



COMMON DENTOFACIAL MANIFESTATIONS OF SYSTEMIC DISEASES: RADIOLOGIST'S PERSPECTIVE

Galal Omami

Oral Diagnosis and Polyclinics, The University of Hong Kong Faculty of Dentistry, Hong Kong.

ARTICLE INFORMATION:

Article History:

Received: 22 December, 2013

Accepted in revised form: 25 May, 2014

Published: 1 August, 2015

Corresponding author:

Galal Omami

E-mail: jellodent@yahoo.com

Keyword:

Dentistry, dentofacial manifestations, systemic diseases, radiology

ABSTRACT:

This pictorial article provides a guide for recognizing the dentofacial manifestations of systemic diseases from a radiographic perspective. For comprehensiveness, imaging findings of a number of dentofacial manifestations of systemic diseases are briefly reviewed and teaching points are delivered in a fashion of mini case studies.

الملخص العربي

المظاهر الفموية و الوجهية للأمراض الجهازية: من منظور إختصاصي الأشعة

جلال العمامي

عيادات تشخيص أمراض الفم ، كلية طب الاسنان، جامعة هونغ كونج ، هونغ كونج

وتنص هذه المادة التصويرية دليل للتعرف على المظاهر الفموية و الوجهية للأمراض الجهازية من منظور الشعاعي. يقدم المقال مراجعة شاملة لنتائج التصوير الشعاعي لعدد من هذا المظاهر بشكل تدريسي مبسط.

معلومات المقال:

تاريخ المقال:

أستلم في: 22 ديسمبر، 2013

قبل بعد المراجعة في: 26 مايو، 2014

نشر في: 1 أغسطس، 2015

المؤلف المسؤول:

جلال العمامي

البريد الإلكتروني: jellodent@yahoo.com

الكلمات المفتاحية:

طب الاسنان ، المظاهر الفموية و الوجهية، الأمراض الجهازية ، الاشعة

INTRODUCTION:

Bone and teeth changes occur with certain metabolic, endocrine, hematological, and other systemic diseases and may be revealed in radiographs. Skeleton contains more than 99% of the total body calcium, so it must be borne in mind that substantial alteration in the amount of calcific material must take place for any radiographic change to appear¹. However, failure to demonstrate disease is not proof of its absence. The effects of systemic diseases of bone are brought about by changes in the number and activity of osteoclasts, osteoblasts, and osteocytes. So the bone structure may be altered in the direction and numbers of trabeculae and/or in the degree of calcification¹. This pictorial article reviews the radiographic findings of a number of systemic diseases manifested in the dento-maxillofacial complex.

Sickle cell anemia and thalassemia:

Sickle cell anemia (SCA) is an inherited form of hemolytic anemia that occurs mainly in black individuals. In SCA, glutamic acid, normally presents on the sixth position of beta chain of hemoglobin, is replaced by valine. This abnormal hemoglobin, under low oxygen tension, results in sickling of the red blood cells (RBCs). These sickled RBCs have reduced oxygen-carrying capacity and rupture more rapidly than normal cells. Thalassemia, also called Mediterranean and Cooley's anemia, is a hereditary chronic anemia which results from a defect in hemoglobin synthesis. Thalassemia is classified into thalassemia minor (heterozygous form) and thalassemia major (homozygous form). The Radiographic findings in SCA and thalassemia result from the hemolysis of RBCs and compensatory hyperplasia of bone marrow resulting in widening of medullary spaces (osteoporosis), fewer but coarser trabeculae, and thinned cortices (Figures 1, 2). In SCA, the remaining trabeculae appear to be lined up in a horizontal "stepladder" pattern in the interradicular processes. However, such trabeculation pattern has been seen non-SCA patients. SCA may be complicated by osteomyelitis if a focus of infection ensues in an area of pronounced hypovascularity^{2,3}. In thalassemia, marrow hyperplasia in facial bones prevents their normal development resulting in peculiar facial deformity featuring pronounced protrusion of premaxilla and malar

