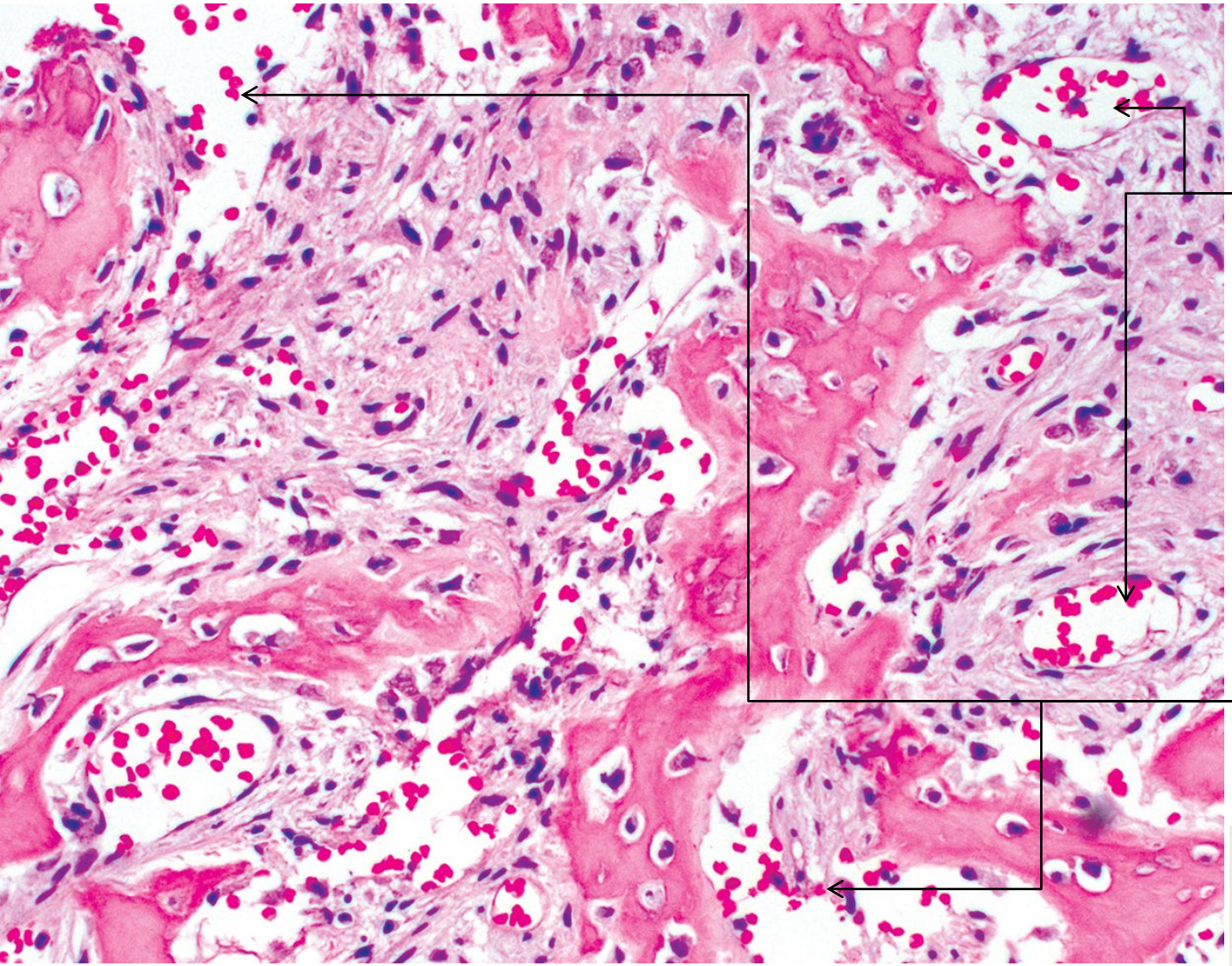

Prognosis and prediction

Once a diagnosis of periapical and focal COD has been established, patients require no treatment and can be monitored during routine dental appointments. Individuals with florid COD may require close clinical follow-up for complications of osteomyelitis

