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A dental approach to Proteus Syndrome: a case report

Abordagem odontológica na Síndrome de Proteus: relato de caso

Abordaje odontológico en el Síndrome de Proteus: reporte de caso

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Vilma Inutuka Pereira Rocha
ORCID: https://orcid.org/0000-0001-7199-0230
Centro Estadual de Reabilitação e Readaptação Dr. Henrique Santillo, Brasil
E-mail: vilmaiprocha@gmail.com
Bruna Yasmin Gomes
ORCID: https://orcid.org/0000-0002-1588-3008
Centro Estadual de Reabilitação e Readaptação Dr. Henrique Santillo, Brasil
E-mail: brunayasmin@egresso.ufg.br
Letícia de Paula Cauhi
ORCID: https://orcid.org/0000-0001-6139-5892
Centro Estadual de Reabilitação e Readaptação Dr. Henrique Santillo, Brasil
E-mail: leticiacauhi@gmail.com
Rosenelle Oliveira Araújo Benicio
ORCID: https://orcid.org/0000-0002-7465-8708
Centro Estadual de Reabilitação e Readaptação Dr. Henrique Santillo, Brasil
E-mail: rosenelle.araujo@gmail.com
Aida Sabbagh Haddad
ORCID: https://orcid.org/0000-0001-6769-8480
Universidade de São Paulo, Brasil
E-mail: aidasabbagh@hotmail.com

Abstract
Proteus syndrome (SP; OMIM #176920) is a rare complex disorder, characterized by the asymmetric postnatal growth of different parts of the body which tends to progress throughout life. Its prevalence is estimated at 1 per 1,000,000 live births. The clinical features are: partial gigantism of extremities, plantar hyperplasia, macrocephaly, cranial hyperostosis and asymmetry of the limbs due to the growth of long bones and soft tissues. Some of those affected may have intellectual impairment. Diagnosis is based on clinical criteria, imaging and genetic exams. The mainstay of treatment is early diagnosis of the disease and medication for symptoms presenting throughout life to improve quality of life. This case report presents a female patient, from 4½ years of age to the present, with asymmetric growth of the face and upper and lower limbs, undergoing dental follow-up at the Dental Service of a Specialist Center for Rehabilitation and Readaptation in Goiânia, Brazil. She presents delay in neuropsychomotor development, pronounced osteopenia, does not speak and is dependent in terms of activities of daily living. The eruption of deciduous teeth began at 4 months and permanent dentition at 4 years of age. During the current clinical examination of the child at 12 years and 2 months we found severe enamel hypoplasia in the teeth on the right side, mandibular prognathism, maxillary atresia, ogival palate, and anterior crossbite. The treatment plan included prevention and maintenance of oral health, monitoring of enamel hypoplasia, rhizolysis of the deciduous dentition and of eruption of permanent dentition, and restoration and extraction of teeth which were causing traumatic lesions on soft tissues. The parents' concerns were the malformations in dental enamel and traumatic lesions in the mucous membranes caused by badly positioned teeth. The interventions performed were composite resin restorations, tooth extractions, oral hygiene guidance and topical application of fluoride. Orthodontic treatment was contraindicated due to the severity of the disease. The dental approach in severe cases of PS must be individualized, with emphasis on oral hygiene and the removal of traumatic factors which damage soft tissues and cause pain. Dental surgeons must be prepared for individualized management.
Keywords: Congenital malformations; Craniofacial abnormalities; Hamartomatous syndrome; Proteus syndrome.

Resumo
A síndrome de Proteus (SP; OMIM #176920) é uma desordem rara, complexa, caracterizada por crescimento pós-natal assimétrico de diferentes partes do corpo que tendem a progredir por toda vida. A prevalência é estimada em 1 para cada 1.000.000 nascidos vivos. As características clínicas são: gigantismo parcial de extremidades, hiperplasia plantar, macrocefalia, hiperostose craniana e assimetria dos membros devido ao crescimento de ossos longos e partes moles. O comprometimento intelectual pode estar presente. O diagnóstico baseia-se em critérios clínicos, exames de imagem e teste genético. A base do tratamento inclui a identificação precoce da doença, medicamentos para sintomas apresentados com o decorrer da doença, buscando qualidade de vida. Apresentamos o caso de uma paciente do sexo feminino, desde os 4 anos e 6 meses de idade até a presente data, em acompanhamento odontológico em um Centro

