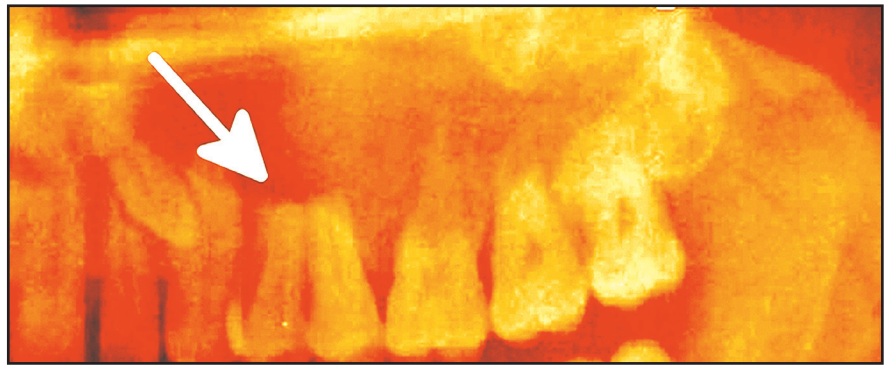


Uncommon mixed and sclerotic jaw lesions

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A variety of benign and malignant conditions can affect the jaws, the maxilla and mandible. The similar image appearance of many pathologies can make achieving a single diagnosis difficult in many situations.¹ The age of the patient, location of the lesion, relationship to the tooth and adjacent structures, margins of the lesion, internal characteristics such as the pattern of mineralization, periosteal reaction, associated soft tissue changes and relevant clinical details, all help to narrow the differential diagnosis.¹ Plain radiography retains its role as the initial mode of evaluation of these lesions.² Cross-sectional imaging, such as CT and MRI, are useful in tissue characterization, assessing extent of extra-osseous soft tissue and bony involvement in terms of cortical expansion, resorption or destruction, osteosclerosis, and calcification; thus, they help in pre-operative evaluation. MRI is superior to CT in differentiating cystic from solid lesions, and in defining the extent of soft tissue and bone marrow infiltration in aggressive jaw lesions. The classification of jaw lesions is summarized in Figure 1.

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Radiologic findings of some less common jaw lesions *Odontogenic keratocyst (OKC) or keratocystic odontogenic tumor with dentinoid formation*

In 2005, the World Health Organization (WHO) reclassified OKC to highlight the aggressive nature and high recurrence rate of this lesion. It is a benign, locally aggressive tumor of odontogenic origin, with a characteristic lining of parakeratinized stratified squamous epithelium.³ It is typically seen in young patients and commonly located in either the body or the ramus of the mandible.^{1,4} On CT, these tumors are seen as cystic structures with cortical expansion without solid components. They can be associated with an impacted tooth, mimicking a dentigerous cyst, and can sometimes be complex with multiple septations, making it difficult to differentiate OKC from ameloblastoma.

Multiple lesions are associated with Gorlin-Goltz syndrome or nevoid basal cell carcinoma syndrome. MR imaging can help differentiate OKC and ameloblastoma based on T2 relaxation time, which is more heterogeneous and short in OKC (contents appear more hypointense on T2-weighted images due to keratin), compared to ameloblastoma, where it is more homogeneous and hyperintense.^{5,6} Other MRI features, such as a thin wall with poor enhancement, are seen in OKC, compared to the thick, enhancing wall and solid component in ameloblastoma.⁵ Deposition of hard tissue like dystrophic calcification or cartilage in the cyst wall of OKC is rare, and the presence of dentinoid-like tissue (Figure 2), even more rare,⁷ can mimic other dentinoid-forming odontogenic cysts and tumors like calcifying epithelial odontogenic tumor, odontoma, ameloblastic fibro-odontoma and adenomatoid odontogenic tumor.⁷

