

Clinical Case Report

Diagnostic challenge in segmental odontomaxillary dysplasia: a case report



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ABSTRACT

Segmented odontomaxillary dysplasia is a fibro-osseous lesion of unknown etiology characterized by segmental enlargement of the bone tissue of the hemimaxilla and surrounding soft tissues, and dental abnormalities on the affected side. The objective is to report clinical, radiographic, and microscopic features of a case of segmented odontomaxillary dysplasia. A 13-year-old male patient presented with mild facial asymmetry and a fissure in the left labial commissure. Physical examination revealed asymptomatic swelling in the left hemimaxilla and lack of dental eruption. Panoramic radiographs and cone-beam and fan-beam [IM1.1] computed tomography showed tooth retention and anomalies of 6 teeth, altered root canals, enlarged pulp chambers, and residual roots of malformed deciduous teeth. The adjacent bone showed expansion from the midline to the maxillary tuberosity, altered trabecular bone, and decreased volume of the nasal cavity and the left maxillary sinus. In addition to bone expansion, the patient had unique radiological findings not described in segmented odontomaxillary dysplasia: reduced nasal cavity and enlarged pericoronal follicle of the unerupted tooth. The patient underwent clinical investigation, followed by incisional biopsy and microscopic analysis for clinical correlation. Segmented odontomaxillary dysplasia is a rare disease, with poor established treatment protocols. Conservative treatments should be considered in young patients who have not yet undergone puberty. (Rev Port Estomatol Med Dent Cir Maxilofac. 2026;67(x):1-8)

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Introduction

Segmental Odontomaxillary Dysplasia (SOD) or hemimaxillofacial dysplasia is a rare disease, defined as a unilateral, non-hereditary, ectodermal dysplastic condition characterized by segmental enlargement of the maxilla and soft tissues,¹ with dento-osseous abnormalities and occasional cutaneous manifestations, including hypertrichosis, facial erythema, lip hypopigmentation, Becker's nevus, pigmented nevus, vermilion border discontinuity, and cheek depression.² It was described for the first time in 1990 by Danforth et al.³, after a series of eight reported cases that showed few alterations compared to cases of hemimaxillofacial dysplasia, recognized by Milhas et al.² in 1987. In 2022, it was classified as a fibro-osseous lesion by the World Health Organization (WHO).¹

SOD's etiology is still unknown. The literature has described genetic mutations in PIK3CA associated with dental alterations and in beta-actin (ACTB) associated with cutaneous alterations.⁴ Clinical changes begin between 5 and 27 years old, and the diagnosis usually occurs in the first decade of life.³ The overall prevalence of SOD is unknown due to its rarity, but most cases involve the right maxilla (55%),⁵ and it is slightly more prevalent in males (1:1.8).³

The main clinical feature of SOD is asymptomatic hemi-maxillary enlargement associated with abnormal bone maturation.⁴ Facial asymmetry may or may not be associated. Other manifestations may be present in the affected region, such as abnormal resorption of deciduous molars, hypodontia involving permanent premolars, reduced volume of the maxillary sinus, adjacent gingival alteration with or without skin lesions on the face, fissure in the labial commissure, and hypertrichosis.^{4,3,6,7}

Imaging studies of SOD have shown several alterations, including localized, unilateral enlargement of the maxilla, which may extend from the canine eminence to the tuberosity region, with associated dental abnormalities.⁸ Areas of coarse bone, sclerosis, and irregular and vertically oriented trabeculae, producing an ill-defined opacity and a ground-glass appearance, have been described. Small vertical radiolucencies, representing the most evident nutrient channels, have also been reported.^{9,10} Deciduous teeth in the affected region may be significantly abnormally resorbed and may be larger than those on the contralateral side.¹⁰ Teeth may be missing (hypodontia), often the premolars, or have abnormal or delayed root development, with changes in the size of the root canals or in the anatomy of the dental crowns, with enlargement of the pulp chambers. Because of these alterations, tooth displacement from the standard position,^{3,8,9} with delayed or absent eruption of the associated teeth, is common. Moreover, cone-beam computed tomography (CBCT) may reveal buccopalatal expansion of the affected region and a decrease in the size of the maxillary sinus on the affected side.¹⁰

