Clinical and Radiographic Features of Chronic Monostotic Fibrous Dysplasia of the Mandible

(Caractéristiques cliniques et radiographiques de la dysplasie fibreuse monostotique chronique du maxillaire inférieur)

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Sommaire

Cet article décrit une dysplasie fibreuse chronique non traitée du maxillaire inférieur chez un homme de 40 ans, en insistant sur les résultats radiographiques. Selon les auteurs, il s'agirait du premier cas du genre décrit dans la littérature. À l'intérieur de cette lésion mandibulaire mature, une vaste radiotransparence a été observée, celle-ci évoquant un kyste osseux simple. Le patient ne ressentait aucun symptôme directement relié à la lésion mandibulaire. Divers aspects du diagnostic, de l'apparence radiographique et du diagnostic différentiel sont examinés. Ces renseignements seront utiles à tous les dentistes, spécialistes en chirurgie buccale et maxillofaciale, médecins et autres fournisseurs de soins, pour reconnaître l'aspect des lésions fibro-osseuses chroniques.

Mots clés MeSH : *diagnosis, differential; fibrous dysplasia of bone; mandibular diseases/diagnosis*

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Fibrous dysplasia is a disturbance of bone metabolism that is classified as a benign fibro-osseous lesion. Fibrous connective tissue containing abnormal bone replaces normal bone. The etiology of fibrous dysplasia is unknown. The radiographic appearance of the irregularly shaped trabeculae aids in the differential diagnosis. Occurring most commonly in the second decade of life, the lesions of fibrous dysplasia can be surgically recontoured for esthetic or functional purposes once they become dormant.[1]

Case Report

A 40-year-old man was referred for panoramic radiography to the oral and maxillofacial radiology clinic at the New Jersey Dental School in Newark by the emergency clinic in the medical centre at the same institution. The patient, a cab driver, initially complained of pain in his back, neck and shoulder radiating to the jaws. The patient reported that clinically evident expansion of his left mandible (**Fig. 1**) had been present for at least 20 years. The clinical examination revealed that the expansion of the left mandible was diffuse and bony hard. There were no apparent changes in the skin or the intraoral mucosa adjacent to the swelling. When the patient presented to the medical centre earlier, extraoral plain film radiographs had been obtained, including a posteroanterior (PA) view, a lateral oblique view of the left mandible and reverse Towne views (**Fig. 2**), and these radiographs were available to the radiology clinic through the hospital's Web-based Picture Archiving and Communication System.

However, the existence of the skull films was not known at the time of the patient's presentation to the radiology clinic, and the requested panoramic radiograph was obtained (**Fig. 3**). A high panoramic view (**Fig. 4**) was subsequently obtained to visualize the most superior extent of the lesion, which was not visible on the primary panoramic film (**Fig. 3**). The panoramic projections revealed diffuse enlargement of the left mandible, extending from the left canine area to the condyle and encompassing the inferior border, the alveolar crest, the ramus and the coronoid process. The anterior border of the lesion appeared reasonably well demarcated because of superimposition of the lesion over adjacent normal bone. Radial expansion of the lesion was evident on the panoramic, PA, cephalometric and reverse Towne views, and the lesion obliterated the cortical bone along the inferior border of the mandible. In fact, the inferior border of the mandible was entirely replaced by the

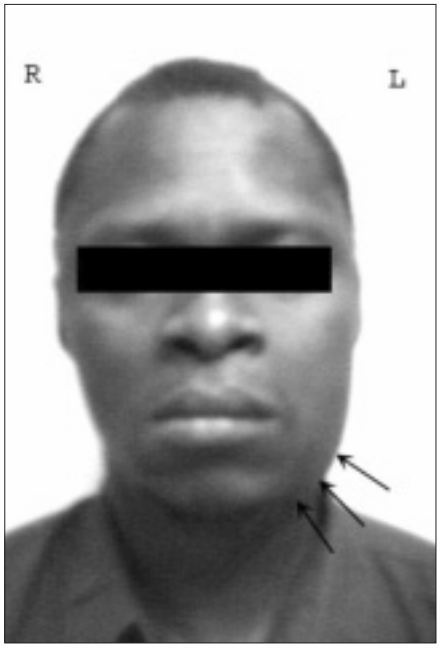

Figure 1: *Photograph of the patient at the time of presentation. Arrows indicate the swelling of the left mandible.*