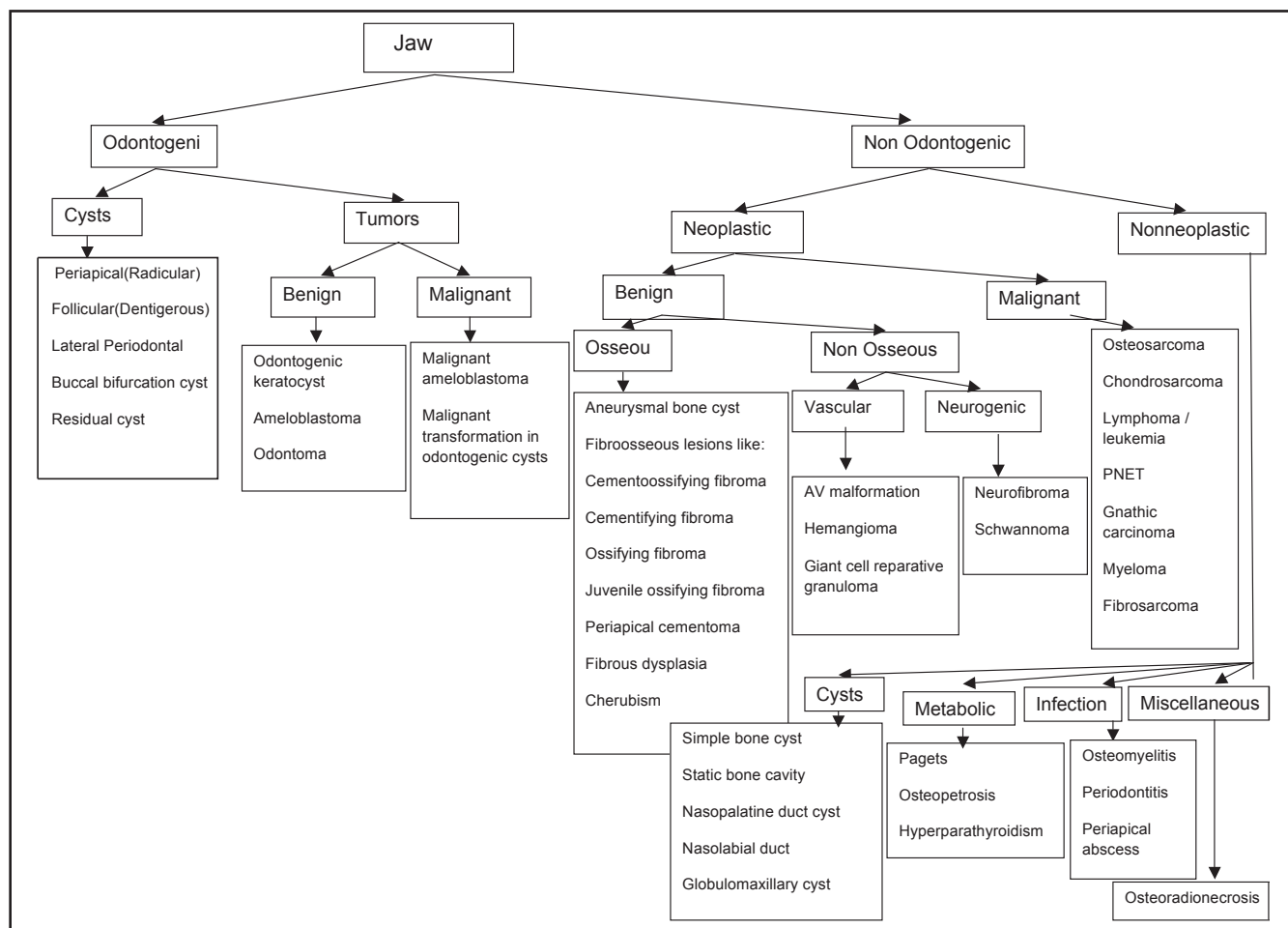


Figure 1. Flowchart of jaw lesions (Adapted and modified from Ref 1).

Odontoma

Odontomas are considered to be “tooth hamartomas” rather than true neoplasms. They appear in children as pericoronal sclerotic lesions with a thin radiolucent rim and are commonly associated with an impacted tooth. Odontomas can be simple (supernumerary teeth), compound or complex.^{1,2,8} Compound odontomas are composed of several well-formed, tooth-like structures (Figure 3A), while complex odontomas show irregular, amorphous hyperattenuating, calcified tissue surrounded by a radiolucency (Figure 3B). Odontomas can be differentiated from osteomas by the thin radiolucent line that surrounds them.⁹

Ameloblastic fibro-odontoma

Ameloblastic odontomas can be classified into ameloblastic fibro-odontoma and odonto-ameloblastoma. Odonto-

ameloblastoma is an ameloblastoma containing mineralized dental tissue. Ameloblastic fibro-odontoma is classified as a benign, mixed-odontogenic neoplasm containing neoplastic epithelium and mesenchyme. It is common in a younger (less than 20 years) age group and consists of variable amounts of components of the developing teeth, ranging from homogenous mass-like structures to loose calcified strands radiating from the center to the periphery of lytic lesions, giving the appearance of wheel spokes.^{1,2,8,10} (Figure 4).

Odontogenic myxoma

Odontogenic myxoma is a locally aggressive, benign tumor with variable radiographic appearances.^{1,2,4,11} It is commonly seen as a multiloculated, radiolucent lesion with internal osseous trabeculae and involving the tooth-bearing

area. Varying radiographic appearances have been divided into 6 types as follows: unilocular, multilocular (including honeycomb, soap bubble and tennis racquet patterns), involvement of local alveolar bone, involvement of the maxillary sinus, osteolytic destruction and a mix of osteolytic destruction and osteogenesis (Figure 5).¹² When features are not classical, the tumor can mimic ameloblastoma, Pindborg tumor, fibrous dysplasia, ossifying fibroma and even osteosarcoma when the tumor perforates the cortex with the formation of multiple, radiating spicules giving a sunburst appearance.¹²

Calcifying epithelial odontogenic tumor or Pindborg tumor

Pindborg tumor consists of epithelial tissue within a fibrous stroma. It is also associated with an unerupted or impacted tooth and is commonly seen

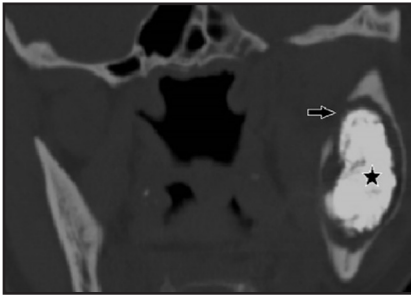


FIGURE 2. Coronal CT images showing odontogenic keratocyst with dentinoid formation in a 30-year-old woman. Cyst in mandibular ramus associated with extensive calcifying tissue or dentin formation (stars). Note the areas of cortical thinning and disruption (black arrow). Deposition of hard tissue like dystrophic calcification or cartilage or dentin like tissue in OKC can occur rarely.

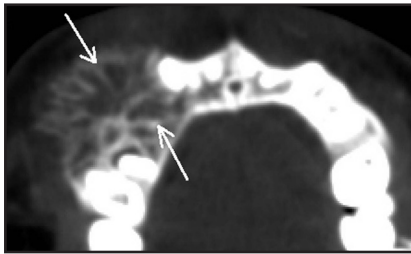


FIGURE 5. Axial CT in bone window show odontogenic myxoma of maxilla in a 15-year-old boy, with an expansile lesion having straight and curved bony septae (arrows) with characteristic "tennis racket-like" appearance

around the mandibular premolar or molar teeth as a radiolucent lesion with variable calcification (Figure 6).^{1,2,4,11,13}

Adenomatoid odontogenic tumor

This is an uncommon, slow-growing, hamartomatous and benign epithelial lesion of odontogenic origin that is frequently misdiagnosed on radiographs as other odontogenic cysts and tumors. Unlike many other cystic lesions of the jaws, these lesions occur more commonly in the maxilla than in the mandible. On radiographs they appear as well-defined expansile lytic lesions with variable internal calcifications.