Especializado em Reabilitação e Readaptação, na cidade de Goiânia-Go, no Centro-Oeste do Brasil com crescimento assimétrico da face, membros inferiores e superiores. Apresenta atraso no desenvolvimento neuropsicomotor, osteopenia acentuada, não fala e é dependente para as atividades da vida diária. A erupção dos dentes decíduos iniciou-se aos 4 meses de idade e a dentição permanente aos 4 anos. Ao exame clínico atual, com a criança aos 12 anos e 2 meses, observamos hipoplasia severa de esmalte nos dentes do lado direito, prognatismo mandibular, atresia maxilar, palato ogival, mordida cruzada anterior. O plano de tratamento consistiu na prevenção e manutenção da saúde bucal, acompanhamento das hipoplasias de esmalte, da rizólise da dentição decídua e erupção da dentição permanente, restauração e exodontia de dentes que causavam lesão traumática em tecidos moles. As preocupações dos pais eram as malformações no esmalte dental e as lesões traumáticas em mucosas provocadas por dentes mal posicionados. As intervenções realizadas foram restaurações em resina composta, extrações dentárias, orientação de higiene bucal e aplicação tópica de flúor. O tratamento ortodôntico foi contraindicado devido à gravidade da doença. A abordagem odontológica nos casos graves da SP deve ser individualizada com ênfase à higiene bucal e à remoção de fatores traumáticos que lesam tecidos moles e provocam dor. Cirurgiões-dentistas devem estar preparados para o manejo individualizado.
Palavras-chave: Anormalidades congênitas; Anormalidades craniofaciais; Síndrome Hamartomatosa; Síndrome de Proteus.

Resumen

El síndrome de Proteus (SP; OMIM #176920) es un trastorno raro y complejo caracterizado por un crecimiento posnatal asimétrico de diferentes partes del cuerpo que tiende a progresar a lo largo de la vida. La prevalencia se estima en 1 por 1.000.000 de nacidos vivos. Las características clínicas son: gigantismo parcial de las extremidades, hiperplasia plantar, macrocefalia, hiperostosis craneal y asimetría de las extremidades por crecimiento de huesos largos y tejidos blandos. El deterioro intelectual puede estar presente. El diagnóstico se basa en criterios clínicos, pruebas de imagen y pruebas genéticas. La base del tratamiento incluye la identificación temprana de la enfermedad, medicación para los síntomas presentados durante el curso de la enfermedad, buscando calidad de vida. Presentamos el caso de una paciente de sexo femenino, de 4 años y 6 meses de edad hasta la actualidad, en tratamiento odontológico en un Centro Especializado de Rehabilitación y Readaptación, en la ciudad de Goiânia-Go, en el Centro-Oeste de Brasil con asimetría crecimiento de la cara, miembros inferiores y superiores. Presenta retraso en el desarrollo neuropsicomotor, marcada osteopenia, no habla y es dependiente para las actividades de la vida diaria. La erupción de los dientes temporales comenzó a los 4 meses de edad y la dentición permanente a los 4 años. En el examen clínico actual, con el niño de 12 años y 2 meses, se observa hipoplasia severa del esmalte en los dientes del lado derecho, prognatismo mandibular, atresia maxilar, paladar alto, mordida cruzada anterior. El plan de tratamiento consistió en prevención y mantenimiento de la salud bucal, seguimiento de la hipoplasia del esmalte, rizólisis de la dentición temporal y erupción de la dentición permanente, restauración y extracción de las piezas dentales que ocasionaron lesión traumática de los tejidos blandos. Las preocupaciones de los padres eran malformaciones en el esmalte dental y lesiones traumáticas en las mucosas causadas por dientes mal posicionados. Las intervenciones realizadas fueron restauraciones de resina compuesta, extracciones dentales, orientación de higiene bucal y aplicación tópica de flúor. El tratamiento de ortodoncia estaba contraindicado debido a la gravedad de la enfermedad. El abordaje odontológico en casos severos de PS debe ser individualizado, con énfasis en la higiene bucal y la remoción de factores traumáticos que dañan los tejidos blandos y causan dolor. Los odontólogos deben estar preparados para un manejo individualizado.
Palabras clave: Anomalías congénitas; Anomalías craneofaciales; Síndrome hamartomatoso; Síndrome de Proteo.