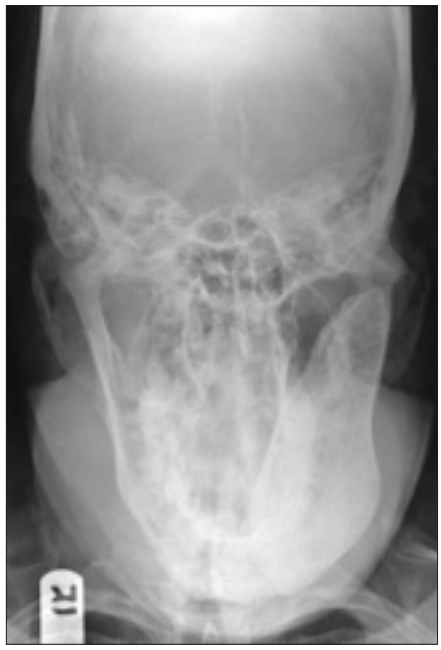

Figure 2: *Reverse Towne view demonstrating the extent of the lesion in the buccolingual and inferio-superior dimensions.*

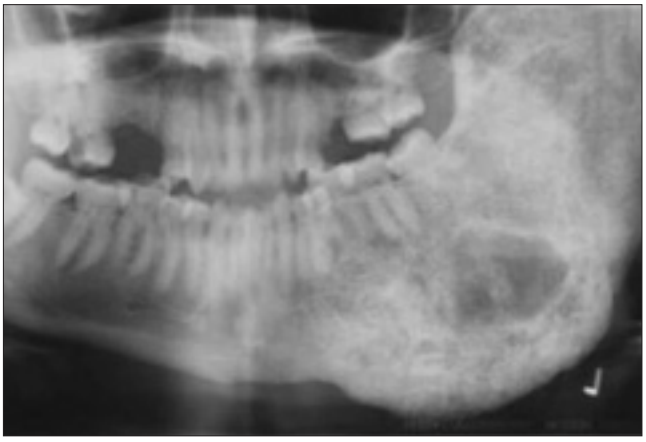

Figure 3: *Panoramic view showing the anteroposterior and inferior extent of the lesion. The superiormost extent of the lesion is not visible.*

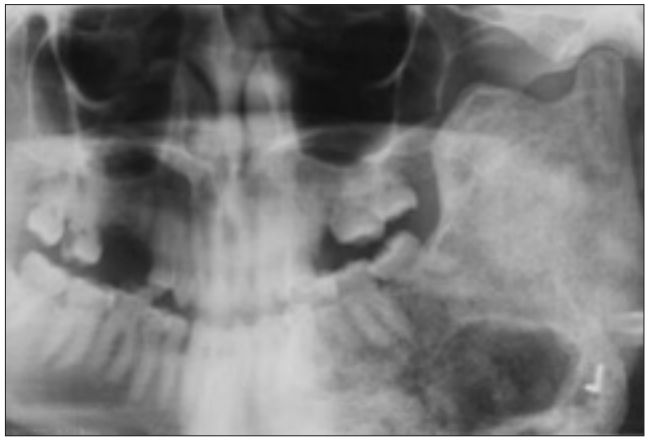

Figure 4: *High panoramic view capturing the remodelled neck of the condyle and the coronoid processes of the mandible.*

lesion. The posterior mandibular teeth on the affected side were displaced superiorly, and the lesion had evidently altered occlusion. Although the head of the condyle appeared to have relatively normal morphology, the neck of the condyle and the coronoid process were entirely involved by the lesion.

Internally, the bulk of the lesion had a mixed radiopaque–radiolucent texture with an altered trabecular pattern. In the centre of the lesion, there was a diffuse radiolucent area about 3 cm x 3 cm surrounded by a hyperostotic rim. This area appeared less ossified than the surrounding areas of the lesion.