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Ossifying fibromas

Synonyms:
Cemento-ossifying fibroma:
central ossifying fibroma;
cementifying fibroma; periodontoma;
juvenile ossifying fibroma:
juvenile active ossifying fibroma;
juvenile aggressive ossifying fibroma

Definition

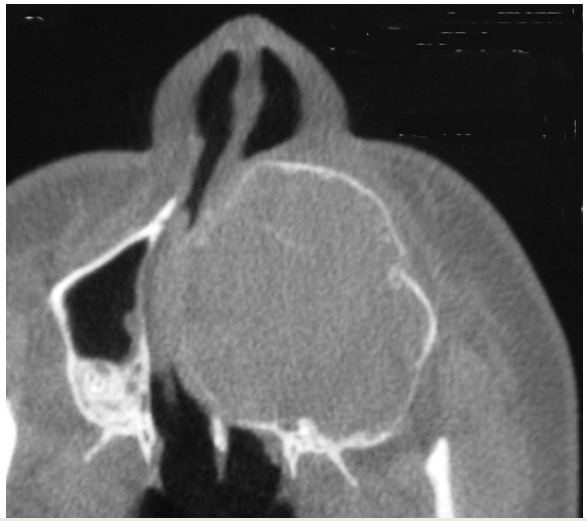
Ossifying fibromas are benign fibro-osseous neoplasms affecting the jaws and the craniofacial skeleton

Localization

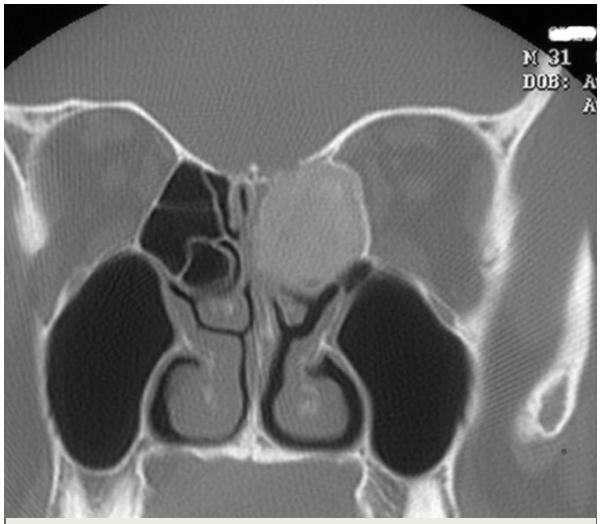
COF occurs exclusively in the tooth-bearing areas of the mandible and maxilla. The mandible is far more commonly involved than the maxilla.

Subtype(s)

1. **Ossifying fibroma of odontogenic origin** – also called cemento-ossifying fibroma (COF)
2. **Juvenile trabecular ossifying fibroma** (JTOF)
3. **Juvenile psammomatoid ossifying fibroma** (JPOF)



Juvenile trabecular ossifying fibroma
CT scan illustrating a circumscribed, expansive lesion of the maxilla; cortical thinning is observed.



Juvenile psammomatoid ossifying fibroma
CT shows an expansive, well defined but incompletely corticated lesion with ground glass appearance at the ethmoid area

Cemento-ossifying fibroma
Radiography showing a well-defined, expansive radiolucent lesion with radiodense areas present at the mandibular molar area.

