Unilateral enlargement of the mandible in a child

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CLINICAL PRESENTATION

A 6-year-old Asian male presented to the oral and maxillofacial surgery department of the Children's Hospital of Philadelphia (CHOP) with a 5-month history of recurrent right mandibular pain, swelling, and trismus, with no associated fever. His private dentist evaluated him at the onset of symptoms and attributed his pain to a carious right mandibular deciduous first molar. The tooth was extracted without resolution of the patient's symptoms. He was subsequently diagnosed with parotitis by his pediatrician and was prescribed amoxicillin. The swelling transiently improved on completion of the amoxicillin treatment but recurred after 1 month. The patient's pediatrician referred him to the CHOP for further management.

The patient's medical and surgical histories were noncontributory, and his childhood immunizations were up to date. He had no known drug allergies. At the time of presentation to our clinic, he was not taking any medications. His family history was significant for rheumatoid arthritis and hyperthyroidism in his paternal grandmother. He was living at home with his parents and one brother and was progressing well in school. He was developmentally appropriate for his age, and the review of systems was normal, except for the presenting complaint.

Clinical examination revealed a diffuse, firm swelling of the right mandible with buccal and lingual expansion and no suppuration. The lesion was tender to palpation, and the patient's interincisal opening was limited. There was no obvious dental source of infection to explain the swelling and no reported history of trauma.

A panoramic radiograph revealed diffuse expansion of the right mandibular body, angle, ramus, condyle, and coronoid process, with flattening of the angular notch (Figure 1). Small osteolytic areas were also noted. A computed tomography scan revealed a mixed density sclerotic/lytic lesion involving the entire right mandible, with buccolingual expansion, thinning of the buccal and lingual cortices, and associated extensive soft tissue swelling (Figure 2).

DIFFERENTIAL DIAGNOSIS

Based on the clinical presentation and radiologic features, our differential diagnosis included fibrous dysplasia (FD), primary chronic osteomyelitis (PCO), congenital hemifacial hyperplasia/hypertrophy, and hemimandibular hyperplasia.

FD is a noninherited developmental skeletal disorder in which normal medullary bone is replaced by immature and disorganized fibro-osseous tissue.[1] Genetic studies have shown that it arises from postzygotic mutations in the *GNAS1* gene located on chromosome 20q13.[2] FD may involve a single bone (monostotic) or may involve multiple bones at the same time (polyostotic). The phrase "craniofacial FD" is used to describe the contiguous involvement of the bones of the craniofacial complex. Most patients are diagnosed in the first and second decades of life, and the lesion stabilizes when skeletal growth is completed.[1] In the maxillofacial region, the most common presentation is that of a unilateral, slowly progressing asymptomatic expansion of the involved bone, which may result in asymmetry and subsequent disfigurement. The maxilla is involved twice as frequently as the mandible, with the lesion mostly located posteriorly.[1] Dental malocclusion and tooth displacement may also occur.

Radiographic presentation of FD varies considerably, depending on disease activity, and can range from a radiolucent lesion to a mixed density or radiopaque lesion. However, the classic presentation in the maxillofacial region is described as having a "ground glass" or "orange peel" appearance with ill-defined borders, and the lesion merges imperceptibly with the surrounding normal bone.[1] FD was considered in the differential diagnosis in this case because of the diffuse bony expansion. The patient's age and the insidious nature of the disease process also suggested FD. However, the 5-month history of recurrent mandibular pain, swelling, and trismus, with transient improvement of the swelling following antibiotic therapy, was inconsistent with a diagnosis of FD.

PCO,[3] an uncommon nonsuppurative inflammatory disease was considered a possible diagnosis for our