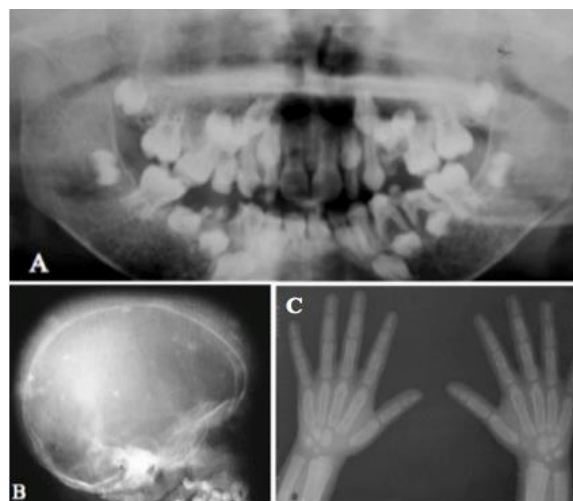


Fig.1. 6-year-old black boy with severe sickle cell anemia. A, C; panoramic and hand films show coarse trabeculation. B; lateral skull radiograph shows the hair-on-end pattern; and distinct foci of radiopacity representing healed infarcts (dystrophic medullary calcification).

bones resulting in "chipmunk" face; and absence of the paranasal sinuses, with exception of ethmoid air cells as they lack red marrow.

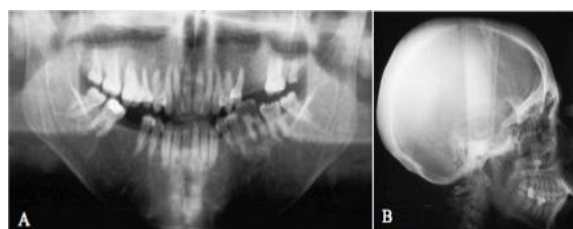


Fig. 2. Thalassemia. A; panoramic radiograph shows thickened mandibular body, reduction in trabeculation, and aplastic maxillary sinuses. B; lateral skull film of the same case shows enlargement of diploic space, thinning of inner and outer tables of calvarium, and subtle linear orientation of the trabeculae.

The roots of the teeth may be short and spikey⁴. In severe cases of SCA and thalassemia, the loss of the outer table of the calvarium results in perpendicular trabeculae radiating outward from the inner table, in response to the intracranial pressure, giving a "hair-on-end" appearance^{2, 3, 4}.

Paget's disease of bone (osteitis deformans)

Paget's disease of bone is a chronic idiopathic disease characterized by uncoordinated episodes of bone desorption and deposition. The new bone is of poor quality and may result in increased bone fragility and a tendency to fracture. The incidence increases in older individuals; it is reported to affect men more than women, with a ratio of 2:1. The most commonly affected bones are the skull, spine, extremities and maxilla.

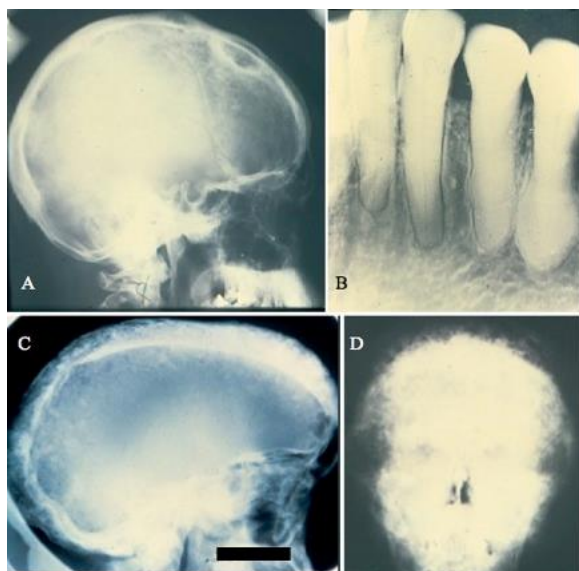


Fig. 3. Paget's disease of bone. A; lateral skull view illustrates early osteolytic stage of Paget's disease as shown by a well-defined radiolucent area in the frontal bone (osteoporosis circumscripta). B; intraoral periapical film of Paget's disease showing hypercementosis of the roots. C; lateral skull view of the osteolytic-osteoblastic phase of Paget's disease. Note the "cotton-wool" appearance of the bone that is pathognomonic for Paget's disease. D; PA skull view of the late osteoblastic stage