SOD's histopathological features include thick bony trabeculae formed by irregular and immature bone, absence of lamellar bone, and osteoblastic/osteoclastic border. There is no cellular mitosis or pleomorphism.³ In soft tissues, nonspecific and non-inflammatory thickening of the connective tis-

sue and fibrosis of the gingival tissues are evidenced.¹¹ The teeth involved show microscopic alterations such as fibrous increase of the pulp in the deciduous teeth, irregular pulp-dentin interface, pseudoinclusions and pulp stones, irregular tubular dentin, and deficient odontoblastic layer.^{3,10,11}

Treatment of SOD is conservative, since the asymmetry it causes is usually mild. Contour osteotomy is not necessarily recommended, while orthodontic correction and prosthetic rehabilitation in edentulous areas are often indicated.¹²⁻¹⁴ When the diagnosis occurs in the first decade of life, the main objective is to maintain the deciduous teeth and postpone definitive treatment until the pubertal growth spurt.¹² Osseointegration and healing of dental implants have been reported successfully.^{9,13} The maxilla is the most affected bone, with right and left sides affected almost equally.¹² The prognosis is uncertain, due to the lack of published studies with long-term follow-up confirming whether it is a self-limited disorder.¹⁴ The objective of this study is to report a unique case of a young patient diagnosed with SOD with delayed tooth eruption and atypical radiological characteristics such as midline bone expansion, nasal cavity reduction, residual roots of malformed deciduous teeth, and enlargement of the pericoronal follicle of an unerupted tooth.

Case Report

The patient and his guardian signed an informed consent form authorizing the publication of the report.

A 13-year-old Caucasian male patient complained of painless swelling on the left maxilla and lack of eruption of permanent teeth. The patient brought several imaging tests (panoramic radiography, CBCT and fan beam and sinus tomography) and reported that other professionals had seen him previously, but no diagnosis had been established. In the anamnesis, the patient's mother reported that a dental surgeon had evaluated changes in the maxillary left primary molars 5 years earlier. His medical history had nothing noteworthy, and he denied the use of medications.

Extraoral physical examination revealed a slight facial asymmetry in the left middle third and a fissure in the labial commissure on the same side (Figure 1). Intraoral examination revealed swelling of the left maxillary ridge, which extended anteroposteriorly from the region of the left maxillary lateral incisor to the maxillary tuberosity, and from the vestibular region to the palate. The alveolar mucosa and gingiva covering the swelling were intact and showed no color changes. The deciduous maxillary left canine tooth had partially erupted, and the permanent left canine, premolar, and molar were missing. Only the distobuccal cusp of the left maxillary first molar was noticeable in the alveolar ridge (Figure 2).

Radiographically, the left maxillary canine, first and second premolars, and second molar were intraosseous and impacted in abnormal bone. The partially erupted left maxillary first molar was angulated, and the left maxillary third molar was developing, with 1/3 of the crown formed. All of them presented developmental anomalies characterized by changes in tooth shape and increased size of the pulp chamber and root canals when compared to those on the contralateral side. Radiography



Figure 1. A) Extraoral photograph focused on the lower third of the face, showing a fissure at the labial commissure. B) Extraoral photograph with focus on the intraoral region, showing, during smiling, the buccal corridor with mild asymmetry and missing teeth in the affected region.



Figure 2. The intraoral view shows enlargement of the alveolar ridge from the canine region to the maxillary tuberosity.



Figure 3. Panoramic radiograph demonstrates an ill-defined opacity in the left maxilla, with alterations in the pulp chambers and shape of the impacted teeth.

also depicted an alteration in the trabecular bone pattern, arranged more vertically in the left maxilla, extending from the canine eminence to the maxillary tuberosity (Figure 3).