1. Introdução

Proteus Syndrome (PS), Online Mendelian Inheritance in Man #176920, is a rare disorder with polymorphic phenotypic presentations which change over time (Cohen, 2014; Lindhurst et al., 2019). Although its first historical records date from the 19th century, it was originally described by Cohen and Hayden in 1979 (Cohen & Hayden, 1979). It was called PS due to its highly variable phenotypic presentation (Wiedemann et al., 1983). The name refers to the Greek god Proteus, because of its ability to change its shape, one of the main characteristics of the syndrome (Twede et al., 2005).

In 1884, Sir Frederick Traves, a British surgeon presented the case of the patient Joseph Merrick, known as the "Elephant Man" (Tibbles; Cohen, 1986). In later reviews, after analyzing the clinical and phenotypic presentations presented, it was concluded that the case met the criteria for PS (Satter, 2007).

It has an estimated incidence of 1 in every 1,000,000 live births and, as it is a sporadic condition, there are no records of family recurrence (Brockmann et al., 2008). Males are predominantly affected, in a proportion of 1.9:1 when compared to

females (Friedrich, 2021). A longitudinal study undertaken with 64 patients diagnosed with PS in São Paulo, showed that 25% had died by the age of 22 (Sapp et al. 2017). The main causes of death were pulmonary thromboembolism, pneumonia, neoplasias and respiratory failure (Cohen, 2001; Sapp et al. 2017).

PS is characterized by the asymmetric postnatal growth of different parts of the body, which usually starts between 6 and 18 months of age, due to a c.49G>A somatic mutation in the AKT1 gene, responsible for encoding a serine-threonine-kinase which leads to activation of the PI3K9 pathway and explains the mosaic pattern of the disease (Cohen, 2005; Biesecker, 2006). That same mutation has been described in various solid tumors and, when germline, is considered incompatible with life. Molecular studies facilitate diagnosis of the syndrome when clinical criteria are inconclusive and should preferably be undertaken in a tissue sample extracted from the affected region, unlike genetic syndromes due to germline mutations, in which the genetic variants causing the condition can be found in a blood or saliva sample (Lindhurst et al., 2011).

The main clinical features of PS are: partial gigantism of the extremities, cerebriform connective nevi, plantar hyperplasia, hemangiomas, lipomas, lymphangiomas, varicosities, linear verrucous epidermal nevus, macrocephaly, cranial hyperostosis and asymmetry of the limbs due to the growth of long bones and soft tissues (Cohen & Hayden, 1979). Other characteristics described in patients include hemimegalencephaly (one of the few congenital alterations associated with the syndrome), specific tumors (bilateral ovarian cystadenoma, monomorphic adenoma of the parotid glands), scoliosis, asymmetric and disproportional growth of the viscera (including spleen and thymus), neuropsychomotor delay and craniofacial dysmorphia (Biesecker & Sapp, 2012; Jamis-Dow et al., 2004; Turner et al. 2004).

In terms of craniofacial abnormalities, the literature points to: tooth agenesis, impacted teeth, ectopic eruption, malocclusion, eruption and heterogeneous maturation, midline displacement, bone exostoses, unilateral macroglossia, degenerative alteration in the temporomandibular joint, enlargement of the mandibular canal and foramen, alteration in maxillary and mandibular vertical and horizontal growth (Munhoz et al., 2021) and enamel hypoplasia (Mason & Roberts, 2009; Valéria et al., 2015).

As overgrowth is a permanent problem, its management is complex. Skeletal overgrowth may occur with functional impairment; hence, the assistance provided by physiatry/rehabilitative medicine, including physical and occupational therapy, is considered paramount. Because of the difference between the length of the legs and the feet, custom shoes or orthotics are also valuable. In terms of scoliosis, corrective surgery is recommended due to the potentially fatal risk of restrictive lung disease. Anticoagulation should be adopted, and special care taken due to the increased risk of thromboembolic events, especially that with potential risk of death from deep vein thrombosis (DVT) and pulmonary thromboembolism (PTE) (Biesecker, 2006; Jamis-Dow et al., 2004).

The clinical diagnosis of PS is based on characteristic findings supplemented by additional specific clinical criteria (Panteliadis; Friedrich, 2022). Therefore, patients with PS benefit from multidisciplinary and multi-professional therapeutic approaches, adapted to the specific needs and manifestations of each individual. Accompaniment should involve a multi-professional team. It should be mentioned that such therapeutic approaches aim to avoid clinical complications, which allows for improved social inclusion (Lougaris et al., 2016).