The patient was eventually referred to the oral and maxillofacial surgery department at the university hospital for management, where the attending surgeon prescribed a computed tomography (CT) examination. The axial CT images confirmed the unilocular radiolucency within the bony lesion located posteroinferiorly within the body of the mandible. With the bone window setting, the lesion appeared roughly round with regular borders and no ossified contents. The majority of the central core had a density consistent with soft tissue (**Fig. 5**). Coronal CT confirmed expansion of the ramus in the mediolateral plane (**Fig. 6**). The bone morphology of the functional portion of the head of the condyle appeared relatively unaltered. Although biopsy was contemplated, the patient refused any further investigative procedures or treatment for the mandibular lesion. He requested treatment for his presenting complaint only, which was unrelated to the mandibular lesion.

Because the patient's initial complaint was the radiating

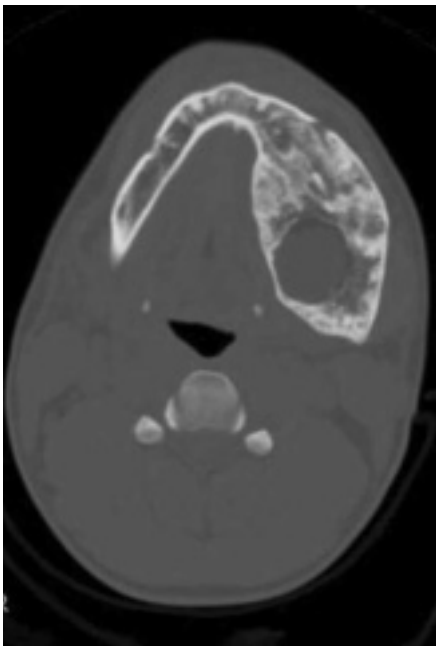

Figure 5: *Axial computed tomography view of the mandible (bone window setting). The centre of the lesion is radiolucent.*

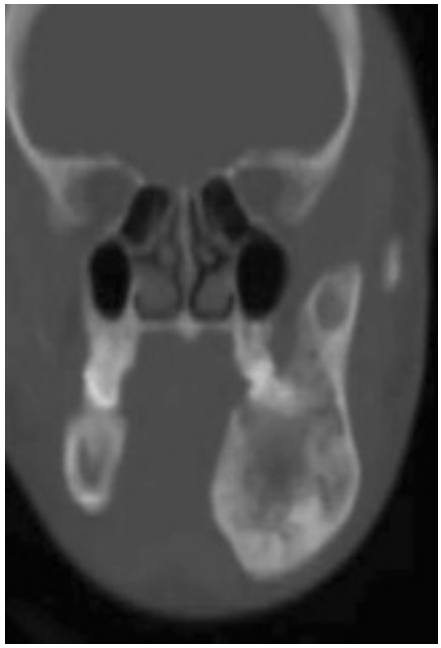

Figure 6: *Coronal computed tomography view of the mandible (bone window setting) demonstrating the unaffected right mandible as well as the affected left mandible.*

pains from his back and shoulder to the jaws, he was treated symptomatically for his musculoskeletal condition of postural origin, possibly related to his occupation as a taxi driver. He was discharged with an appointment for follow-up regarding his musculoskeletal problem.

Discussion

Fibrous dysplasia is a developmental anomaly in which normal bone is replaced with fibrous connective tissue. As the lesion matures, the fibrous connective tissue is replaced with irregularly patterned trabecular bone.[1] Fibrous dysplasia is a localized abnormality, which can involve one (monostotic) or multiple (polyostotic) bones. A recent study described various forms of fibrous dysplasia occurring within a Hong Kong population.[2] The forms were distributed as follows: 74% monostotic, 13% polyostotic and 13% craniofacial.[2] The monostotic form generally occurs during the second decade of life and becomes dormant by the third decade. Hormonal changes, such as those seen in pregnancy, can reactivate a dormant lesion.[2] The craniofacial form of fibrous dysplasia can be diffuse and may involve multiple bones. When the anatomic spaces and foramina are constricted because of encroachment of the lesions, the patient may experience a variety of symptoms, including headaches, loss of vision, proptosis, diplopia, loss of hearing, anosmia, nasal obstruction, epistaxis, epiphora and symptoms mimicking sinusitis.[3]