CBCT demonstrated increased bone volume beginning slightly in the anterior midline region and gradually increasing to the posterior maxilla. In a sagittal view, the left maxillary canine showed an enlarged root canal and a palatal location, with its crown in close contact with the root of the left maxillary lateral incisor (Figure 4a). A coronal view revealed a pericoronal follicle slightly larger than normal and in close relation to the left maxillary deciduous canine, which also presented alteration of the pulp chamber and root canal and physiological external root resorption in its apical third (Figure 4b). Sagittal sections of the affected side, more laterally (Figures 4c and 4d), exhibited atypical anatomy of the left maxillary first and second premolars and first and second left maxillary molars, with enlarged pulp chambers and root canals, abnormal bone with evident vertical nutrient canals, and root

remains of malformed deciduous teeth located buccally to the maxillary second premolar. The Computed tomography (CT) and fan beam scan of the sinuses showed reduced ipsilateral nasal cavity and maxillary sinus when compared to the contralateral side (Figures 5a and 5b).

The panoramic X-ray performed 5 years earlier already showed poorly defined permanent teeth (from the left maxillary canine to the second molar) still in development, bone with a ground-glass appearance, and anomalies in the anatomy of the deciduous teeth on the same side (Figure 6).

The diagnostic hypothesis was SOD. The proposed treatment plan was extraction of the left maxillary deciduous canine associated with an incisional biopsy of soft tissue and bone for microscopic analysis. A small bone fragment was removed, respecting the proximity to permanent impacted teeth. Microscopic analysis of the gingival tissue demonstrated a stratified non-keratinized squamous epithelium with areas of hyperplasia, acanthosis, spongiosis, and hydropic