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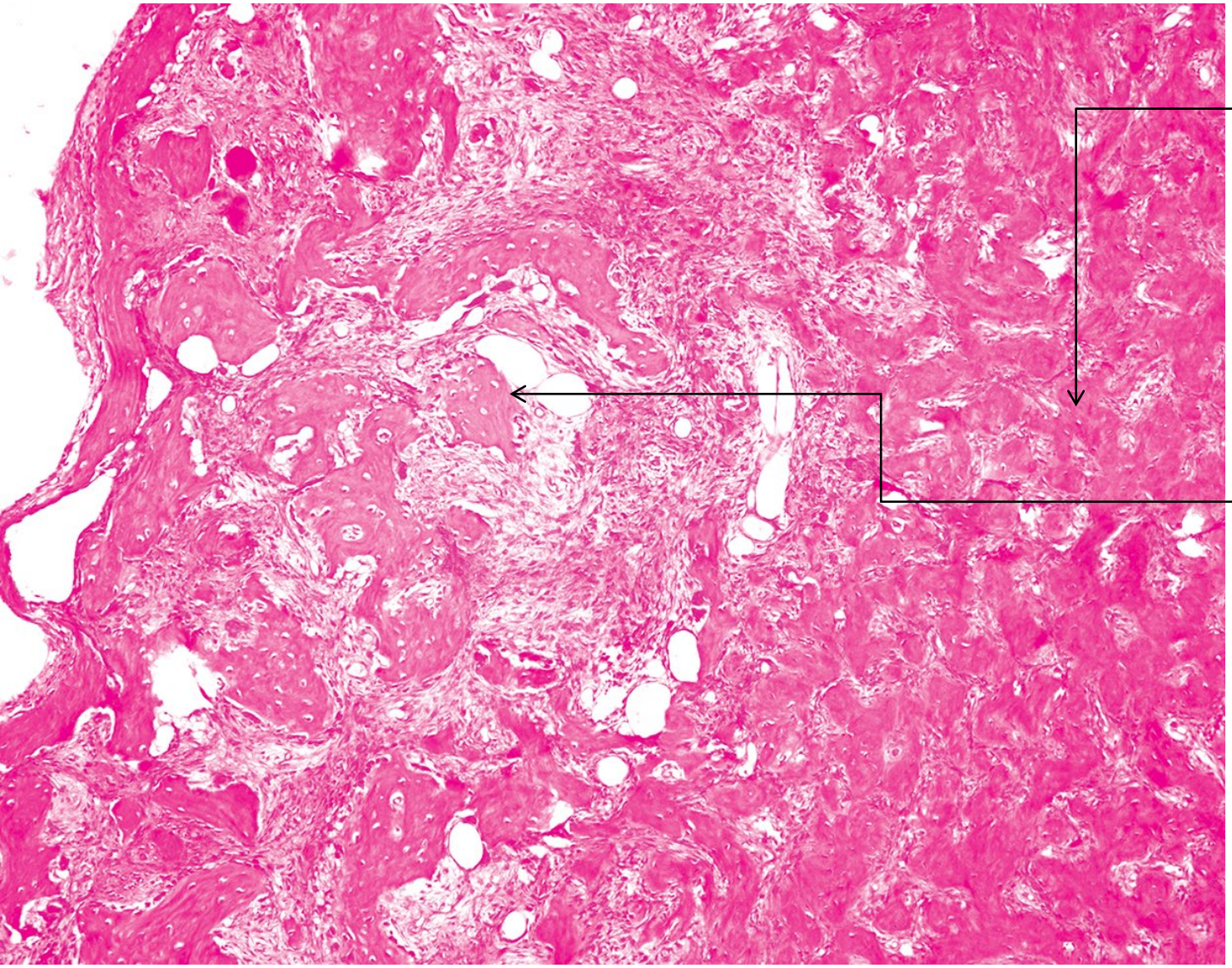
COF Histopathology