Like dentigerous cysts, they are associated with impacted teeth; however, these tumors usually extend beyond the cemento-enamel junction and include the tooth root with variable calcification around the embedded tooth (Figure 7).^{1,2,4,11,8,13}

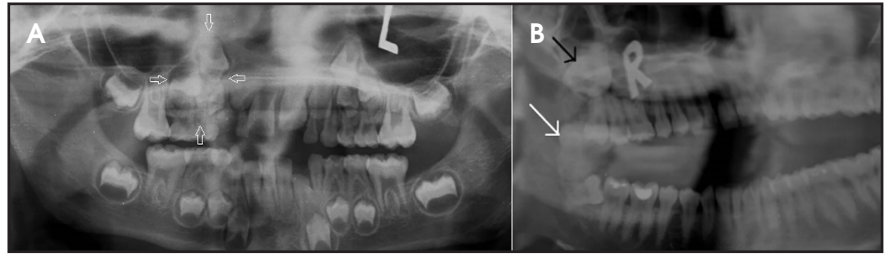


FIGURE 3. Orthopantomogram showing a compound odontoma of maxilla (A) in an 8-year-old boy and complex odontoma of maxilla and mandible (B) in a 32-year-old woman. "Cluster of tooth-like structures of varying shape" surrounded by radiolucent rim (arrows in A) in the maxilla is seen in a compound odontoma and well-delineated mass of dental tissues in posterior mandible (white arrow) and maxilla (black arrow), hampering normal eruption of the third molar is seen in a complex odontoma. Odontomas are considered to be tooth hamartomas rather than true neoplasms. Compound odontomas are commonly seen in anterior maxilla and complex odontomas in molar regions as illustrated in these cases.

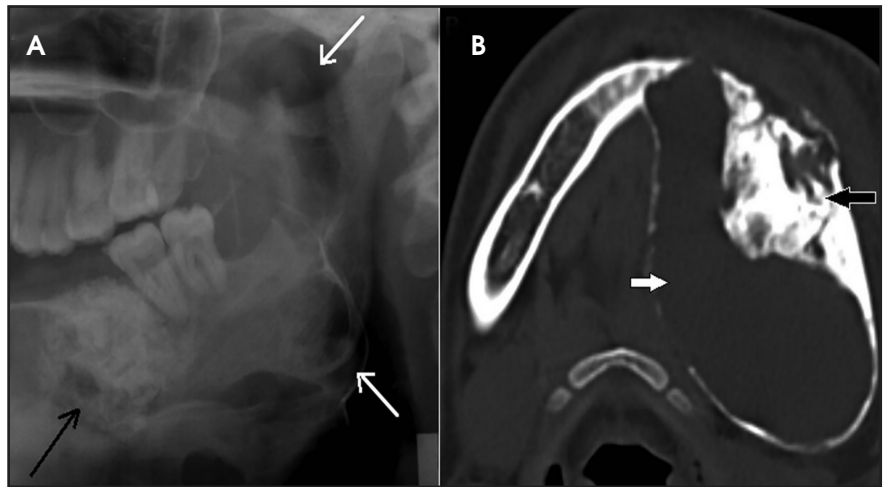


FIGURE 4. Orthopantomogram (A), axial (B) CT show ameloblastic fibro-odontoma of mandible in a 16-year-old girl. A well-circumscribed expansile radiolucent lesion (white arrows) is seen in the body and ramus of mandible with eccentric amorphous calcified mass (black arrow). CT confirms the dentin like mass suggesting complex odontoma (black arrow in B) within the large cystic lesion (white arrow in B). Larger cystic component helps to differentiate ameloblastic fibro-odontoma from complex odontoma.

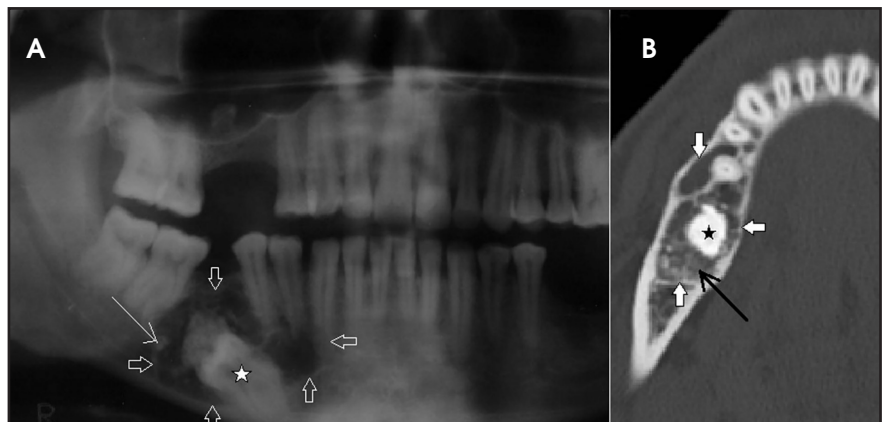


FIGURE 6. Orthopantomogram (A) and axial CT section (B) show calcifying epithelial odontogenic (Pindborg) tumor of mandible in a 49-year-old male. A well-defined expansile mixed density lesion (open arrows in A, white arrows in B) is seen involving the mandible, containing small peripheral calcifications (thin arrow in A, black arrow in B) and inferiorly displaced unerupted first molar (star in A, B).