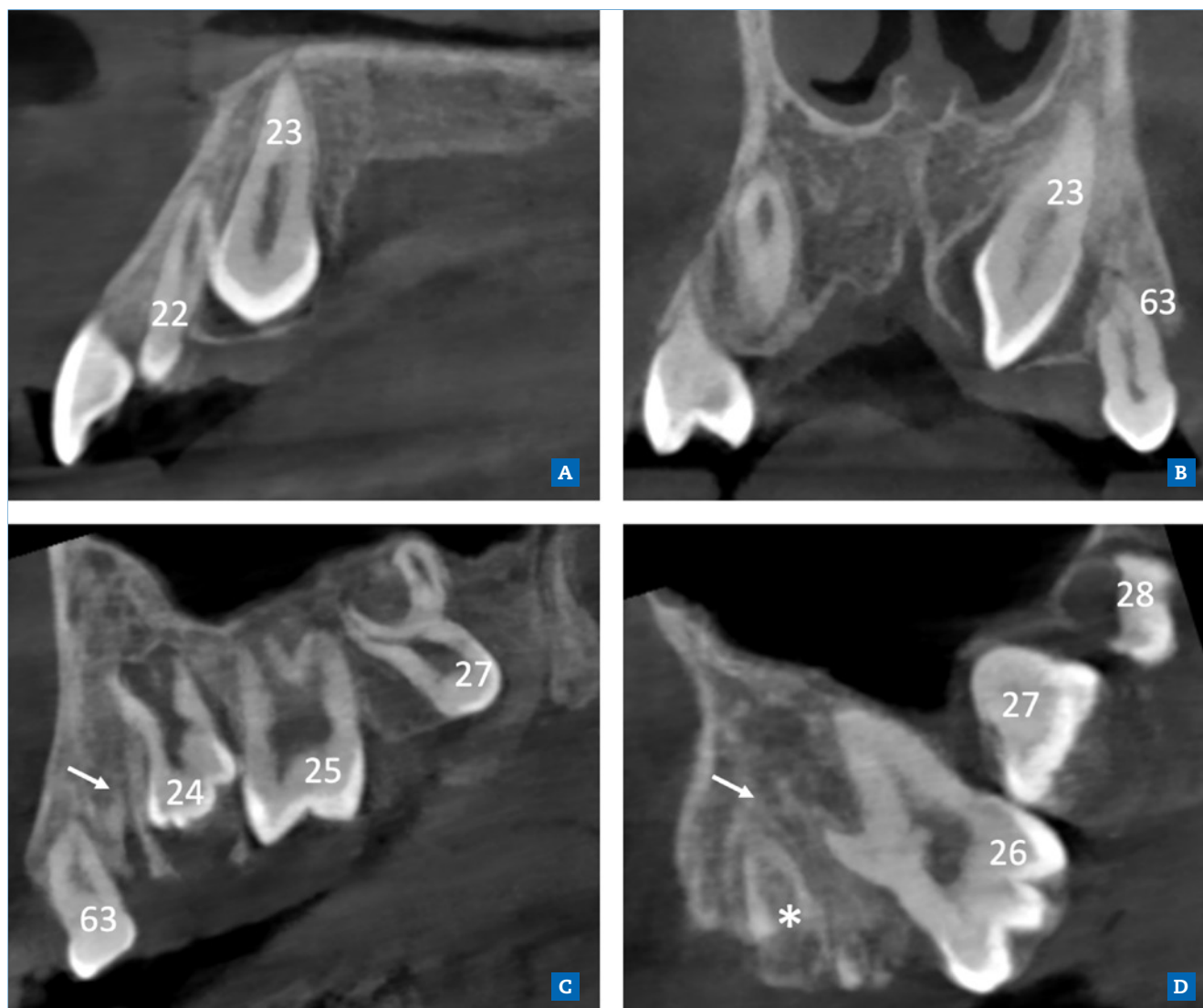


Figure 4. Cone-beam computed tomography (CBCT). A) Sagittal section showing the impacted left canine in proximity to the left maxillary lateral incisor. B) Coronal section showing enlargement of the pericoronal follicle of the left maxillary canine and partially external root resorption of the left maxillary deciduous canine. C) Sagittal section showing impacted atypical anatomy of the left maxillary first and second premolars and left maxillary first and second molars, and enlarged pulp chamber and root canals. The bone shows a frosted-glass appearance, and vertical nutrient canals (arrow) are noticeable. D) Lateral sagittal section showing partially erupted left maxillary first molar, left maxillary third molar in development, presence of vertical nutrient canal (arrow) (asterisk).

degeneration. Underneath, densely collagenized fibrous connective tissue was observed, exhibiting a discrete predominantly mononuclear inflammatory infiltrate in the subepithelial and perivascular regions, in addition to normal blood vessels. The bone tissue fragment analysis revealed irregular bone spicules exhibiting lamellar architecture and atrophic medullary spaces. Osteocytes were noted in their osteoplasts, with an absence of osteoblastic and osteoclastic activity (Figure 7a and 7b).

In view of the clinical and microscopic findings and imaging tests, the final diagnosis was SOD. The proposed treatment plan was orthodontic treatment, and the patient is being followed up, given that his pubertal growth spurt has not yet started.

Discussion and Conclusions

SOD is a rare disease. Although it was described more than 30 years ago, knowledge remains limited, especially regarding treatment protocols. A recently published paper reported that there are only 72 cases of SOD documented.¹⁵

The etiology of SOD is uncertain, and its occurrence is uncommon.^{12,16} Although it has been hypothesized to result from a localized abnormality in a developmental field or a postzygotic mutation or similar phenomenon in a progenitor cell, such alterations are thought to produce an altered cell clone that influences the morphogenesis of ectomesodermal tissues in a segment of the maxilla and overlying facial tissues.¹² Viral or bacterial infection of the branches of the max-