There is usually progressive enlargement of the skull, bowing of long bones, and spinal kyphosis. Enlargement of the skull is usually associated with neurological deficits, e.g., blindness, deafness, facial paralysis. The jaws are involved in only 20% of cases; the maxilla is more frequently affected than the mandible (3:1 ratio). [5] The progressive enlargement of the maxillary and/or mandibular alveolar process(s) results in spacing of the teeth and malocclusion. Dental extraction could be complicated by prolonged socket bleeding and osteomyelitis in the early and late stages of the disease; respectively. Dentures may have to be replaced to accommodate the continued enlargement of the alveolar ridges. The enormously increased vascularity in pagetoid bone results in a high cardiac output, cardiac enlargement, and eventually cardiac failure. Other complication of this disease is sarcomatous transformation in about 10% of patients. [6] In Paget's disease of bone, the serum alkaline phosphatase level is extremely increased although serum calcium and phosphate levels are normal. [5, 7] The radiology of Paget's disease varies with the stage. In the early stage, the affected bone shows diminished density. In the skull these changes are called "osteoporosis circumscripta". These lytic lesions consist of single or multiple circumscribed areas of reduced density, usually starting in the outer cranial table.

Citation: Libyan Dent J 2015, 5: 17121232 -<http://dx.doi.org/10.5542/LDJ.v5i0.17121232>

The affected bones are softened during the early phase and subject to weight effects. If the cranial base is involved, softening results in platybasia (basilar invagination), i.e., abnormal flattening of the skull base. The appearance is described as a "Tam O'Shanter" skull, which is named after the Scottish cap. [6] In the biphasic stage, the pagetoid bone randomly fills the lytic spaces resulting in patchy radiopacities described classically as "cotton-wool" appearance (Figure 3). Involvement of the maxilla and malar and frontal bones produces a leonine appearance described as "leontiasis ossea". As the disease advances, deposition exceeds resorption. The lamina dura around the teeth in the involved regions may be absent. The teeth are usually hypercementosed, typically the premolars. [6, 7]

Hyperparathyroidism

Hyperparathyroidism is an endocrine disorder in which there is persistent excess of circulating parathyroid hormone (PTH) levels. PTH increases blood calcium and phosphate levels by mobilizing calcium from the skeleton and decreasing renal tubular reabsorption of phosphate. Hyperparathyroidism occurs in three clinical forms: primary, secondary, and tertiary.

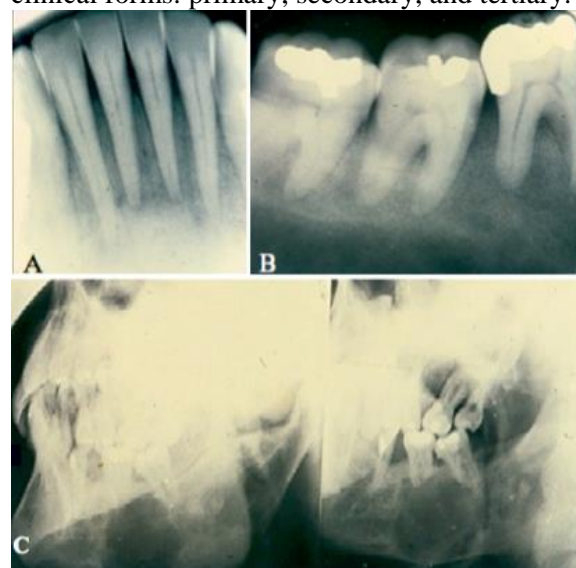


Fig. 4. Hyperparathyroidism. A, B; intraoral radiographs of cases with secondary hyperparathyroidism demonstrate loss of lamina dura and granular pattern of alveolar bone. C; lateral oblique radiographs of a different case with primary hyperparathyroidism shows a brown tumor of the mandibular body causing cortical expansion and root resorption.

Primary hyperparathyroidism is usually caused by a benign tumor (adenoma) of the parathyroid gland, producing excessive amounts of PTH. Secondary hyperparathyroidism is usually the

result of chronic renal insufficiency (hence called renal osteodystrophy) that in turn results in persistent hypocalcemia, thereby the parathyroid glands are prompted to secrete excessive PTH in response to the low serum calcium levels. Secondary hyperparathyroidism, when chronic, called tertiary hyperparathyroidism. [8] Hyperparathyroidism could be complicated by formation of renal stones. The classic radiographic features of hyperparathyroidism include subperiosteal bone resorption, generalized demineralization, localized ill-defined giant cell lesions, known as brown tumors and osteitis fibrosa cystica; loss of lamina dura, and metastatic calcification of soft tissues (Figure 4). The skeleton undergoes trabecular alterations that are manifested radiographically as “ground glass” appearance owing to the granular trabeculation pattern. [9] Observation of widened PDL spaces and tapered roots is relative to the loss of lamina dura.