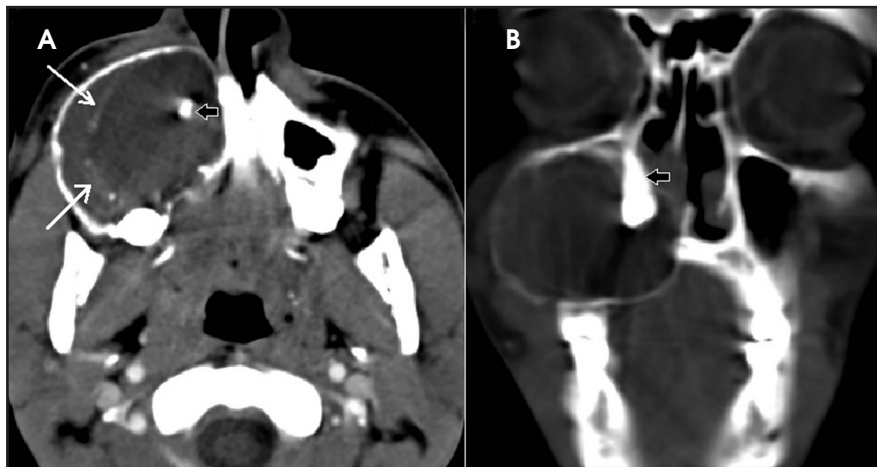


FIGURE 7. Axial (A) and coronal (B) CT sections showing adenomatoid odontogenic tumour of maxilla in a 23-year-old male, as a well marginated unilocular expansile lytic lesion with curvilinear calcification (white arrows) and impacted tooth (black arrows), which is completely inside the lesion. Extension of the lesion above the cement-enamel junction with the tooth completely inside the cyst helps to differentiate it from dentigerous cysts, where only the crown of the tooth is within the cyst.

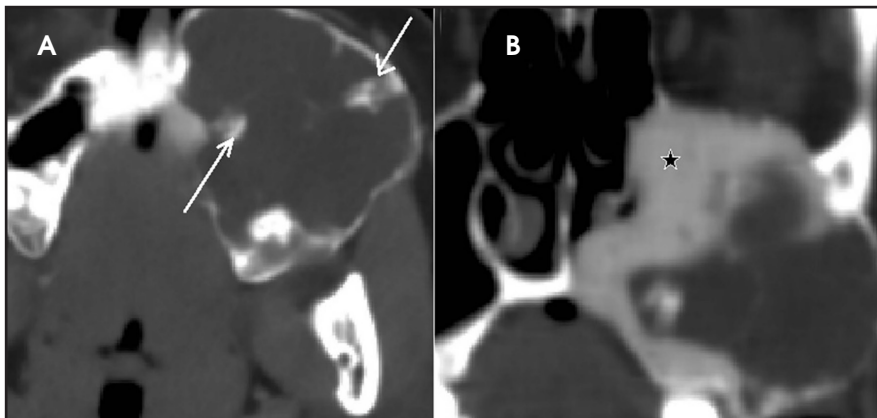


FIGURE 8. Axial (A) and coronal (B) CT sections show cementifying fibroma of maxilla in a 21-year-old-female, with an expansile lobulated well circumscribed mixed density mass with homogenous dense eccentric, heavily ossified area (stars), few scattered linear and nodular calcific foci (arrows).

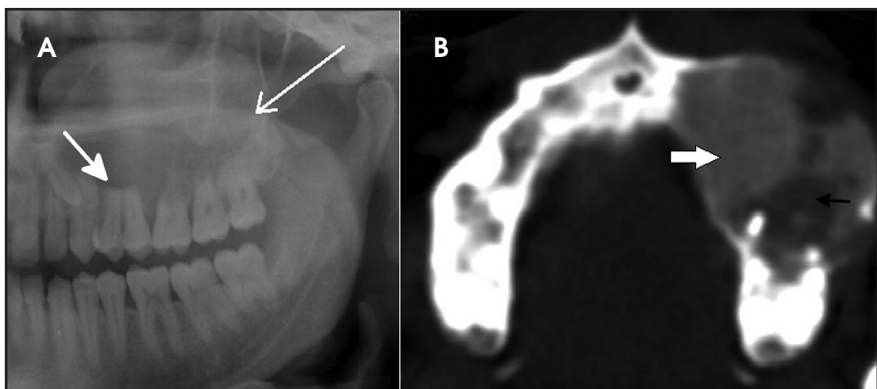


FIGURE 9. Orthopantomogram (A) and axial (B) CT images show ossifying fibroma of maxillary alveolus in a 31-year-old male. A mixed density expansile lesion (arrow in A) is seen involving the maxillary alveolus with erosion of roots of adjacent teeth (thick white arrow), internal ground glass matrix (arrow in B), non-mineralized areas (black arrow), with ballooning of cortices without frank disruption.

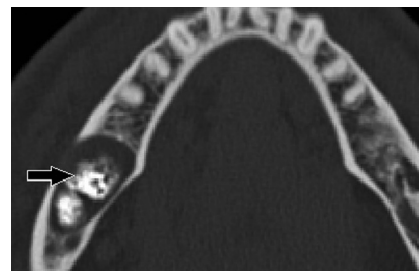


FIGURE 10. Axial CT image at the level of tooth roots show periapical cemento-osseous dysplasia of right mandibular molars in a 54-year-old woman. Periapical well-marginated calcified masses (arrow) are seen, surrounded by a halo of lytic area.