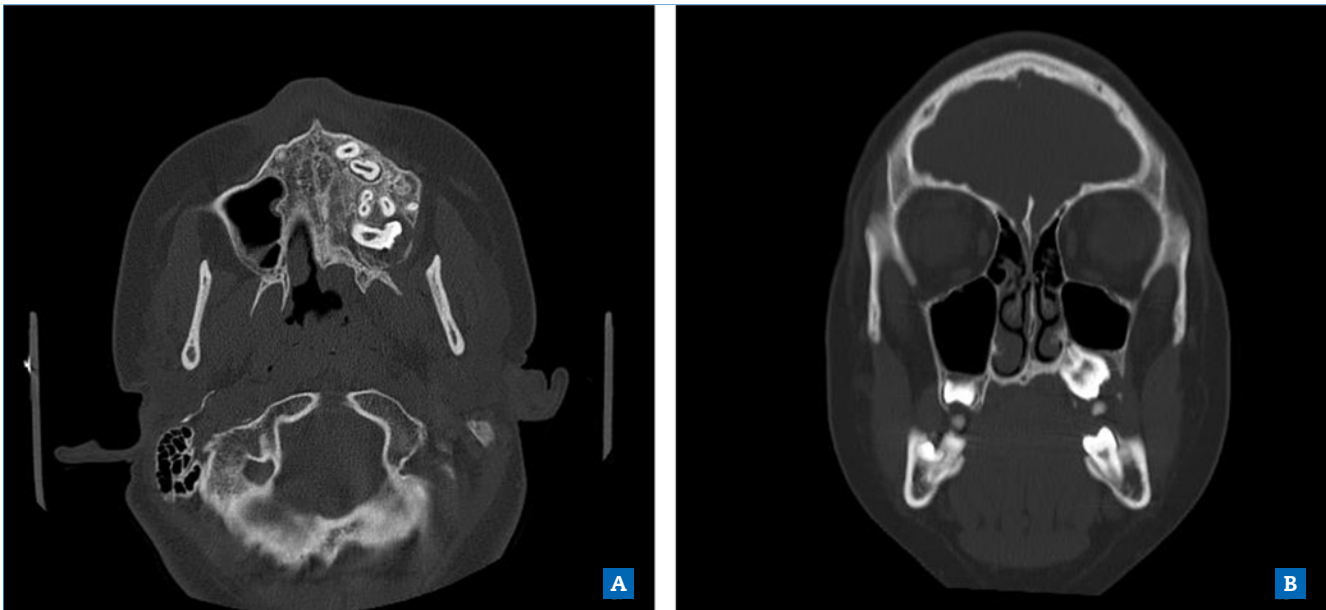


Figure 5. Axial section of sinus tomography. A) Evidenced bone expansion and dental/coronal malformation. B) Reduction in the nasal cavity volume and maxillary sinus on the affected side.

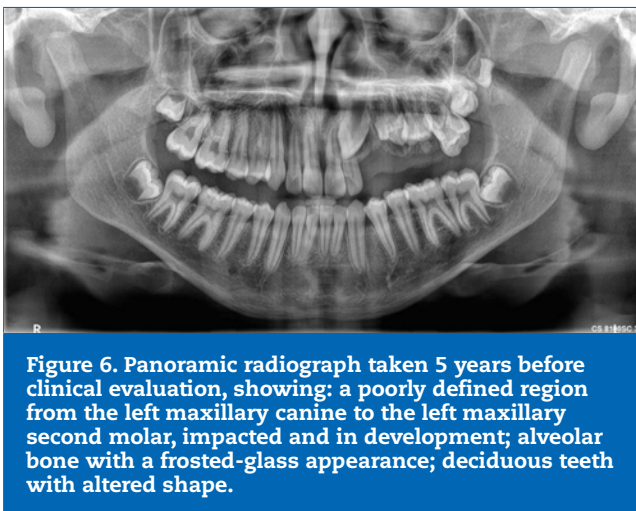


Figure 6. Panoramic radiograph taken 5 years before clinical evaluation, showing: a poorly defined region from the left maxillary canine to the left maxillary second molar, impacted and in development; alveolar bone with a frosted-glass appearance; deciduous teeth with altered shape.

illary division of the trigeminal nerve has also been proposed as an initial causal factor in SOD.^{12,17} Most reported cases seem to be a sporadic occurrence, and there is no hereditary pattern.^{12,18} This patient's medical history was not contributory, as previously reported. SOD is slightly more prevalent in males,^{5,15,19} and it does not seem to be associated with genetic inheritance.⁴

Clinical and imaging findings contribute to the diagnosis, and histopathological findings can be valuable. Although SOD is usually diagnosed in the first decade of life, diagnostic errors or even failure to recognize this abnormality can postpone diagnosis to adulthood.¹² Absent or noncontributory past medical history is reported in cases of SOD.