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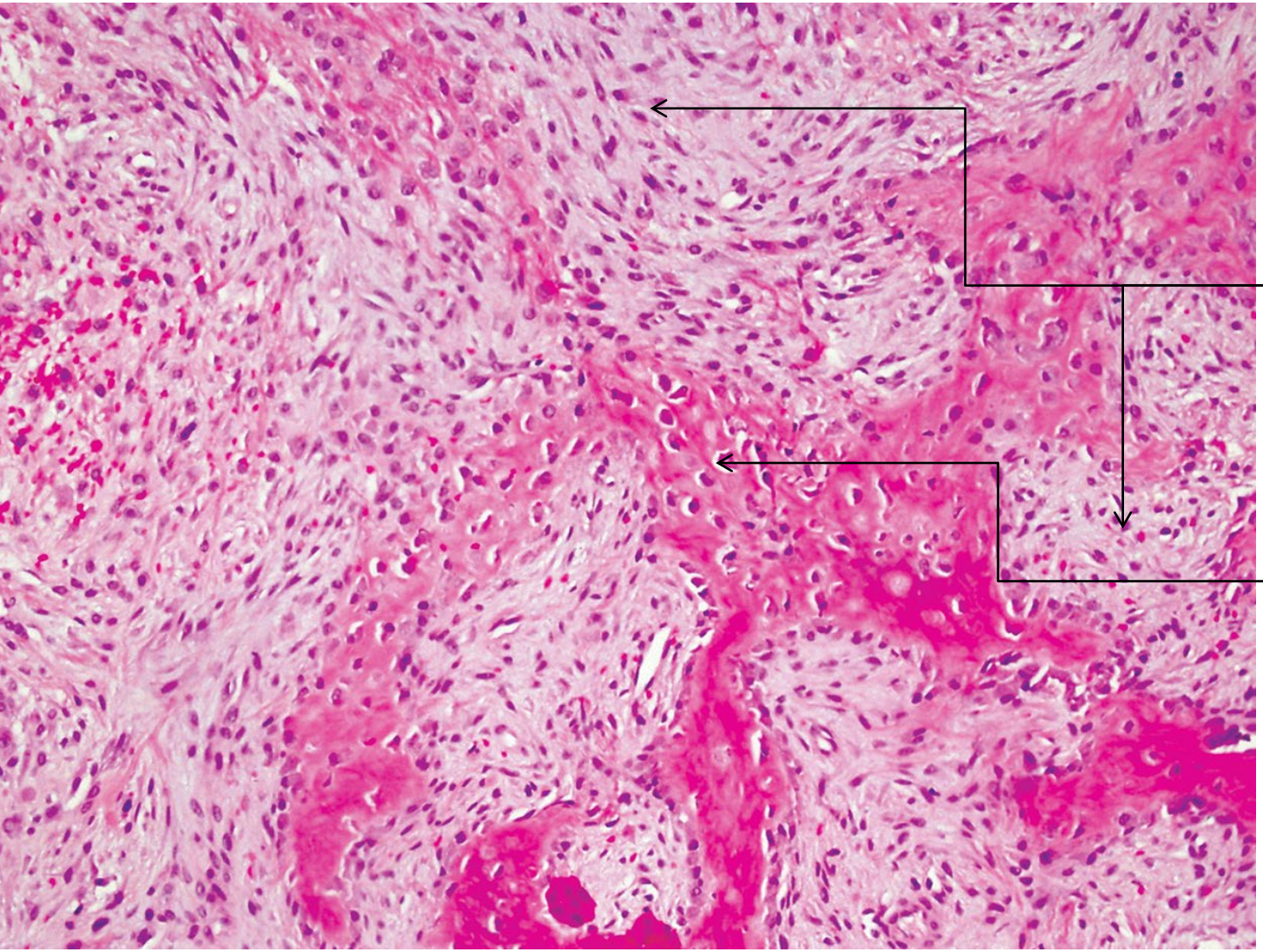
JTOF Histopathology