**Benign fibro-osseous lesions:
Ossifying fibroma, cementifying
fibroma and cemento-ossifying
fibroma**

Benign fibro-osseous lesions of the jaw include a spectrum of developmental, reactive or neoplastic lesions characterized by replacement of normal bone with fibrous tissue that, over time, is infiltrated by osteoid or cementoid tissue. These lesions are believed to arise from cells of the periodontal membrane. They are broadly classified as fibrous dysplasia and ossifying fibroma. Ossifying fibromas include cementifying fibromas, cemento-ossifying fibromas and juvenile ossifying fibromas. Psammomatoid and trabecular types of juvenile ossifying fibromas are recognized.⁹ Dysplastic conditions around the teeth, such as periapical, focal or florid cemento-osseous dysplasias, are also included in some classifications like that proposed by Charles A Waldron.¹⁴ Depending on their evolution, these lesions can vary in appearance, with a more lucent appearance seen initially due to predominant fibrous tissue and deposition of dense osteoid or cementum-like material⁹. Cementifying fibromas (Figure 8) and cemento-ossifying fibroma are very similar to ossifying fibromas, but they have a greater tendency to form cementum-like material that is more ovoid and heavily calcified. However, in ossifying fibroma, the calcifications are more spiculated, with a ground-glass appearance (Figure 9). Ossifying fibromas show a narrow transition zone from surrounding normal bone, thus



FIGURE 11. Axial (A) and coronal (B) CT images of a 27-year-old male with left molar tooth pain for 4 months shows cementoblastoma involving the left maxilla. A left periapical lucent lesion with central calcification (white arrow) is seen, which fuses with the tooth root (black arrow).

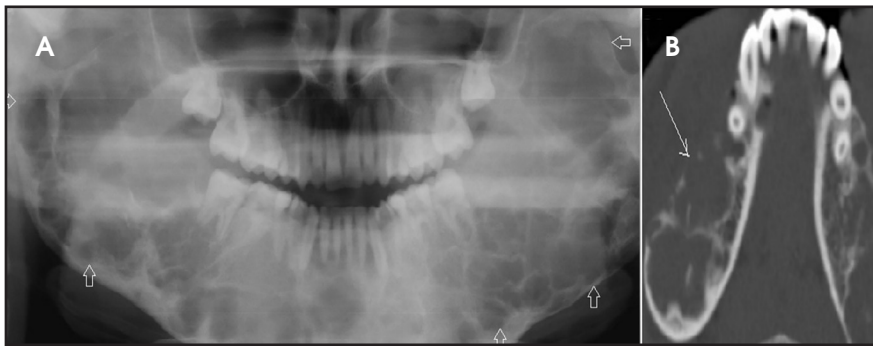


FIGURE 12. Orthopantomogram (A) and axial (B) CT sections show cherubism (familial multilocular cystic disease of the jaws) in a 21-year-old male. Diffuse multilocular expansion of mandible (open arrows in A) is noted with cortical thinning and disruption (long thin arrows in B).

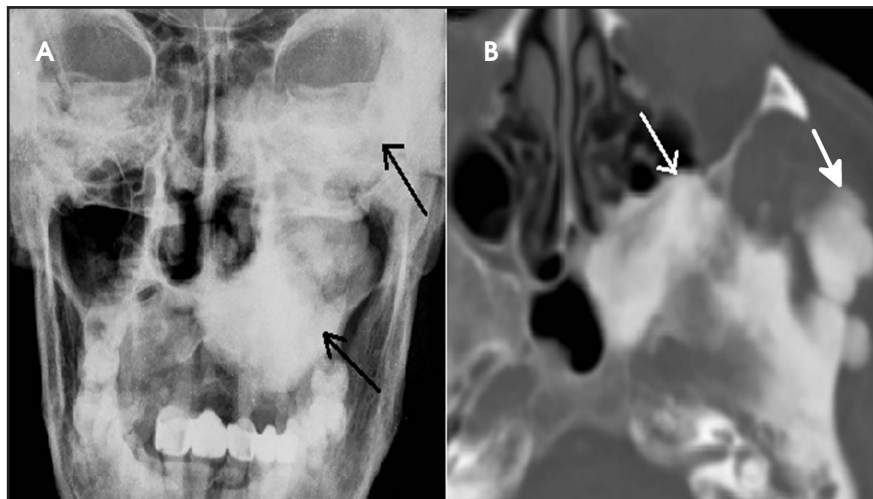


FIGURE 13. Skull radiograph (A) and axial CT section (B) show osteomas of cranio-facial bones in a 43-year-old man. Multiple lesions of cortical density (black arrows in A, white arrows in B) in cranio-facial bones suggesting ivory osteomas in sphenoid and maxillary bones.

differentiating them from fibrous dysplasias. Another differentiating feature is displacement of teeth in ossifying fibroma, unlike fibrous dysplasias.⁹

Cemento-osseous dysplasia/periapical osseous dysplasia

The 2005 WHO classification divides these bone-related lesions into periapical, focal, florid cemento-osseous dysplasias and familial gigantiform cementomas.³ These appear as single or multifocal, well-defined, cement-like sclerotic lesions in a periapical location with surrounding lucent halo (Figure 10). Middle-aged women are most commonly affected. The closest differential is cementoblastoma (Figure 11), which occurs in children and fuses to the tooth root, unlike cementososseous dysplasia. Like ossifying fibromas, these lesions also show temporal evolution in appearance from early lucent lesions to progressive increase in density. Early lucent lesions can mimic periapical inflammatory lesions; however, the latter is unlikely in a normal tooth with no caries or previous treatment.⁹

Fibrous dysplasia - cherubism

Fibrous dysplasia is a disturbance of bone metabolism where the fibrous connective tissue containing abnormal bone replaces normal bone. It can exist in monostotic and polyostotic forms². In its typical form it has ground glass matrix however, atypical forms can mimic other fibro-osseous lesions like ossifying fibroma in the craniofacial region. A rare variant of this entity called Cherubism is a familial form affecting children and adolescents and typically involving only the maxilla and mandible^{2, 15}. On imaging it is seen as expansile remodeling of the maxilla and mandible with thinning of the cortices, multilocular radiolucencies and a coarse trabecular pattern (Figure 12).