Hyperpituitarism

Hyperpituitarism is an endocrine disease where there is increased production of growth hormone, causing continued growth of body organs. This disorder is usually caused by a benign tumor of the anterior lobe of the pituitary gland. If hyperpituitarism occurs during childhood, it results in gigantism; if it occurs during adulthood, the result is acromegaly. In acromegaly, there is no further increase in height because the adult bones are incapable of further growth as the onset of the abnormality occurs after epiphyseal and sutures closures of the appendicular and craniofacial skeletons; respectively.



Fig. 5. Acromegaly. A, B; PA and lateral skull views show excessive growth of the mandible resulting in skeletal class III malocclusion, enlarged paranasal sinuses and mastoid processes, and prominence of frontal bone and supraorbital ridges.

However, there is progressive subperiosteal deposition of bone, resulting in increased thickness of extremities, skull, and jawbones. Progressive growth of the mandible (mandibular prognathism) and tongue (macroglossia), results

in spacing and flaring of anterior teeth and development of class III skeletal malocclusion (Figure 5). The posterior teeth may be hypercementosed in response to increased functional demands. [1] The trabecular pattern is relatively normal though. A lateral skull radiograph of an acromegalic patient typically demonstrates a prognathic mandible, enlarged paranasal sinuses, enlarged sella turcica, and frontal and occipital bossing. [10]

Rickets and Osteomalacia

Rickets and osteomalacia result from deficiency in serum calcium and phosphate required for normal calcification of bone and teeth due to any disorder in the vitamin D-phosphorus-calcium axis. Rickets occurs in infants and children, whereas osteomalacia occurs in adults.

In rickets, hypomineralized bone matrix results in bowing of weight-bearing bones (e.g., femur and tibia), increased widening of their epiphyses, and thinning of cranial tables. Greenstick fractures do occur in rare cases of rickets. Jaws and teeth may show loss of cortication, hypoplastic enamel, delayed eruption, and abnormally large pulp chambers (Figure 6). In severe cases, no cancellous bone is seen and the teeth appear to be floating in air. [11] In osteomalacia the teeth are not affected because they are fully developed before the onset of the disease.

Osteoporosis

Osteoporosis is a generalized reduction of bone mass without any change in size. The decreased mass is the result of the reduction in thickness and trabeculation of cortical and cancellous bones; respectively. Trabecular bone is affected earlier than cortical bone with the vertebral column is the primary involvement site. Osteoporosis usually occurs in the elderly, postmenopausal women, Cushing's syndrome, and patients on corticosteroids. During menopause, there is diminished production of estrogen, which tends to protect the skeleton against the demineralizing action of PTH. [12] Osteoporotic bones are more prone to fracture. On a radiograph, osteoporosis is evident as generalized rarefaction and cortical thinning of jaws, most notable in the mandible (Figure 7). There is reduction in the overall quantity of trabeculae in the cancellous component. [13]

Systemic Sclerosis (Scleroderma)

Systemic Sclerosis is a rare connective tissue disease that results in excessive production and sclerosis (hardening) of collagen fibers. It is more