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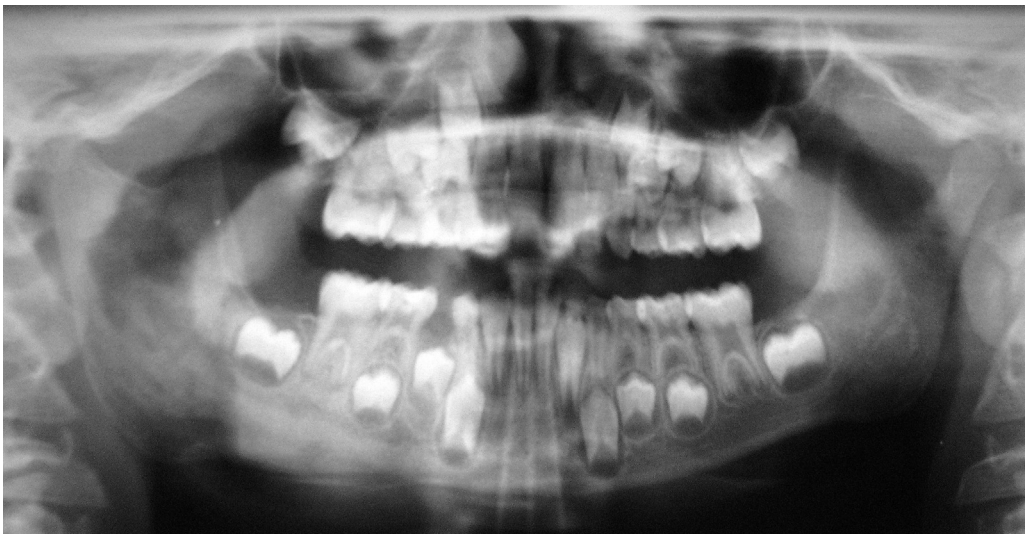

Fig. 1. Panoramic radiograph. There was diffuse expansion of the right mandibular body, ramus, condyle, and coronoid process, with flattening of the mandibular notch.

patient. PCO mostly involves the mandible and is not restricted to any age group. It is usually characterized by diffuse enlargement of the mandible, accompanied by intermittent episodes of pain, swelling, trismus, and regional lymphadenopathy.[3] PCO is generally associated with a protracted clinical course without an acute stage and is often refractory to antibiotic treatment.[3,4] The episodes occur periodically, with varying degrees of intensity and can last from a few days to several months. Unlike chronic (secondary) osteomyelitis, PCO is not associated with sequestration, fistulae formation, or purulence, and there is no obvious dental source of infection.[3,4]

Radiographically, PCO presents as a mixed-density lesion with areas of osteolysis and sclerosis with disruption of normal trabecular pattern.[3] Our patient's clinical as well as radiographic presentation was reminiscent of PCO. The absence of purulence, lack of an odontogenic source of infection, recurrence on completion of antibiotic therapy, and the radiographic presentation of a mixed-density lesion further favored PCO as a possible diagnosis.

Hemifacial hyperplasia/hypertrophy is a rare congenital anomaly characterized by unilateral overdevelopment and overgrowth of bone, teeth, and soft tissue on the affected side.[5] The deformity is usually present at birth but becomes more accentuated with age, being more noticeable at puberty. The abnormal growth terminates when skeletal maturity is completed.[6] The etiology is unknown, but various factors have been considered, including hormonal imbalance; neural, lymphatic, and blood vessel abnormalities; incomplete twinning; chromosomal abnormalities; and localized alteration of the intrauterine environment.[6] The teeth on the affected side are abnormal in their crown size, root size and shape, and rate of development. Soft tissue abnormalities can involve the uvula, tonsils, lips, buccal mucosa, and tongue.

We considered hemifacial hyperplasia in the differential diagnosis because of the unilateral enlargement of the orofacial region in the young patient. However, his teeth were similarly sized in both mandibular quadrants, and he had no soft tissue abnormalities involving the uvula, tonsils, lips, buccal mucosa, and tongue. Also, his radiographs revealed the presence of a mixed-density mandibular lesion, thereby making the diagnosis of hemifacial hyperplasia unlikely.

Hemimandibular hyperplasia (hypertrophy) is a developmental disorder characterized by progressive unilateral enlargement of the mandible.[7] Patients present with asymmetry and diffuse overgrowth of the mandibular condyle, ramus, and body, and the overgrowth terminates at the symphysis. The deformity invariably results in malocclusion. In contrast to congenital hemifacial hypertrophy, in which the defect is present at birth, in mandibular hemihypertrophy, the enlargement occurs postnatally, usually at the age of 5 to 8 years and occasionally later.[7] The etiology of hemimandibular hyperplasia remains obscure. It may be a multifactorial disorder, with heredity, trauma, hypervascularity, and hormonal influences all being considered in the etiology.[7]

Hemimandibular hyperplasia was considered in the differential diagnosis because our patient presented with unilateral expansion of the mandible at 6 years of age, and his panoramic radiograph revealed diffuse expansion involving the right mandibular condyle, coronoid process, ramus, and body. However, his presentation involved pain, swelling, and trismus, which are not usually associated with hemimandibular hyperplasia.[7] Also, his computed tomography scan showed a mixed-density lesion with thinning of the cortices, which is not consistent with hemimandibular