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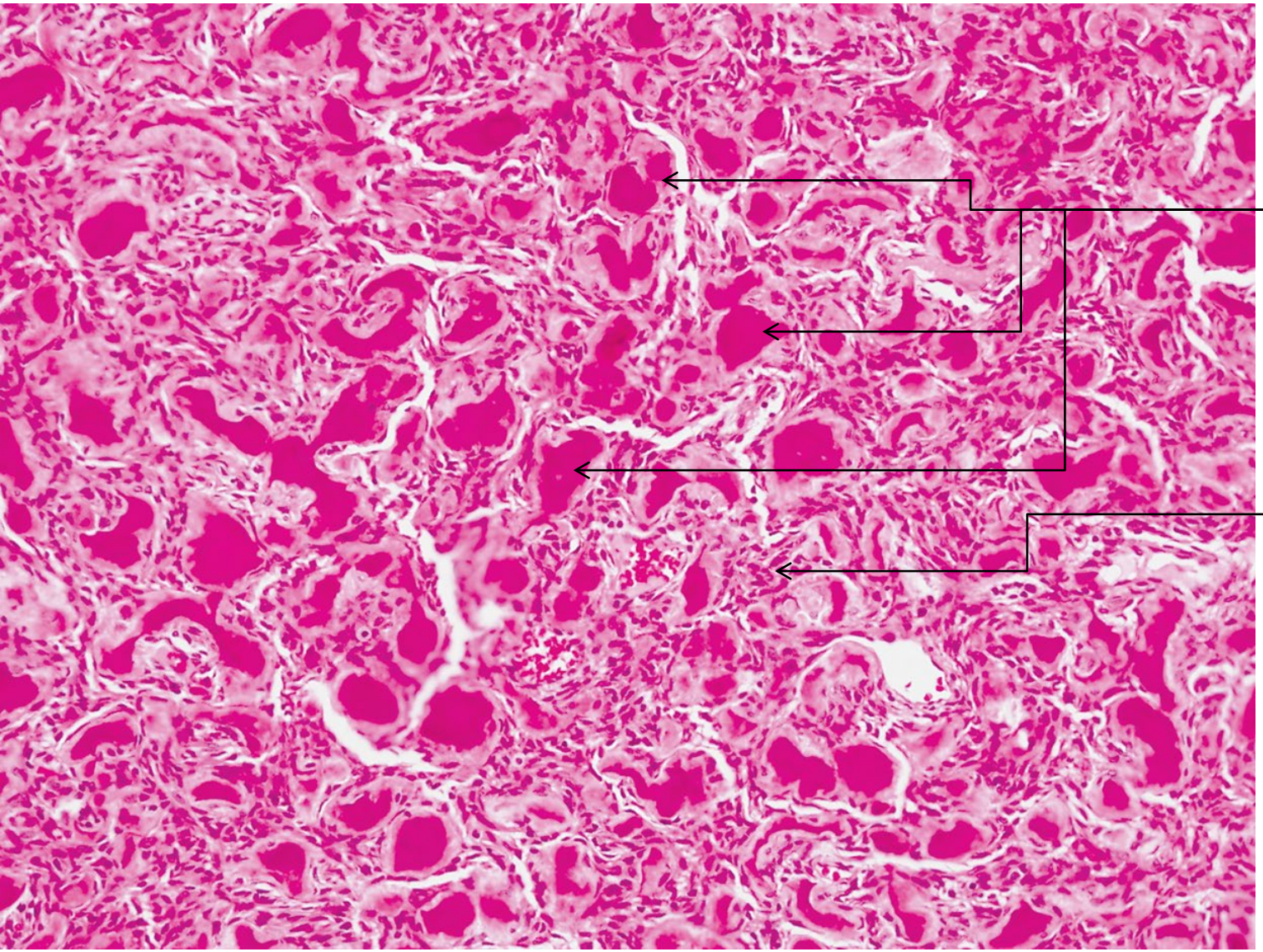
JPOF Histopathology



Prognosis and prediction
COF is a slow-growing benign neoplasm. It can be surgically excised conservatively, with no recurrence in most cases.

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Osteoporosis

Reduction in bone mass due to increased bone porosity, which predisposes bones to fracture

- Usually refers to postmenopausal or senile loss of bone severe enough to cause fractures
- Affects entire skeleton due to metabolic bone disease, but may be localized due to limb disuse
- Usually due to increased bone resorption, with normal levels of bone formation

Primary causes: due to postmenopausal condition, older age (15 million cases in US) or idiopathic

Secondary causes (due to identifiable conditions): endocrine (hyperparathyroidism, thyroid disorders, hypogonadism, pituitary tumors, type I diabetes, Addison's disease), neoplasms (myeloma, carcinomatosis), gastrointestinal disturbances (malnutrition, deficiency of vitamins C or D), drugs (corticosteroids, chemotherapy), osteogenesis imperfecta, immobilization, homocystinuria, anemia