Consequently, this article set out to report the clinical findings of a patient with PS, evaluating their physical aspects, general changes, as well as oral findings, and report the multi-professional clinical management with emphasis on the dental procedures undertaken, showing the need for individualized preventive, restorative and therapeutic treatment in order to provide quality of life for the patient.

2. Methodology

It is a case report study in which a description and subsequent discussion of a case of scientific interest and for knowledge is presented (Estrela, 2018). In the present study, a report was made of a child with Proteus Syndrome treated at the dental service of a referral hospital for rehabilitation and rehabilitation in the city of Goiânia, Goiás, Brazil.

Prior to data collection, this work was sent to the Research Ethics Committee (CEP) Leide das Neves Ferreira of the Goiás State Health Department and was approved on 12/16/2021 with number 5,170,738. Those responsible for the child read and signed the free and informed consent form, agreeing to carry out this case report.

3. Case Report

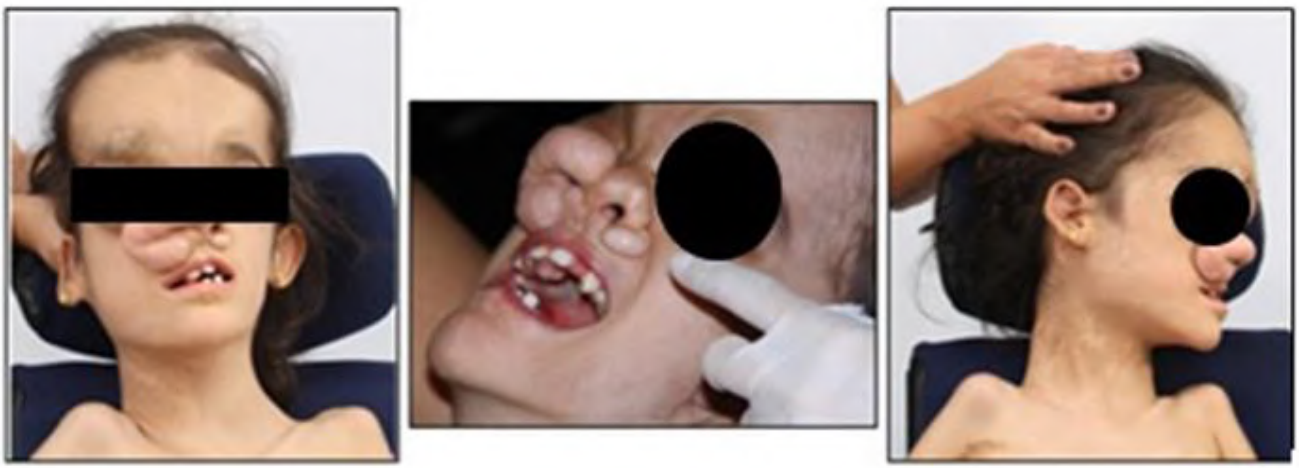
A 4½ year old female with leukoderma presented at the Dental Service of a Center for Rehabilitation and Readaptation, in Goiânia, in Brazil's Midwest, having been referred by a geneticist who works at the hospital where the Dentistry Division is located. Her mother complained about the position of the mandibular tooth which was causing trauma to the maxillary gingival tissue and her difficulty in performing oral hygiene.

The patient is the first child of a healthy non-consanguineous couple who have another healthy child. Despite the mother planning a normal delivery, the child was born by cesarian section at the 39th week of pregnancy, on medical advice, due to the increased head circumference. At birth, the child weighed 3,200 grams (between the 25th and 50th percentiles for sex and gestational age), measured 53.5 cm (between the 90th and 97th percentiles for sex and gestational age), and had an Apgar score of 6 in the first minute and 9 in the fifth minute. At birth, macrocrania (a head circumference of 39 centimeters above the 97th percentile for sex and gestational age), asymmetric growth of the lower limbs (a characteristic which was confirmed by the plantar impression of both feet made in the early hours of life) and ala of the nose were seen. Since birth, craniofacial and trunk impairments have been observed. However, around the age of 6 years, the growth of the right nasal ala and ocular proptosis were accentuated. The child has marked neuropsychomotor developmental delay (NPMD), does not speak and is dependent for activities of daily living, although she is able to express joy and discontent.