With initial development of fibrous dysplasia the patient usually reports facial swellings and asymmetries. Although the lesion is usually asymptomatic, encroachment on canals and foramina, as well as limitations of movement, may engender complaints of pain and discomfort. In general, males and females are thought to be affected evenly, although recent research has shown a slight female preponderance.[2] However, McCune-Albright syndrome, a form of polyostotic fibrous dysplasia associated with café-au-lait pigmentation and multiple endocrinopathies such as precocious puberty, pituitary adenoma or hyperthyroidism, almost always affects females.[1]

The lesions of fibrous dysplasia are twice as common in the maxilla as the mandible, and the posterior aspects of the jaw are more frequently affected than the anterior. Treatment usually involves bony recontouring at the affected site to improve esthetics and function. Recurrence is rare in adults, but the lesions can show surprising growth potential if they are surgically altered during their active growth phase.[1] Other lesions to be considered in the differential diagnosis include inflammatory lesions, fibro-osseous lesions, and benign and malignant neoplasms.[4]

In a systematic review of previous studies of fibrous dysplasia, McDonald-Jankowski[2] determined that a greater proportion of females than males were affected and that the maxilla is the most common facial bone affected.[2] The most common presenting complaints were swelling in 94% of reported cases and pain in 15%.[2]

The case described here is unusual because the patient reported that the lesion had been present, without surgical intervention, for more than 20 years. In addition, the radiographic appearance of the internal architecture of the lesion was not consistent with the common descriptions of fibrous dysplasia, which typically refer to a salt-and-pepper, orange peel, ground glass or thumb print appearance.[1] The trabecular pattern appeared irregular and thickened, with no discernible orientation. Radiolucent lesions resembling cysts

occasionally occur in mature lesions of fibrous dysplasia.[5] These bone cavities are analogous to simple bone cysts.[1] Such a bone cavity was present in this case.

In McDonald-Jankowski's study, the most common radiographic presentation of fibrous dysplasia was a poorly defined, ovoid (fusiform) area of dysplastic bone exhibiting a ground glass appearance.[2] In fibrous dysplasia of the mandible, the mandibular canal may be displaced either inferiorly or superiorly. Petrikowski and others[6] suggested that upward displacement of the mandibular canal may be unique to fibrous dysplasia and could be pathognomonic. In the case presented here, the mandibular canal appeared to have been repositioned inferiorly. Although the borders of fibrous dysplasia are known to be ill-defined,[1] they can appear well defined in panoramic and plain skull films (as in this case) if the border of the expansile portion of the lesion is superimposed over the mandible (**Fig. 5**). Loss of lamina dura due to replacement of normal bone may be one of the diagnostic signs of fibrous dysplasia.[6] In chronic cases, the lesion tends to become increasingly more radiopaque.

The literature suggests that fibrous dysplasia in women can be reactivated during pregnancy.[7–10] This association is more commonly seen with the polyostotic form. Cystic lesions resembling aneurysmal bone cysts have been noted in association with the monostotic form.[11]

Panoramic, reverse Towne, PA and lateral skull views are often adequate to visualize lesions in the mandible. It is desirable to have at least 2 images, exposed at right angles, to assess the extent of the lesion in all dimensions. Because of the complexity of the anatomy, CT is helpful for assessing lesions in the maxilla.[2,3,12]

Differential diagnosis of the initial radiolucent stage must include the following: central ossifying fibroma (COF), central giant cell granuloma (CGCG), aneurysmal bone cyst, osteomyelitis and early fibro-osseous lesions.[13] Because these lesions represent a variety of disease processes with different behaviours, including infection and endocrine dysfunction, prompt diagnosis incorporating clinical, radiographic and, occasionally, histologic findings, is essential.

COF is a benign neoplasm that commonly has a radiographic and histological appearance similar to that of fibrous dysplasia.[14] Tissue sections of COF show a cellular or sclerotic fibrous connective tissue stroma containing numerous osseous trabeculae of various sizes associated with prominent osteoblasts.[15] A mixture of lamellar and woven bone is typically seen. Often, there are also scattered ovoid calcifications that resemble cementum. In contrast to fibrous dysplasia, a well-defined capsule occasionally surrounds the lesion. Radiographically, a well-defined margin is consistent with COF, whereas the margins of fibrous dysplasia tend to merge with the surrounding normal bone. On occasion, a sclerotic border, absent from fibrous dysplasia, is also seen in COF. COF occurs mostly in the third and fourth decades of life, whereas fibrous dysplasia is most often discovered in the second decade. COF is more common in the mandible, tends to occur in anterior regions and is smaller in size, whereas fibrous dysplasia is more common in the posterior maxilla, and the lesions tend to be larger. Both lesions tend to expand the bone cortex.[9]

