Research, Society and Development, v. 12, n. 4, e23912441211, 2023
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Complexity of dental management of a pediatric patient with lamellar ichthyosis: case report

Complexidade do manejo odontológico de um paciente pediátrico com ictiose lamelar: relato de caso

Complejidad del manejo dental de un paciente pediátrico con ictiosis lamelar: reporte de caso

Received: 03/27/2023 | Revised: 04/10/2023 | Accepted: 04/11/2023 | Published: 04/16/2023

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Abstract
The purpose of this case report is to describe the complexity of dental management of a pediatric patient with lamellar ichthyosis. A 9-year-old male patient was referred by the pediatrician to the special needs' clinic with a major complaint of "tooth in the roof of the mouth", according to the person in charge. The patient's mother reported that he was diagnosed with lamellar ichthyosis at birth and regularly followed up with a medical team, including a cardiologist, due to the presence of mild pulmonary valve stenosis. The physical examination revealed presence of skin peeling, everted lip, mouth breathing, cognitive deficit, and speech delay. The intraoral clinical examination showed mixed dentition, absence of caries lesions and presence of supernumerary teeth on the palate, between teeth 11 and 21, which was confirmed by radiographic examination. Although he was cleared by the pediatrician and cardiologist for the procedure under general anesthesia, this procedure in the hospital setting was interrupted at the last minute and contraindicated by the anesthesiologist, as it would be a high risky due to the patient's airway conditions. Thus, one month later, the patient's surgery was performed under local anesthesia on an outpatient basis under sedation and protective stabilization. The patient remains on follow-up and receiving preventive treatment. Thus, it is fundamental that there is a multidisciplinary approach of these patients to prevent possible complications resulting from dental procedures. In addition, regular dental monitoring is essential to avoid subjecting the patient to invasive procedures.
Keywords: Ichthyosis, lamellar; Tooth, supernumerary; Pediatric dentistry.

Resumo
O objetivo deste relato de caso é descrever a complexidade do manejo odontológico de um paciente pediátrico com ictiose lamelar. Paciente do sexo masculino, 9 anos, foi encaminhado pelo pediatra ao ambulatório de necessidades especiais com queixa principal de "dente no céu da boca", segundo o responsável. A mãe do paciente relatou que ele foi diagnosticado com ictiose lamelar ao nascimento e acompanhado regularmente pela equipe médica, incluindo um cardiologista, devido a presença de estenose leve da válvula pulmonar. O exame físico revelou presença de

descamação cutânea, lábio evertido, respiração bucal, déficit cognitivo e atraso na fala. O exame clínico intraoral mostrou dentição mista, ausência de lesões de cárie e presença de dentes supranumerários no palato, entre os dentes 11 e 21, o que foi confirmado por exame radiográfico. Embora autorizado para o procedimento de anestesia geral pela pediatria e cardiologista, após o risco cirúrgico, no momento deste procedimento em ambiente hospitalar, o mesmo foi interrompido de última hora e contraindicado pelo anestesista de plantão devido ao elevado risco de vida em função das condições das vias aéreas do paciente. Assim, um mês depois, a cirurgia do paciente foi realizada sob sedação e estabilização protetora, com anestesia local em ambulatório. O paciente permanece em acompanhamento e recebendo tratamento preventivo. Assim, é fundamental que haja uma abordagem multidisciplinar desses pacientes para prevenir possíveis complicações decorrentes de procedimentos odontológicos. Além disso, o acompanhamento odontológico regular é essencial para evitar submeter o paciente a procedimentos invasivos.
Palavras-chave: Ictiose lamelar; Dente supranumerário; Odontopediatria.

Resumen
El presente reporte de caso tiene como objetivo describir la complejidad del manejo dental de un paciente pediátrico con ictiosis lamelar. Un paciente masculino de 9 años de edad fue derivado por el pediatra a la consulta externa de necesidades especiales con una queja principal de "muela en el techo de la boca", según el responsable. La madre del paciente informó que fue diagnosticado con ictiosis lamelar al nacer y fue seguido regularmente por el equipo médico incluido un cardiólogo, debido a la presencia de estenosis valvular pulmonar leve. Al examen físico destacaba la presencia de descamación de la piel, labio evertido, respiración bucal, déficit cognitivo y retraso del habla. El examen clínico intraoral mostró dentición mixta, ausencia de lesiones cariosas y presencia de dientes supernumerarios en el paladar, entre los dientes 11 y 21, lo cual fue confirmado por examen radiográfico Aunque autorizado para el procedimiento de anestesia general por el pediatra y el cardiólogo, luego del riesgo quirúrgico, al momento de realizar este procedimiento en un ambiente hospitalario, fue interrumpido a última hora y contraindicado por el anestesista de turno por el alto riesgo de vida debido a las condiciones de las vías respiratorias del paciente. Así, un mes después, el paciente fue intervenido bajo sedación y estabilización protectora, con anestesia local de forma ambulatoria.. El paciente permanece en seguimiento y recibiendo tratamiento preventivo. Por lo tanto, es fundamental que exista un abordaje multidisciplinario de estos pacientes para prevenir posibles complicaciones derivadas de los procedimientos dentales. Además, el seguimiento odontológico periódico es fundamental para evitar someter al paciente a procedimientos invasivos.
Palabras clave: Ictiosis lamelar; Diente supernumerario; Odontología pediátrica.