Diagnosis

Radiographic measurement of bone density, iliac crest biopsy

Microscopic (histologic) description

1. Thin trabeculae disconnected from each other
2. Increase in osteoclastic activity (may be uneven) or increased percentage of surface with resorptive pitting

Paget disease

Also called osteitis deformans

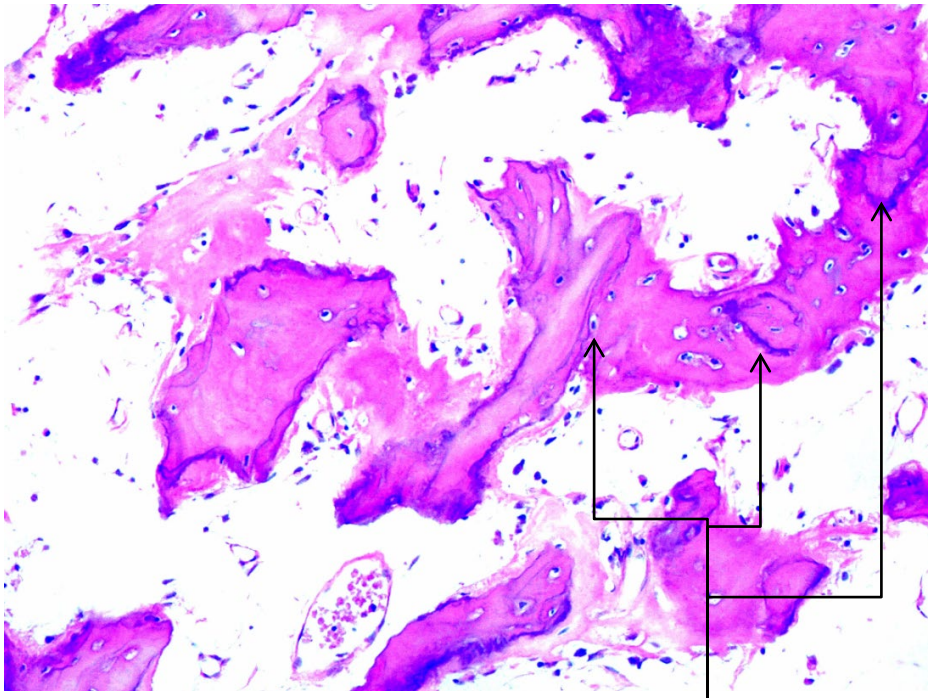
is a condition involving cellular remodeling and deformity of one or more bones. The affected bones show signs of dysregulated bone remodeling at the microscopic level, specifically excessive bone breakdown and subsequent disorganized new bone formation.