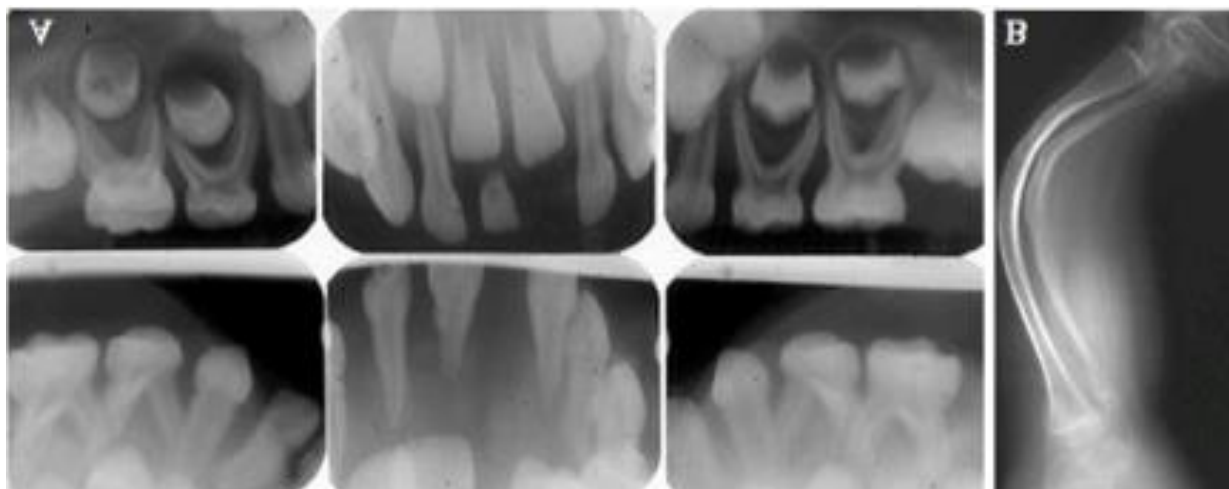


Fig. 6. 6-year-old girl with rickets. A; full mouth series shows loss of follicular cortication and delayed eruption. B; leg radiograph shows severe bowing deformity of tibia and fibula.

common in women than in men (10:1). Progressive fibrosis of visceral organs like the heart, lungs and kidneys may result in serious complications and death. The facial skin and oral mucosa are rigid and tense, and thickening of the perioral tissues may result in limited mouth opening (microstomia). [14] Widening of periodontal ligament space marks thickening of periodontal fibers (Figure 8). Mandibular erosive bone changes at areas of muscular attachment are seen in approximately 25% of patients. [15]

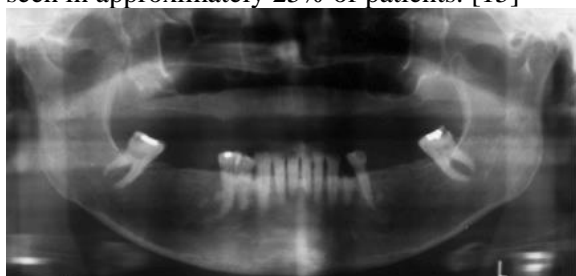


Fig. 7. Osteoporosis. Panoramic radiograph of a 65-year-old white woman shows cortical thinning of the mandibular inferior cortex, external oblique ridges, and inferior alveolar canals. The cancellous bone is darker than normal although the teeth are exposed correctly; trabeculae are fewer in number.



Figure 8. Systemic sclerosis. Intraoral periapical film of a patient with systemic sclerosis showing widened periodontal ligament space more pronounced in the coronal portion of the roots.

Multiple myeloma

Myeloma is a neoplastic proliferation of plasma cells that may occur in several forms. Multiple myeloma is the most common bone malignancy in adult. Approximately 80% of cases occur between the ages 40 to 70 years, with twice the incidence in males as in females. [16] The disease is associated with urinary hypergammaglobulinemia and Bence-Jones proteins, which cause the urine to be foamy. Radiographically, multiple myeloma is characterized by sharp non-corticated lytic lesions lacking any signs of bone reaction “punched-out”, ranging in size from a few millimeters to several centimeters. Mandibular lesions are observed in about 33% of myeloma patients and may be the first bony manifestation of the disease. [16] Aggressive little lesions may coalesce, giving the appearance of multilocularity. Other radiographic features include generalized osteopenia and cortical effacement especially in the spine and pelvis (Figure 9).

CONCLUSION:

Abnormalities of the teeth and jaws are occasionally potential indicators of underlying systemic diseases. Oral and maxillofacial radiologists must be acquainted with systemic conditions manifesting in the dento-maxillofacial complex, so that early diagnosis can be made. The author highlights specific conditions that are radiographically manifested in the oral and maxillofacial region, and hope to provide ample insight for radiologists, dentists, and physicians.



Fig. 9. 73-year-old patient where lateral and PA skull views show a widespread and almost patternless areas of radiolucencies that were suggestive of multiple myeloma (A, B). Punched-out radiolucencies were also noticed on an intraoral film of the mandibular right premolar (C). Further radiographic examination showed areas of radiolucencies in the bones of the hands (D), the radius (E), and widespread destruction of the pelvic bones (F). Bone biopsy confirmed the diagnosis of multiple myeloma, although urine analysis was negative for Bence-Jones proteins.

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