1. Introduction

Ichthyosis is a group of keratinization disorders that is clinically and etiologically heterogeneous and characterized by dryness, flaking, and peeling of the skin (Gulasi et al., 2016), with a multitude of complications in severe variants (Neema et al., 2015). These manifestations are due to mutations in genes mostly involved in skin barrier formation (Takeichi et al., 2016), with TGM1 (transglutaminase 1) being one of the major (Ramar et al., 2014). There are a wide range of ichthyosis phenotypes, including harlequin ichthyosis, lamellar ichthyosis (LI), congenital ichthyosiform erythroderma (CIE), and self-improving collodion ichthyosis (SICI) (Oji et al., 2009; Akiyama et al., 2011).

Lamellar ichthyosis is the rarest form (Pranitha et al., 2014) with estimated incidence of 1:200,000 ± 300,000 (Yang et al., 2001) and has an equal incidence in men and women (Fitzpatrick et al., 1999). Cicatricial ectropion, the most common eyelid abnormality in LI, decreases the ocular surface defense mechanism and leads to corneal exposure and ulceration (Hosal et al., 1999). The condition usually presents at birth and the baby is found to be unsheathed in a membrane (Lefèvre et al., 2006; Fleckman et al., 2008). In addition, xerostomia may also be an important oral manifestation of this disease, which can consequently increase the risk of caries in these patients (Ramar et al., 2014). Other manifestations may include abnormal deciduous and permanent teeth (Pranitha et al., 2014).

The symptoms vary from the mildest types such as ichthyosis vulgaris, which may be mistaken for normal dry skin, up to life-threatening conditions such as harlequin ichthyosis (Ramar et al.,2014). However, little is known about dental treatment and its possible limitations and there is a considerably low number of studies and dental case reports on patients with lamellar ichthyosis (Matsuno et al., 2014; Reis et al., 2022), mainly studies that report the possible complexity of a surgical procedure in a patient with this condition.

In this context, the aim of the present paper was to report the management and dental care of a pediatric patient with lamellar ichthyosis, to perform a good care and avoid possible risks and complications.

2. Methodology

This is descriptive report, of qualitative nature, being characterized as a case report (Pereira et al., 2018), that addresses the difficulty and ideal care when setting up a treatment plan for a patient with lamellar ichthyosis. The single patient reported in this case needed a surgery to remove a supernumerary tooth; the proposed plan was to perform the surgery in a hospital environment, under general anesthesia, but the difficulty in manipulating the patient's airways made surgery impossible and it was necessary to decide on a new treatment plan. The remaining alternative was oral sedation associated with protective stabilization, performed in an outpatient setting. After the execution, the patient was in regular follow-up and preventive treatment. Regarding ethical aspects, this study followed the ethical principles of the Declaration of Helsinki of Resolution nº 466/2012 of the National Health Council (NHC) and the person responsible for the patient signed the Informed Consent Form.

3. Case Report

A nine-year-old boy presented with his mother at the Special Dentistry Clinic of the Universidade Federal do Rio de Janeiro, Rio de Janeiro, Brazil, referred by the pediatrician. A major complaint, reported by the mother was "tooth in the roof of the mouth".

During the anamnesis, the mother reported that the patient had been diagnosed with Ichthyosis Lamellar at birth, without complications during pregnancy and absence of systemic involvement among those responsible. In addition, the patient has an older sibling, who has no disease, who is completely healthy and has a 3-year-old younger sibling, who has the same condition.