Benign bone and cartilage-forming tumors — multiple osteomas in Gardner's syndrome

Osteomas are slow-growing, benign, bone-forming tumors commonly

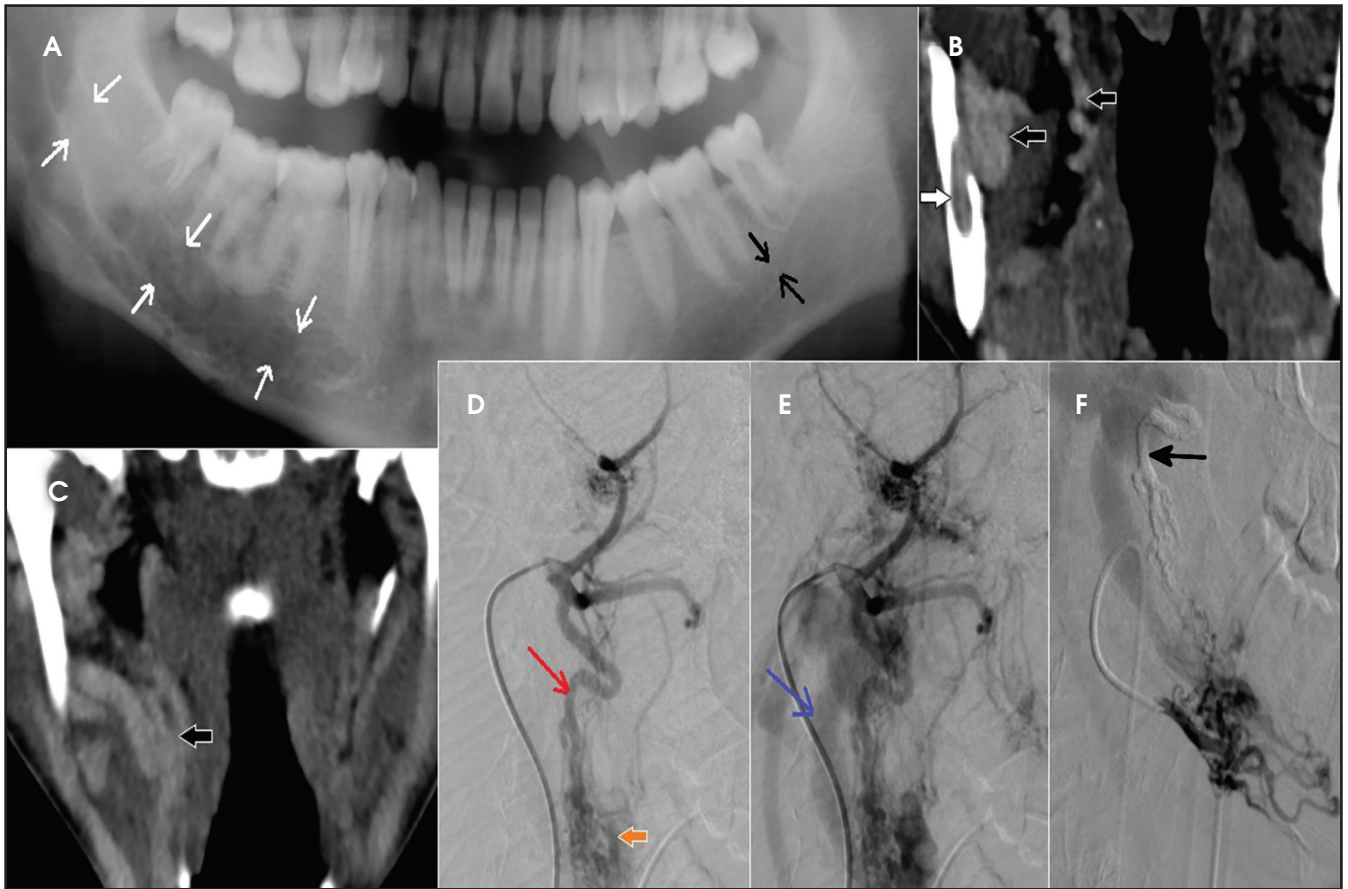


FIGURE 14. Orthopantomogram (A), coronal contrast-enhanced CT (B,C), digital subtraction angiogram (DSA; D,E,F) show intraosseous arteriovenous malformation of mandible in a 17-year-old male. Ill-defined lytic areas in the mandible are seen on the right side, with enlarged ipsilateral mandibular canal (white arrows) as compared to contralateral side (black arrows in A). Multiple enlarged vessels (black block arrows) seen in and around the mandible with intraosseous extension through the widened mandibular canal (white arrow in B). DSA demonstrates the nidus (orange arrow), arterial feeders mainly from branches of internal maxillary artery (red arrow in D) and facial artery and draining vein into the external jugular vein (blue arrow in E). The arterial feeders were embolized (glue cast of arterial feeders in F, black arrow).