Whitt JC, Rokos JW, Dunlap CL, Barker BF., 2011¹⁰ classified SOD based on the presence and type of facial skin manifes-

tations. Type I (classic) features intraoral dentoalveolar and gingival manifestations without extraoral facial involvement, while Type II (cutaneous) exhibits combined dermatological, dentoalveolar, and gingival involvement. Type II is further subclassified into four subtypes: hypertrichotic (IIa), pigmentary (IIb), erythematous (IIc), and commissural defect (IId). Combinations between Type II subtypes are possible; for example, Type IIab with a hypertrichosis-pigmentary combination and Type IIabd with combinations of hypertrichotic, pigmentary, and commissural defect subtypes have been observed.¹⁰ In this case, no hypertrichosis or erythema was observed, so the cleft in the labial commissure indicated the classification as Type IId, according to Yassin and Rihani.¹¹ Cleft lip is a classic clinical sign of SOD.^{10,16} A discontinuity of the vermillion of the lip in the region of the commissure or fissure was first described by Packota et al.⁶ Skin changes may develop later, in adolescence or adulthood.¹⁹

On physical examination, six permanent maxillary teeth were missing, with only the distobuccal cusp of the left maxillary first molar visible in the left alveolar ridge in the buccopalatal direction. Regarding dentoalveolar and gingival alterations, the literature reports that delayed eruption is common, usually of the first molars,¹¹ in about 30% of the cases.⁵

Swelling of the left hemimaxilla was an important clinical finding associated with the patient's complaint. Unilateral enlargement of the maxilla, including gingiva, bone, or both, is a constant finding.¹⁹ Although this patient presented with swelling on the left side, a systematic review demonstrated a slight prevalence in the right maxilla, in 55% of the reported cases.⁵

It is noteworthy that the expansion in this case extended from the anterior region to the maxillary tuberosity, as seen either on clinical or radiographic examination. To the best of our knowledge, the literature reports only one case describing expansion from the anterior region up to the midline.¹²

