Case Report

Segmental odontomaxillary dysplasia

An underrecognized entity

Molly Housley Smith, DMD; Donald M. Cohen, DMD, MBA, MS; Joseph Katz, DMD; Indraneel Bhattacharyya, DDS, MSD; Nadim M. Islam, DDS, BDS

ABSTRACT

Background and Overview. Segmental odontomaxillary dysplasia (SOD) is a characteristic developmental abnormality that demonstrates posterior maxillary enlargement, dental abnormalities, altered bone trabeculation, and possible cutaneous findings. Only 62 cases have been reported in the English-language literature.

Case Description. The authors described 3 newly diagnosed cases of SOD, all found in the left posterior maxilla in adolescent female patients; they reviewed the literature to elucidate this rare entity.

Conclusions and Practical Implications. Because of the complexity and variety of associated dental and craniofacial features, patients with SOD may seek diagnosis from various dental and medical providers. The signs of SOD are characteristic, yet the condition is largely underrecognized among health care professionals, which may lead to unnecessary treatment.

Key Words. Segmental odontomaxillary dysplasia; fibrous dysplasia; oral; maxillary expansion; HATS; missing teeth.

JADA 2018:149(2):153-162
https://doi.org/10.1016/j.adaj.2017.08.007

Segmental odontomaxillary dysplasia (SOD) is a rare developmental condition that affects the posterior maxillary region. To date, only 62 well-documented cases, including 3 we share in this article, have been reported in the English-language literature.[1-26] The condition was originally termed "hemimaxillofacial dysplasia" by Miles and colleagues[16] in 1987 but was later renamed "segmental odontomaxillary dysplasia" by Danforth and colleagues[7] in 1990 to highlight the dental abnormalities important in its diagnosis.

SOD is characterized by unilateral expansion of the maxillary alveolus, dental abnormalities (for example, congenitally missing, overretained, impacted, or hypoplastic teeth), mild facial asymmetry, altered radiographic pattern, and possible skin manifestations. Because of the wide range of features, patients may seek care from multiple different dental and medical providers before a correct diagnosis is made, including general dentists, oral and maxillofacial surgeons, pediatric dentists, orthodontists, endodontists, periodontists, oral pathology or medicine specialists, oral radiologists, dermatologists, plastic surgeons, otolaryngologists, pediatricians, or primary care physicians. Although the signs of SOD are classic, lack of widespread recognition of the entity may lead to underrecognition among health care providers.

We describe 3 newly diagnosed cases of SOD, review the English-language literature, and discuss clinical and histopathologic characteristics to promote early recognition of this distinctive developmental anomaly and prevent unnecessary treatment.

CASE 1

A 14-year-old girl was brought to our dental clinic for a comprehensive examination. Her medical history was significant for attention deficit/hyperactivity disorder (ADHD), asthma, and seasonal allergies.

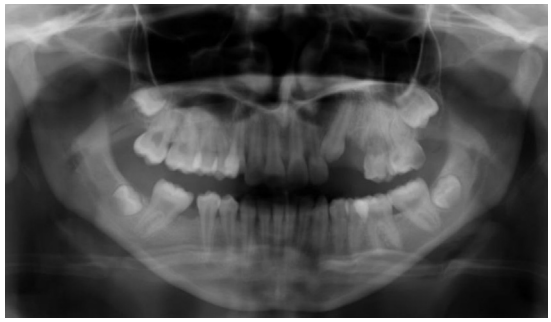

Figure 1. Panoramic image of the patient in case 1, a 14-year-old girl, who had ill-defined, coarse trabeculation of the left posterior maxilla with impacted canine, missing permanent premolars, and overretained primary molar.

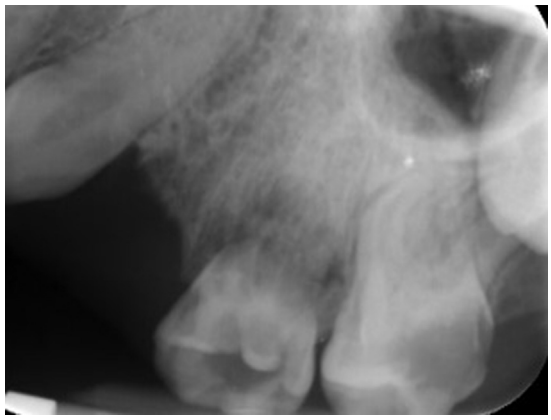

Figure 2. Periapical radiograph of maxillary left alveolus in case 1 showing blending of the dental roots osteocytes with abnormal, vertically oriented, bony trabeculation.

Her medications included

- methylphenidate (Concerta, Janssen), clonidine, and aripiprazole (Abilify, Otsuka) for ADHD;
- an albuterol inhaler and fluticasone propionate and salmeterol (Advair, GSK) for asthma;
- mometasone furoate monohydrate (Nasonex, Merck) for seasonal allergies.

The patient reported no personal or familial history of syndromes, and her family history for craniofacial anomalies was negative. Extraoral examination revealed no abnormal cutaneous changes.

On clinical examination, generalized gingivitis, significant plaque accumulation, and multiple carious lesions were noted. Numerous dental abnormalities were noted, including overretained tooth no. J; missing teeth nos. 12, 13, 14, and 30; unerupted teeth nos. 1, 16, 17, and 32; and impacted tooth no. 11.

Her clinician obtained panoramic and periapical radiographs, which revealed abnormal bony trabeculation in the mandibular left quadrant with vertically oriented trabeculation (Figures 1 and 2). The clinician suggested a diagnosis of fibrous dysplasia based on radiographic presentation.

Because of significant caries and extensive root resorption, her clinician extracted tooth no. J. Tooth no. 15 showed caries extending to the pulp and dilacerated roots. Her clinician attempted root canal therapy on tooth no. 15; however, recurrent infections occurred, and the tooth was eventually extracted, as well. The clinician took a bone biopsy specimen during the extraction. Microscopic examination of the hard tissue specimen revealed fibromyxoid connective tissue with dense viable bone (Figure 3). The bony trabeculae were laid down in both a woven and lamellar fashion. Only sparse osteoclasts were seen, and 1 focus of osteoblastic rimming was noted. The histomorphology combined with the clinical and radiographic features were compatible with segmental odontomaxillary dysplasia (SOD).

The patient had delayed healing at the extraction site, which was eventually resolved after diligent oral hygiene instruction and chlorhexidine mouth rinsing.

ABBREVIATION KEY

ADHD: Attention deficit/hyperactivity disorder.
F: Female.
L: Left.
M: Male.
NE: Not evident.
NS: Not specified.
R: Right.
SOD: Segmental odontomaxillary dysplasia.