She presents a stable primary neoplasm in the pineal region, controlled epilepsy, pronounced osteopenia, myopia, and hearing loss on the right side. Epileptic seizures started when she was around 9 months and are currently under control. The child is on continuous use of 60 mg Oxcarbazepine (9 ml 2x/day), 250mg Levetiracetam (4 ml 2x/day) and 10 mg Clobazam (1/2 tablet 2x/day) in adequate doses. She has been on a gastrostomy tube diet (G-tube) since she was 5 years old. At 8 years and 7 months, she started compassionate and experimental use of an AKT1 inhibitor, initially developed for cancer treatment and which is under investigation for use in PS. She was hospitalized for periods with PTE at 9 years and 11 months.

The general physical examination, performed when she was 6 years of age, described by the geneticist at the hospital, showed: hypoplasia of the shoulder girdle, leg length discrepancy with a marked increase in the right foot, bilateral macrodactyly, and marked dextroconvex thoracolumbar scoliosis. She also presented asymmetry of the ear pinna, asymmetric macrocrania with bulging of the forehead to the right, drooping palpebral fissures, ocular hypertelorism, bilateral ocular proptosis more pronounced on the right, where there was also associated chemosis, tumors on the right nasal ala (circumferential and large) and left nasal ala (smaller and below the nasal ala) (Figure 1).

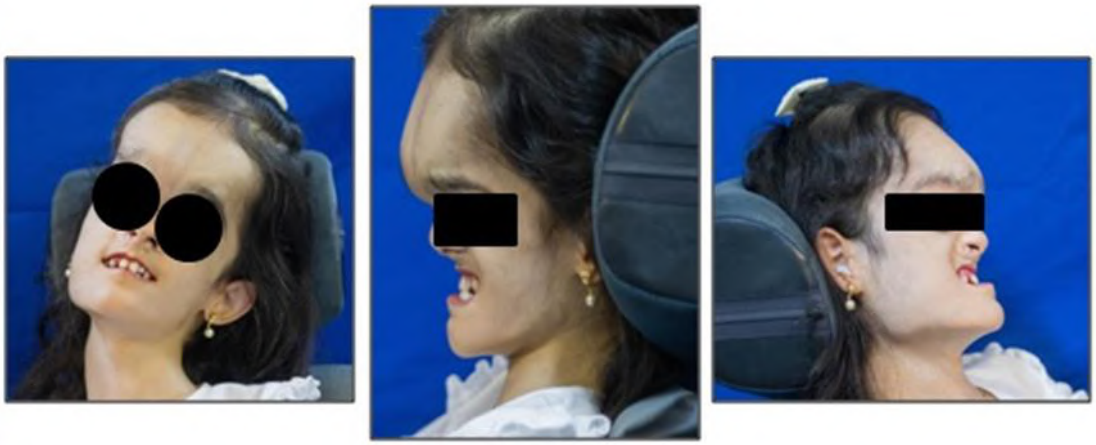
Figure 1. Sequence of images of the 7-year-old patient with Proteus Syndrome, front view and left and right profiles.



Source: Patient's chart.

At the age of 7, the patient underwent plastic surgery on the nasal ala at another hospital to remove the tumors and reduce proptosis of the eyeball. These results can be seen in the patient at 12 years and 2 months (Figure 2).

Figure 2. Sequence of images of the patient with Proteus Syndrome aged 12 years and 2 months, showing front view, and left and right profiles after plastic surgery.



Source: Patient's chart.

Eruption of the deciduous teeth started at 4 months and permanent teeth at 4 years old. During an intraoral examination at the age of four the child presented with mixed dentition with these deciduous teeth: right upper canine (53), right upper first molar (54), right upper second molar (55), upper left lateral incisor (62), upper left canine (63), upper left first molar (64), upper left second molar (65), lower left central incisor (71), lower left lateral incisor (72), lower left canine (73), lower left first molar (74), lower left second molar (75), lower right canine (83), lower right first molar (84), and lower right second molar (85). The following permanent teeth were observed: right upper central incisor (11), right upper first molar (16), left lower first molar (36), right lower central incisor (41) and right lower first molar (46). There was no lower left lateral incisor. According to the mother's report the child had that tooth removed at another health center due to trauma to the maxillary mucosa. Hypotonic lips and tongue and a high-arched palate were also seen.