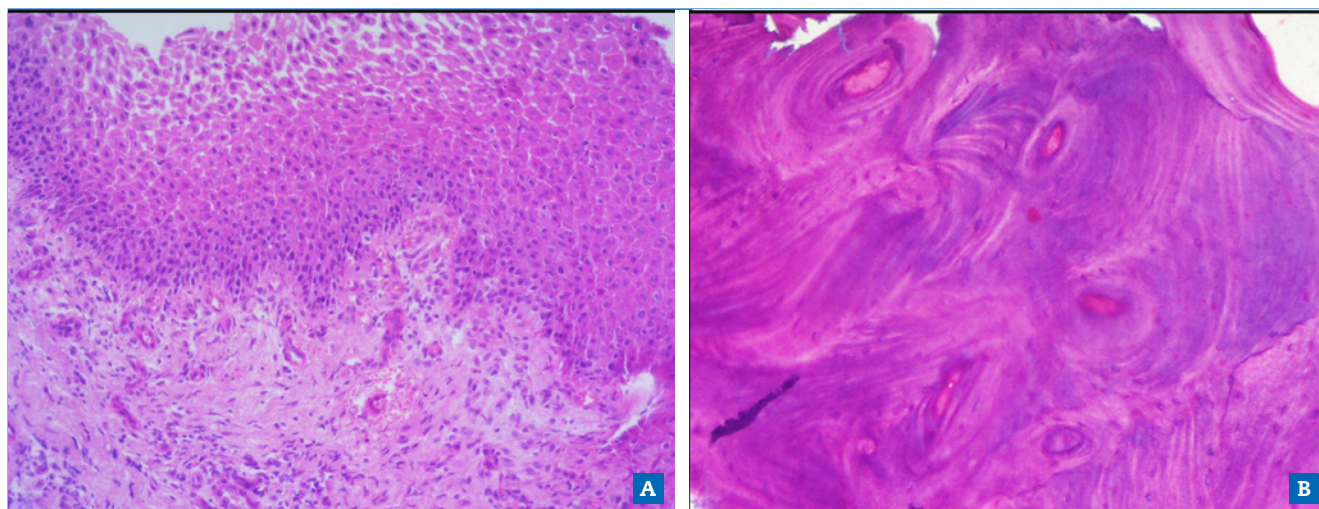


Figure 7. Histological section of gingival and bone tissue. A) Gingival tissue showing stratified non-keratinized squamous epithelium with epithelial hyperplasia and underlying fibrous connective tissue with mild mononuclear inflammatory infiltrate. B) Irregular bone spicules with lamellar architecture and atrophic marrow spaces. Hematoxylin and eosin staining; original magnification $\times 100$.

This case is unique because the imaging studies (panoramic radiography, CBCT, and CT) showed several bone and dental alterations. Regarding imaging characteristics, Packota et al.⁶ described parallel, vertically arranged, coarse trabeculae on periapical radiographs, a common finding of SOD. In the present case, the images showed poorly demarcated radiopaque bone trabeculae resembling ground glass, bone expansion, and reduced volume of the maxillary sinus and nasal cavity on the affected side. Moreover, dental changes were present, characterized by abnormal tooth development, enlargement of the pulp chamber and root canals, and changes in tooth anatomy, including the crowns, number of roots, and unerupted teeth. Dental anomalies as well as anomalies in the size of the maxillary sinus and nasal cavity are frequently reported^{5,12,13}. Hypodontia is common,⁶ especially of the permanent premolars,^{12,19} but it was not observed in this case. Residual roots of malformed deciduous teeth were observed in the affected region with alterations in dental anatomy and a ghostly appearance. The delay in the eruption of all permanent teeth was a striking feature, especially compared to the contralateral side. Delayed eruption is commonly observed in SOD.^{4,6}

The differential diagnosis includes disorders that cause volume increases in young patients. One example is fibrous dysplasia, due to the atypical bone pattern seen on radiographic examination, but this lesion does not cause alterations in tooth anatomy.¹⁵ Hemifacial hyperplasia causes swelling, but it affects the mandible,¹⁵ and early tooth eruption is reported.¹⁷ Regional odontodysplasia can be considered due to the absence of eruption and swelling in soft tissues, and there are no structural bone changes.¹⁷ These possibilities make the diagnosis challenging, because similar conditions can lead to an erroneous diagnosis, and small details are decisive in excluding diagnostic possibilities. Correlating clinical characteristics with imaging findings is essential for greater precision in the investigation.

In this case, a careful clinical evaluation was performed, and previous imaging exams were reviewed, making it possible to monitor disease progression. The patient underwent an incisional biopsy, which revealed densely collagenized fibrous connective tissue with slight signs of inflammation, consistent with the findings described in Shah's study.¹² In this case, collecting bone material was a challenge in the biopsy because of bone scarcity in the region caused by bone irregularity and a type IV bone, in addition to the proximity to the crowns of the unerupted teeth. The microscopic findings were not essential for the diagnostic conclusion, but the analysis detected the absence of osteoblastic and osteoclastic activity, resulting in inert bone.²¹ Although the microscopic findings are not specific in SOD, biopsy may be useful to rule out other diagnostic possibilities and to better guide the dentist in establishing the diagnosis. The diagnosis is based primarily on the correlation of clinical, dental, and radiographic findings.¹⁵

Treatment of SOD should be tailored to the patient's age, clinical presentation, and functional needs. While surgical interventions such as bone recontouring or dental implant placement may be considered in selected adult cases to address severe asymmetry or agenesis of teeth,^{15,17} conservative management is generally preferred, particularly in pediatric and adolescent patients.^{11,19} Conservative approaches, including orthodontic guidance of eruption and surgical exposure of teeth, were the treatment plan for this case, considering the patient's age. These approaches aim to facilitate eruption and alignment of the permanent dentition, thereby optimizing function and occlusion without invasive procedures.^{18,20}