Differentiation of these 2 lesions is critical because the treatment protocols are quite different. COF, although benign, must be enucleated due to its potential to recur. Fibrous dysplasia is generally self-limiting and does not require treatment except for cosmetic reasons, pain, discomfort or impaired function.[2] If undertaken, the treatment, consisting of recontouring or resection, should be postponed until after cessation of skeletal growth, since early treatment may accelerate growth of the lesion.

Early-stage fibrous dysplasia should also be differentiated from CGCG, which more commonly appears in the anterior mandible, results in generally painless expansion of bone and appears radiographically as unilocular or multilocular radiolucent defects with well-delineated, noncorticated margins.[9]

Fibrous dysplasia may also mimic Paget's disease of bone on clinical examination, particularly if a patient with fibrous dysplasia does not present until later in life. In addition to the predilection of Paget's disease for an older population, certain radiographic and clinical features help to distinguish this lesion from other radiographically similar lesions. These features include thickening of the cortices, cotton wool appearance of the involved bone and increased blood levels of alkaline phosphatase.[1] The most useful clinical feature for distinguishing Paget's disease from fibrous dysplasia is that the former tends to occur bilaterally in the jaws, whereas the latter affects only one side. Histologically, Paget's disease exhibits many osseous trabeculae with prominent reversal lines showing simultaneous osteoblastic and osteoclastic activity.[10] The affected bone resides within a well-vascularized fibrous connective tissue stroma.

Although osteomyelitis demonstrates sequestra in the later stages, it may resemble fibrous dysplasia in the early stages, especially if there is associated swelling. Usually, inflammatory signs and the presence of draining sinus tracts are indicative of osteomyelitis. Periosteal new bone, manifesting as one or more laminations occurring parallel to the outline of the jaw, often occurs in osteomyelitis affecting young patients and is a useful indicator of the condition. Osteomyelitis may occur secondary to odontogenic infections of pulpal origin, although hematogenous spread from distant sites has also been reported. Once the offending tooth has been treated, the lesion often resolves spontaneously.[9] Chronic osteomyelitis superimposed on fibrous dysplasia can mimic a malignant lesion even when advanced imaging studies like magnetic resonance imaging (MRI) are utilized. Chang and others[4] described a 6-year-old girl who presented with local pain over the right chin after severe trauma. The lesion was originally diagnosed as chronic osteomyelitis, but after failure of antibiotic treatment, a malignant bone tumour was suspected on the basis of imaging studies including MRI. Fibrous dysplasia of the mandible in association with chronic osteomyelitis was confirmed in this case by repeat biopsy one year after initial onset. Reasons for the delay in diagnosis in this case might have included the similar clinical and radiographic characteristics of fibrous dysplasia of the mandible and chronic osteomyelitis; also, the initial biopsy was not performed at the appropriate site.[4]

Diffuse sclerosing osteomyelitis (DSO) is a sequela of chronic jaw infection and inflammation. It presents radiographically as an ill-defined radiopacity, often encompassing large areas of bone, which may exhibit small radiolucent zones.[16] The affected bone does not show expansion unless the infection involves the cortical plate, which may induce a periosteal reaction. Histologically, DSO demonstrates sclerotic bone showing alternating areas of apposition and resorption. Between the bone trabeculae lies fibrous connective tissue infiltrated by chronic inflammatory cells. Fibrous dysplasia can often be differentiated from osteosarcoma on the basis of radiographic appearance.[6] The radiographic features of osteosarcoma are orthoradial striations, destruction of cortices with an outgrowth of the soft-tissue component, generalized widening of the periodontal ligament spaces and destruction of the lamina dura. Histopathologic examination of the bone is indicated in all cases where DSO or osteosarcoma is suspected.

Although there was no compelling indication to seek a biopsy in the case described here, any sudden change in the clinical presentation or behaviour of the lesion might warrant further investigation. ◆

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