4. Dental treatment

The dental therapeutic plan consisted of prevention and maintenance of oral health, with monitoring of the rhizolysis of the primary dentition, eruption of the permanent dentition, enamel hypoplasia, extraction of the teeth traumatizing soft tissues, and oral hygiene. For the clinical approach, the tell-show-do technique and music therapy were used. As the patient is not collaborative when receiving dental care, the service was performed, when possible, in the patient's own wheelchair. On occasions when supragingival scaling and restorations were required, the treatment was undertaken in the dental chair, involving the dentist and two oral health care assistants, with written consent, in short sessions of a maximum of 30 minutes. In both situations, the child's head had to be stabilized due to involuntary movements. However, protective stabilization (bands of fabric) was not used due to the child's bone fragility and abdominal distension. Aspiration was repeatedly performed, with two aspirators, and a silicone monobloc attached to dental floss was used to maintain the mouth opening.

On certain occasions, assistance was given in the dental chair, but with the mother sitting in the chair supporting the child's lower limbs on her legs, and physically restraining the child's upper limbs with her hands. The chair was set at an angle of about 30° in order to prevent bronchoaspiration.

All the procedures in the dental treatment plan were previously discussed with those responsible for the patient. The risks and benefits of the treatment were presented and, only after signing the Free and Informed Consent Term (FICT), was the dental treatment performed as described below:

1) Extraction of the lower right central incisor (41) and deciduous lower right first molar (84) at 4 years and 7 months in a single session. Tooth 41 was extracted due to traumatic injury to the maxilla and rhizolysis, respectively.

2) Restoration using nano-hybrid composite resin (Tetric/Ivoclar) in the upper right central incisor (11) at 4 years and 9 months, with relative isolation of cotton rolls and continuous aspiration. Reason: enamel hypoplasia. Her mother complained of the darkened appearance of the tooth resembling a caries.

3) Extraction of the following deciduous teeth: upper right canine (53), lower left canine (73), lower right canine (83) at 4 years and 11 months in a single session. Reason: complete rhizolysis.

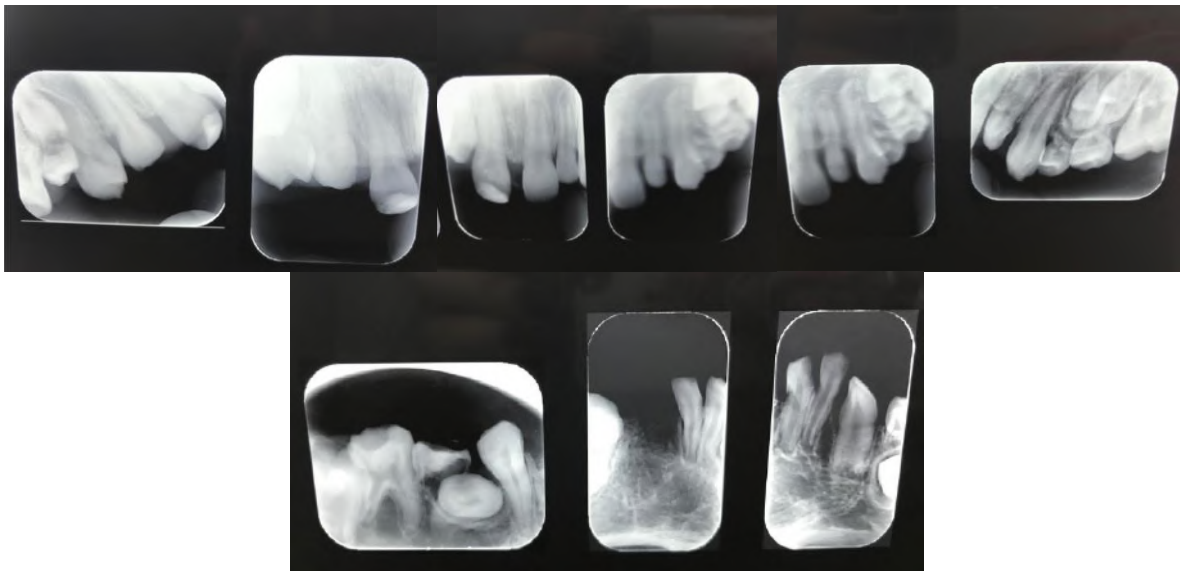
4) Extraction of the following deciduous teeth: upper left lateral incisor (62), upper left canine (63) at 5 years and 6 months. Reason: complete rhizolysis.

5) Extraction of the following teeth: lower right second premolar (45), lower right second molar (85) at 6 years and 4 months. Reason: ectopic eruption and complete rhizolysis, respectively (Figure 3 - periapical X-ray).