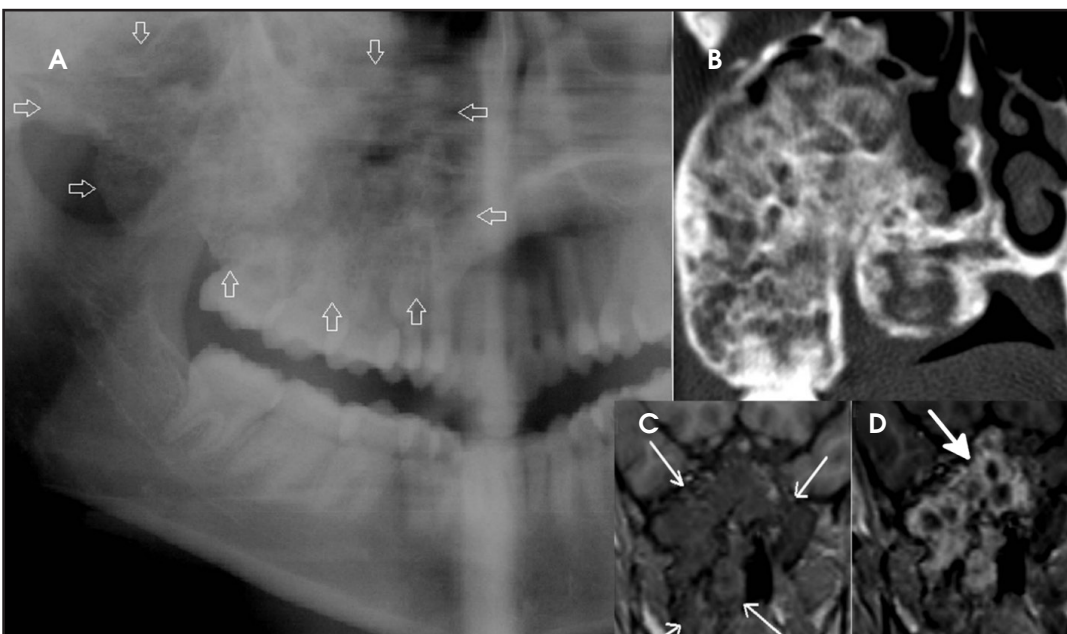


FIGURE 15. Orthopantomogram (A), axial (B) CT and T1W pre- (C) and post-gadolinium (D) MRI with intraosseous haemangioma of maxilla and adjacent bones in a 30-year-old woman. There is marked expansion of right maxillary sinus, sphenoid and petrygoid bones with rarefaction and radiating trabeculations (open arrows in A), giving a “honeycomb-like” appearance. On MRI, the lesion is iso to hypointense (white arrows in C) with fine radiating lines of high signal intensity, heterogenous contrast enhancement (white arrow in D).

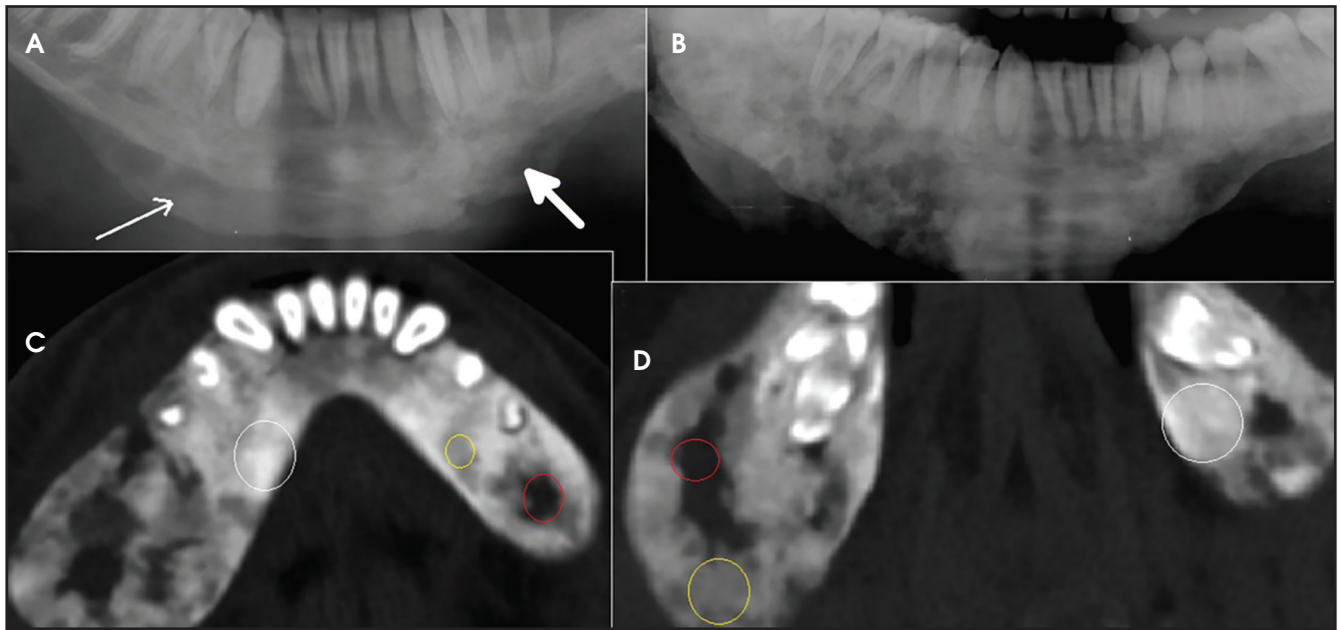


FIGURE 16. Initial Orthopantomogram (A), and one two years later (B), and CT (C, D) show chronic osteomyelitis of mandible in a 14-year-old boy. Ill-defined mixed lytic and sclerotic areas (thick white arrow in A) are seen in central mandible with subperiosteal new bone formation (thin white arrow); which has progressed considerably in B, 2 years later, with bone expansion, remodeling and diffuse lytic and sclerotic foci. CT images show all the three patterns of CT findings in osteomyelitis with bone-defect (red circle), frosted-glass (yellow circle) and compact-bone patterns (thin white circles).