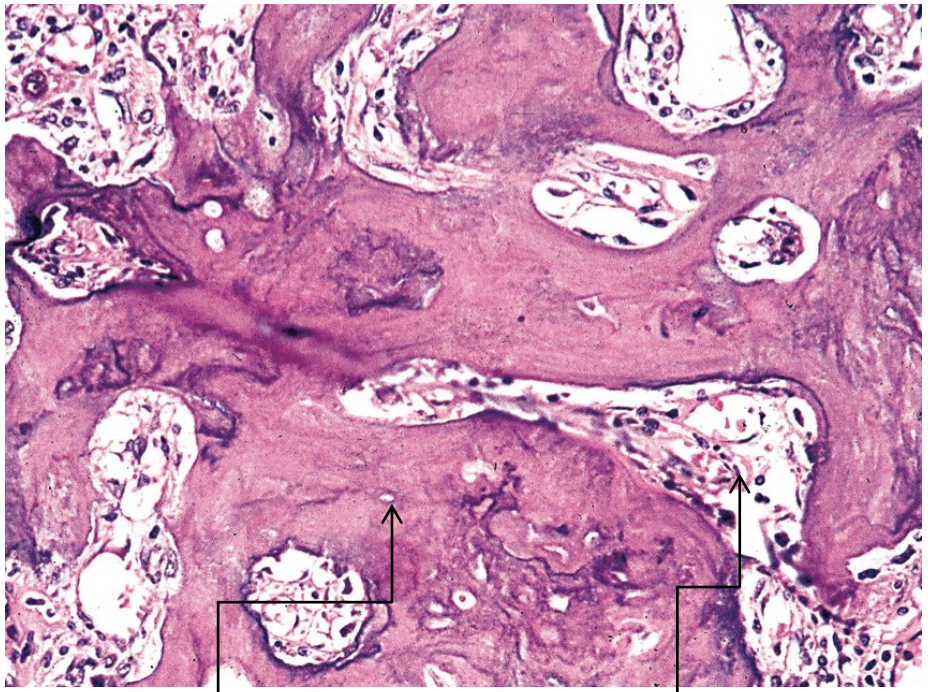
90% are over age 55, rare before age 40

Sites: 85% of presenting patients are polyostotic (pelvis, spine, skull)

Pernick N Paget disease. PathologyOutlines.com website.
https://www.pathologyoutlines.com/topic/bonepagets.html. Accessed January 15th, 2022.

PD Microscopic (histologic) description





focal mosaic pattern of lamellar bone, resembles puzzle

prominent irregular cement lines

thick trabeculae and thicker bones

fine fibrosis of marrow

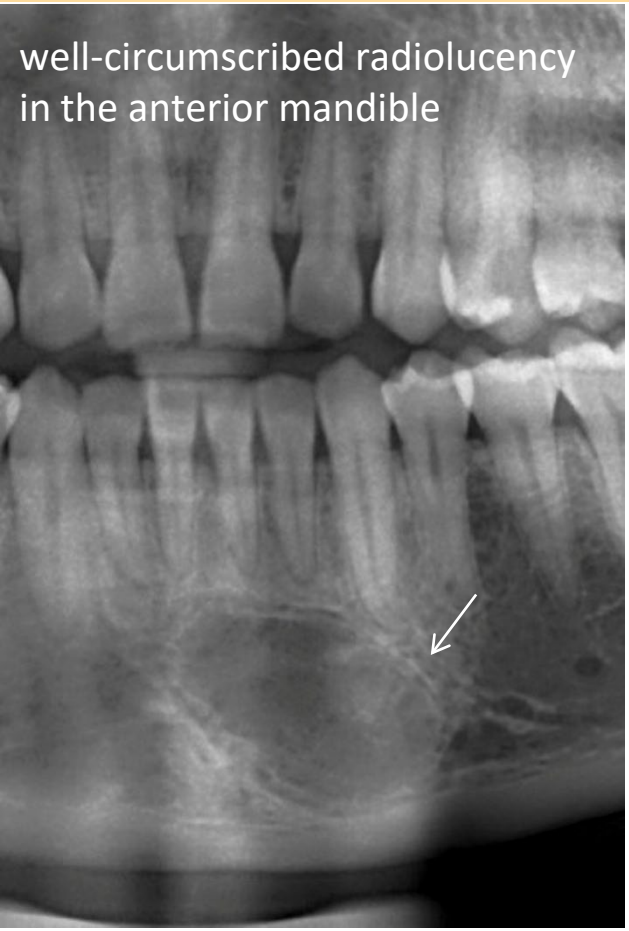
Reticulin (highlights disorganization of lamellar bone)

Pernick N Paget disease. PathologyOutlines.com website.
https://www.pathologyoutlines.com/topic/bonepagets.html. Accessed January 15th, 2022.

Central giant cell granuloma (CGCG)

Related terminology
Central giant cell lesion;
reparative giant cell granuloma (obsolete)

Definition
Central giant cell granuloma (CGCG) is a localized, benign but sometimes aggressive osteolytic lesion of the jaws characterized by osteoclast-type giant cells in a vascular stroma.



Localization
The lesions are more frequent in the anterior jaws, in particular the mandible.

Clinical features
slow-growing, asymptomatic, expansile, well-defined radiolucencies, without tooth resorption.