The association of orthodontic treatment with prosthetic rehabilitation has been shown to effectively improve both function and aesthetics, particularly in patients with missing teeth or altered dental development.^{18,20} Importantly, facial asymmetry in SOD is typically non-progressive, and, for most patients, contour osteotomy is unnecessary. The primary therapeutic

objective is to support normal dental eruption and establish proper occlusion through minimally invasive interventions.^{19,21}

Longitudinal follow-up studies have highlighted the importance of ongoing clinical monitoring. Minett and Daley²⁰ documented a child with SOD followed from age 5 to 16, demonstrating stabilization of the craniofacial features by late adolescence. Similarly, Whitt et al.¹⁰ emphasized that long-term observation—from diagnosis through adolescence—is critical to guide treatment planning and anticipate future developmental outcomes. The present patient is under follow-up and undergoing orthodontic treatment. Regular clinical and radiographic monitoring are recommended every six months in the first year post-diagnosis and annually thereafter, to assess dentoalveolar development and determine the appropriate timing for interventions.^{15,19}

In the present case, because the patient was prepubertal and not showing active growth acceleration, a conservative strategy based on orthodontic treatment and close follow-up was adopted. This approach aligns with current evidence supporting early diagnosis, careful developmental monitoring, and individualized treatment timing as key factors in the successful management of SOD.

This case highlights clinical and radiographic features consistent with SOD, including some atypical radiological findings such as nasal cavity reduction and pericoronar follicle enlargement. The case reinforces the importance of individualized diagnosis and careful follow-up, particularly in young patients, where conservative management has been encouraged.

Conflict of interest

The authors have no conflicts of interest to declare.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed their work center protocols on access to patient data and for its publication.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

CREDIT AUTHORSHIP CONTRIBUTION STATEMENT

Jéssica Hálíce Noronha: Conceptualization, Investigation, Methodology, Writing – original draft, Writing – review & editing. **Luiz Henrique Padilha:** Investigation, Resources. **Thays Regina Ferreira da Costa:** Writing – review & editing. **Heliton Gustavo de Lima:** Data curation, Investigation, Validation, Writing – review & editing. **Melissa Rodrigues Araújo:** Conceptualization, Investigation, Supervision, Writing – review & editing.

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Desafio no diagnóstico da displasia odontomaxilar segmentar: relato de caso

R E S U M O

A displasia odontomaxilar segmentada é uma lesão fibro-óssea de etiologia desconhecida, caracterizada por aumento segmentar do tecido ósseo da hemimaxila e tecidos moles circundantes, além de anomalias dentárias no lado afetado. O objetivo é relatar as características clínicas, radiográficas e microscópicas de um caso de displasia odontomaxilar segmentada. Paciente do sexo masculino, 13 anos, com assimetria facial leve e fissura na comissura labial esquerda. O exame físico revelou edema assintomático na hemimaxila esquerda e falha na erupção dentária. Radiografias panorâmicas e tomografias computadorizadas de feixe cônico e fan-beam mostraram retenção dentária e anomalias em 6 dentes, canais radiculares alterados, câmaras pulpares aumentadas e raízes residuais de dentes decíduos malformados. O osso adjacente mostrou expansão da linha média até a tuberosidade da maxila, osso trabeculado alterado e volume reduzido da cavidade nasal e do seio maxilar esquerdo. Além do comprometimento da expansão óssea, o paciente apresentou achados radiológicos únicos, não descritos na displasia odontomaxilar segmentada: redução da cavidade nasal e alargamento do folículo pericoronário do dente incluso. O paciente foi submetido a investigação clínica, seguida de biópsia incisiva e análise microscópica para correlação clínica. A displasia odontomaxilar segmentada é uma doença rara, com protocolos de tratamento pouco estabelecidos. Tratamentos conservadores devem ser considerados em pacientes jovens que ainda não atingiram a puberdade. (Rev Port Estomatol Med Dent Cir Maxilofac. 2026;67(x):xxx-xxx)

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Palavras-chave:

Odontodisplasia

Radiologia

Anormalidades dentárias