6) Extraction of the lower left second molar (75) at 7 years and 2 months. Reason: complete rhizolysis.

7) Correction using nano-hybrid composite resin (Tetric/Ivoclar) of restoration fractured by trauma, in the upper right central incisor (11) at age 11, resulting from intubation in the operating room during surgery to remove a benign tumor in the right ear.

Figure 3. Periapical radiographs of the patient with Proteus Syndrome at 6 years and 5 months.

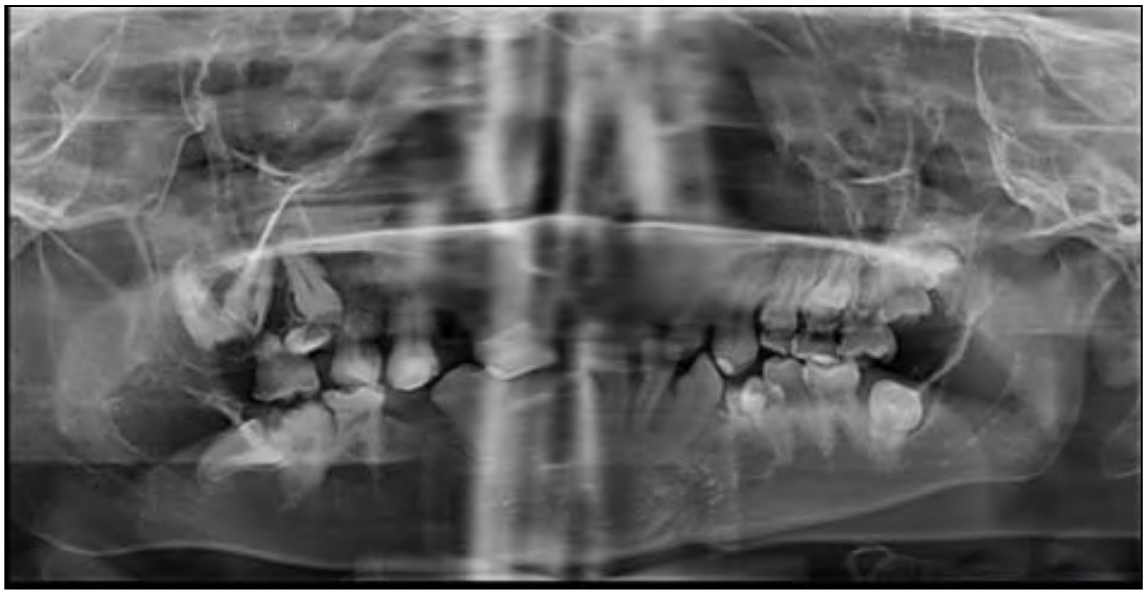


Source: Patient's chart.

In the current clinical examination with the child now 12 years and 2 months, the following conditions were noted: severe enamel hypoplasia in the teeth on the right side, mandibular prognathism, maxillary atresia, anterior crossbite, gingival proliferation located in the incisive canal and right upper molar regions. It was not a generalized gingival hyperplasia caused by drugs.

Analysis of the current panoramic radiograph showed advanced dental and bone development in the right dental hemiarch. The deciduous teeth still present were the upper right molars (54 and 55) with rhizolysis; upper left molars (64 and 65) with rhizolysis and lower left first molar with rhizolysis and in infra-occlusion (74). With regard to permanent teeth, agenesis of the upper right lateral incisor (12), lower right first premolar (44), and lower left third molar (38) was seen. Missing teeth: lower right central incisor (41) and lower right lateral incisor (42). There was a supernumerary calcifying tooth near the crown of the upper right second premolar (15). Smooth apical root resorption was also seen in the upper right central incisor (11) and in the upper right first premolar (14). Erupting upper right and left third molars (18 and 28), restorations in the upper right and left central incisors (11 and 21) and right maxillary canines (13) were also seen. All other permanent teeth are located intra alveolar, upper right second premolar (15), upper right second molar (17), upper left first premolar (24), upper left second premolar (25), upper left second molar (27), lower left second molar (37), lower right second molar (47) and lower right third molar (48) (Figure 4).

Figure 4. Panoramic radiograph of the patient with Proteus Syndrome aged 12 years and 2 months, with distortion due to positioning difficulties during image acquisition.



Source: Patient's chart.