Epidemiology
Most cases occur in females and in patients aged < 20 years

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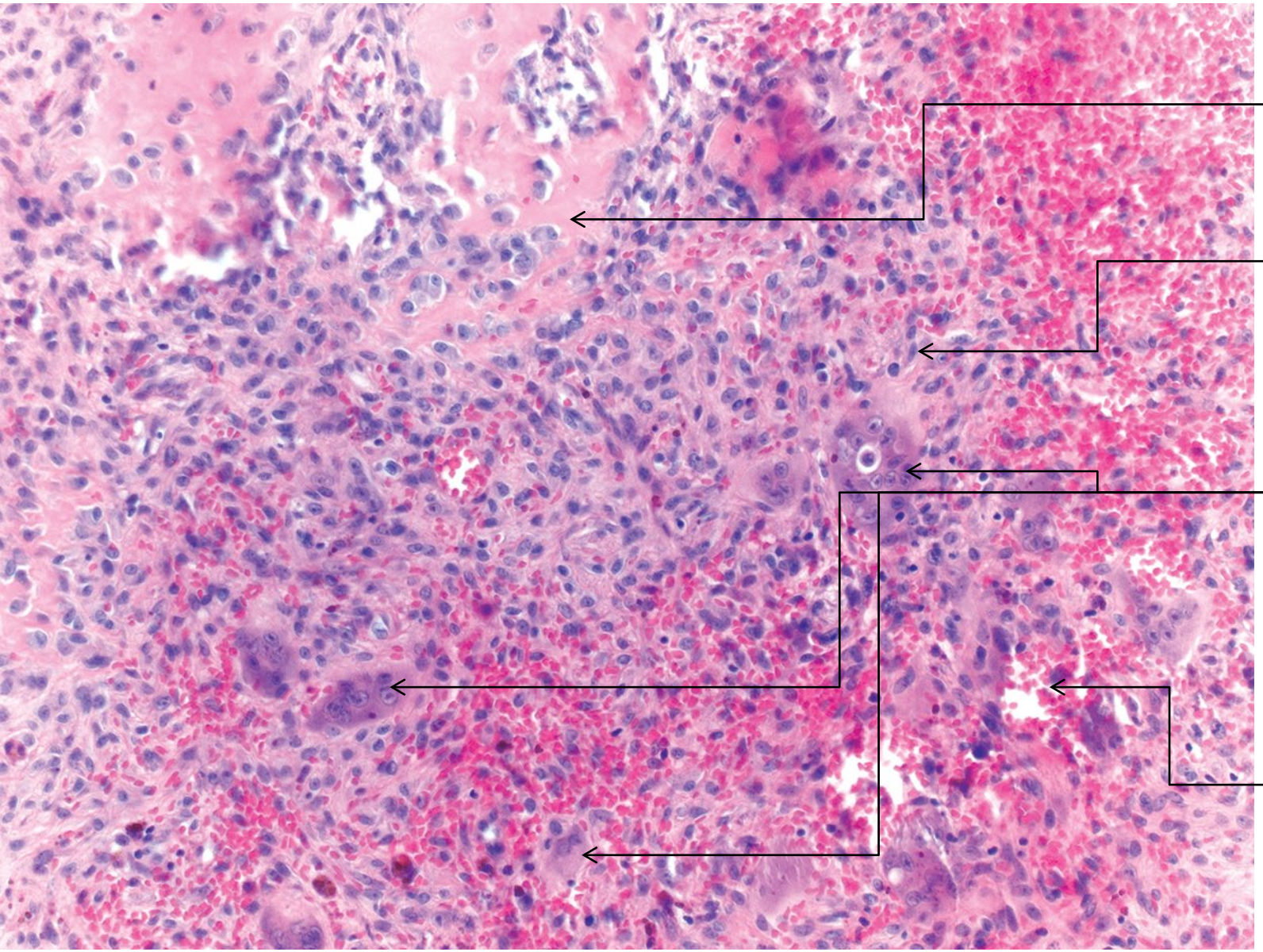
CGCG Histopathology



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Literature and links

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Thanks for attention!