FIGURE 17. Orthopantomogram shows Caffey disease (infantile cortical hyperostosis) in a 5-month-old baby. Note diffuse lamellated subperiosteal new bone formation giving "double contour" appearance of the cortex (arrows).

occurring in the craniofacial bones and characterized by the proliferation of compact and/or cancellous bone. In the jaws, they occur along the lingual side of the ramus or the inferior mandibular border below the molars. On imaging, an osteoma appears as a well-defined, non-tooth related, dense and radiopaque mass with no surrounding radiolucent line, with or without bone expansion. In the absence of expansion, differentiation from idiopathic osteosclerosis may be difficult.⁹ Multiple craniofacial os-

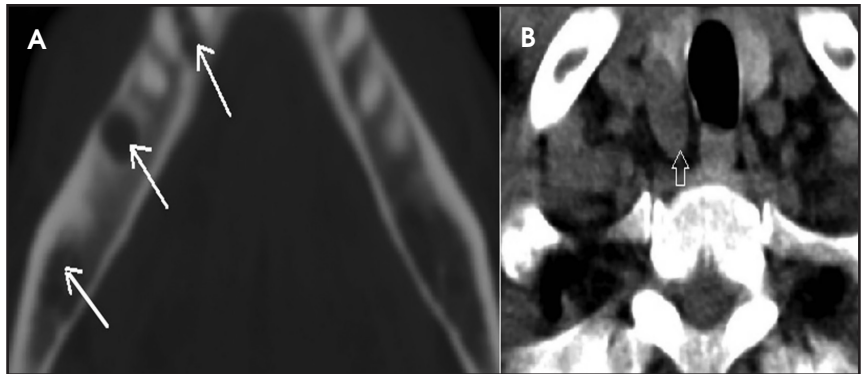


FIGURE 18. CT sections through the mandible (A) and lower neck (B) show parathyroid adenoma and brown tumors of jaw in a 67-year-old male. Multiple well-defined lytic lesions (white arrows) are seen in the mandible. Parathyroid adenoma is seen as an oval isodense lesion (open arrow in B) posterior to right lobe of thyroid gland.

teomas, along with cutaneous and soft-tissue tumors, can suggest a diagnosis of Gardner's syndrome² (Figure 13).

Vascular lesions *Arteriovenous malformation/ hemangioma*

Intraosseous AVM is rare and recognition is important because extraction of an associated tooth may lead to fatal hemorrhage.^{2,4} In the jaw, they usually occur within the ramus and posterior mandibular body.¹ Radiographically,

they appear as multilocular cystic lesions, and angiography is usually necessary to demonstrate the vascular nature of the lesion (Figure 14).² Angiography clearly demonstrates the feeding and draining vessels and also helps in treatment planning.

The intraosseous or central hemangioma is frequently found in the vertebrae and skull and rarely seen in the jaws.¹⁶ These lesions are also great mimickers, with different radiographic appearances. Most commonly, lesions

show a multilocular radiolucency with small (honeycomb appearance) or large (soap-bubble appearance) loculations. Phleboliths or vascular calcifications may also appear as rounded or oval radiopacities.¹⁶ They can mimic ameloblastoma, keratocyst and fibro-osseous lesions such as fibrous dysplasia when they have honeycomb or soap-bubble appearances¹⁷ (Figure 15). CT and MRI allow clear identification of the lesion's vascular nature and also can delineate the extent of involvement. Honeycombed appearance and periosteal reaction are described as rare CT findings.¹⁷

Infective and inflammatory conditions

Osteomyelitis

Periodontitis, periapical abscess and osteomyelitis are important entities in this group of infective/inflammatory conditions. Apical periodontitis includes a spectrum of periapical cysts, granulomas and abscesses. They are mostly related to periodontal diseases,² such as dental caries and irreversible pulpitis. Distinguishing apical cysts and granulomas on imaging is difficult. Thickening of the periodontal ligament space can be the earliest sign. If apical abscess forms, the enhancing rim can be seen on images. Progression of the infection to osteomyelitis or inflammation of the bone and bone marrow is rare due to the early administration of antibiotics. Acute osteomyelitis may not demonstrate any changes on plain radiography, while chronic osteomyelitis may appear as focal or diffuse lytic or sclerotic areas with periosteal reaction.¹⁸ All three patterns of CT findings in bone in chronic osteomyelitis are illustrated in Figure 16.

Systemic disease affecting the jaws

Systemic diseases that can commonly affect the jaws include Paget's disease, osteopetrosis, Caffey's disease, eosinophilic granuloma, brown tumor of hyperparathyroidism and, rarely, amyloid deposits. Associated changes in other bones, clinical features and biochemical

parameters will help complete the final diagnosis in these systemic conditions.²

Caffey disease

Caffey disease, or Infantile Cortical Hyperostosis, is a rare, self-limiting disorder of unknown etiology and affecting infants, usually younger than 6 months of age. It is characterized by inflammation of the periosteum and the overlying soft tissue along with systemic changes of irritability and fever. The mandible, ribs, clavicles, scapula and long bones can be involved. Radiographs show classical cortical hyperostosis beneath regions of soft-tissue swelling¹⁹ (Figure 17). Osteomyelitis, chronic hypervitaminosis A, scurvy and child abuse are common differentials.

Hyperparathyroidism

Brown tumors are non-neoplastic lesions found in hyperparathyroidism and can involve the maxilla or mandible.¹¹ They arise from a change in bone metabolism and present clinically and radiologically as expansile masses. One or more lesions may be seen in a bone. They have variable well- or ill-defined margins (Figure 18). Associated changes, such as bone density, granular texture of bony trabeculae, and loss of the lamina dura in other teeth, can point towards hyperparathyroidism. They are very similar histologically and radiologically to other giant-cell lesions and differential diagnosis includes giant-cell reparative granuloma, true giant-cell tumor and aneurysmal bone cyst. The familial type of hyperparathyroidism with associated lesions in the jaw and kidney is called hyperparathyroidism—jaw tumor (HPT-JT) syndrome. The jaw lesions are usually fibro-osseous and the renal involvement may be either as cysts or as tumors, such as Wilm's tumor.²⁰

Conclusion

Jaw lesions are fairly common in radiology practice. Familiarizing oneself with different conditions and their appearance on imaging can help radiologists to reach a reasonable differential diagnosis.

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