In all 30 preventive return visits, no caries were found. However oral hygiene was unsatisfactory and there was gingivitis. Her mother experienced difficulty in carrying out oral hygiene, especially on the lingual part of all lower teeth and the buccal part of the upper molars. Supragingival periodontal scaling procedures were performed with McCall 13/14, 17/18 and morse tip 0-00, prophylaxis with a Robson brush and fluor therapy with 5% Sodium Fluoride Varnish (Duraphat/Colgate). The parents were instructed on the technique of performing oral hygiene, brush size and dental floss rod. The child has a portable secretion aspirator at home.

Assistance for this patient was possible due to the parents' collaboration and trust in the treatment proposals presented by the dental surgeons. The mainstay of treatment for PS includes early identification of serious medical problems and use of prophylactic and symptomatic treatment. Thus, in addition to the high phenotypic variability of the syndrome, changes have to be made to the approach to dental treatment in order to individualize each case treated, according to its peculiarities.

5. Discussion

PS is an extremely rare disease characterized by excessive growth and with distribution in a mosaic of multiple tissues. Due to its rarity and high phenotypic variability, it has been difficult to establish sensitive and specific diagnostic criteria (Cohen; Hayden, 1979; Cohen, 2001; Cohen, 1993).

Even though most cases are associated with changes in the growth pattern begun after the first months of life, the literature describes hemimegalencephaly from the intrauterine period, with could be compatible with the macrocrania identified in an ultrasonography carried out during the prenatal period (Biesecker, 2006; Biesecker; Sapp, 2019; Cohen, 2005). The delayed neuropsychomotor development identified in the patient in this study is also in accordance with the clinical descriptions, although some patients can present normal neuropsychomotor development (Biesecker et al., 1999; Jamis-Dow et al., 2004; Wiedemann et al., 1983).

The main differential diagnoses of PS involve syndromes associated with genes which act in the same oncogenic pathway as PI3K (in particular, PTEN and PI3KCA). In the present case, the patient met all the general criteria (mosaic

distribution, progressive evolution and sporadic occurrence), as well as the following specific criteria: asymmetric and disproportionate growth of limbs, visceral and organ hypergrowth, facial phenotype with dolichocephaly, drooping eyelids, anteverted nostrils, thromboembolic events and lipodystrophy, totaling 16 points, sufficient for diagnostic definition in patients without molecular confirmation (Keppler-Noreuil et al., 2019; Lindhurst et al., 2019; Wiedemann et al., 1983).

A facial phenotype was previously described, characterized by dolichocephaly, long face, slightly drooping palpebral fissures, mild ptosis, low nasal bridge, wide or anteverted nostrils, and open mouth at rest. This maxillofacial phenotype, previously associated with PS (Becktor et al., 2002), is present in this case, as well as intraoral manifestations, such as tooth agenesis on the affected side. As previously shown, some germs present early tooth eruption and maturation (Lindhurst et al., 2011).

However, contrary to the description above (Canabarro et al., 2008), enamel hypoplasia affected the teeth bilaterally and the maxillary central incisors present vertical hypoplasia (Mason; Roberts, 2009). The patient does not present lingual gingival hyperplasia and in the radiographic findings, early dental age on the affected side, enlargement of the mandibular canal and mandibular foramen, could be seen (Jamis-Dow et al., 2004; Korbmacher et al., 2005). In the craniofacial findings, the increased head circumference at birth, of 39 centimeters, was one of the characteristics which led to the early investigation, as well as the asymmetry of the lower limbs.

Spinal deformations and enlargement of one or more vertebrae are present in the skeletal phenotype of PS (Kaiser et al. 2015). The literature points out that orthopedic surgeries may be necessary when spinal compressions lead to functional complications (Tosi et al., 2011). In this case, despite the involvement of the spine, the child is able to remain seated in a wheelchair and there was no indication for surgery.

Once a diagnosis is made, the treatment and follow-up must be carried out by a multi-professional and interdisciplinary team, with an on-going focus on maintaining quality of life and preserving health. Psychological aspects of patients and family members must be considered, as it is an uncommon condition which leads to several comorbidities which are accompanied by serious deformities and limitations in daily life. Patients are at increased risk of premature death, usually caused by deep vein thrombosis (DVT), PTE and pneumonia (Sapp et al., 2019).

Therefore, dental surgeons' awareness of this condition can contribute to better diagnosis of the oral conditions and planning and dental treatment in light of the particularities of Proteus Syndrome in each individual, with a view to offering a better quality of life.

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