The patient is accompanied by a pediatrician, dermatologist, ophthalmologist, otorhinolaryngologist, neurologist and cardiologist, the latter being due to the presence of mild pulmonary valve stenosis. Also, according to the neurologist, the patient has a mental retardation and difficulty communicating through speech. Dermatological treatment consists of regular use of a body moisturizer based on urea and 2 tablets per day of Acitretin. In relation to the ophthalmological treatment, daily use of Lacrifilm eye drops is used to aid in the lubrication of the eyes and the Epitegel ophthalmic gel, which decreases possible irritations. The patient does not have any allergies and has never received local anesthesia.

General physical examination revealed dry and intensely desquamated skin, getting a brownish color, especially on the face and hands, eversion of the eyelid and lacrimation, mouth breathing, speech delay and moderate cognitive deficit (Figure 1). According to the mother's report, the desquamation worsens when the temperature is higher, that is, in the summer. It was also observed a limitation of mouth opening, due to the dryness of the surrounding skin.

Figure 1 - Physical examination.: peeling of the skin giving a brownish appearance: A) frontal view of the patient's face and eyes. B) lateral view showing the ear. C) patient's hand.

Source: Authors.

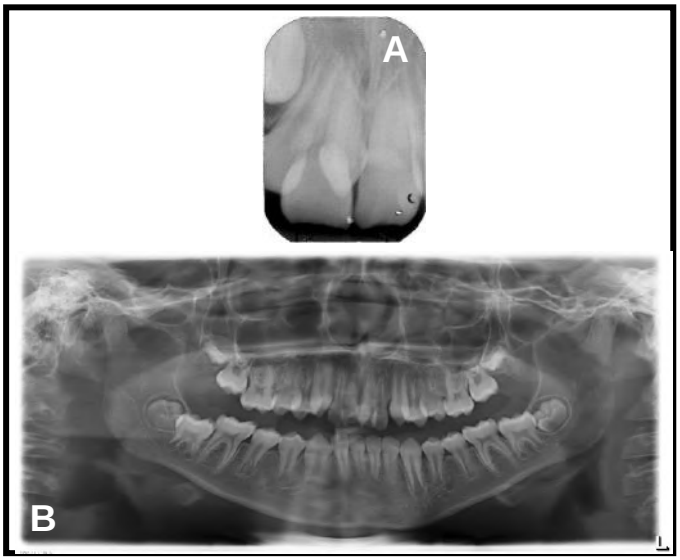
At the intraoral clinical examination, performed with a clinical mirror and applying petroleum jelly around the mouth, the patient has a mixed dentition, poor oral hygiene with biofilm accumulation in the upper central incisors, absence of carious lesions, marked overjet, inadequate lip seal, difficulty opening the mouth and cheilitis lesions in the labial commissure. It was observed the presence of a supernumerary conoid tooth in the region between the teeth 11 (right maxillary permanent central incisor) and 21 (left maxillary permanent central incisor) (Figure 2). According to the mother, this supernumerary tooth injured the patient because when occluding, that tooth was in contact with the lower lip, causing a lot of stress on the child.

Figure 2 - Intraoral clinical examination: A) biofilm accumulation in the upper central incisors and inadequate labial sealing. B) supernumerary tooth in the palate region.

Source: Authors.

A periapical radiograph of the upper central incisors was performed to visualize the correct position of the supernumerary tooth (Figure 3A). On the same day, the patient was also asked to have a panoramic radiograph for a general evaluation of the remaining teeth and structures (Figure 3B).

Figure 3 - Radiographic examinations: A) Periapical radiography of the anterior region evidencing the presence of the supernumerary tooth. B) Panoramic radiography without other changes.



Source: Authors.

After the panoramic radiography, surgery was planned to remove the supernumerary. In addition, the presence of the third molars germs and no space for its future eruption was observed, so a preventive surgery was indicated to avoid a more complicated surgery of these teeth in the future. As the difficult in the surgical management due to limited mouth opening and a non-cooperative behavior, it was initially decided to perform the procedure under conscious sedation with nitrous oxide. However, the sedation test did not have a good result since the use of mask became an additional stress factor by staying in touch with the patient's skin.

Thus, after clarification of all the information to the mother, a surgical risk was requested, to perform the surgery in a hospital environment, with a buccomaxillofacial surgeon, under general anesthesia. The patient underwent all the necessary exams and the surgical risk indicated that he was able to the proposed surgery, presenting a low risk. Nevertheless, on the scheduled day, the anesthetist of the team responsible for the procedure, interrupted the procedure in a hospital environment, contraindicating general anesthesia. The patient was already prepared for surgery and waiting to be called into the operating room but, after a new physical examination by the anesthetist. Due to the Ichthyosis Lamellar, he considered that there is a very high difficulty in manipulating the airways, which could increase the risk of bronchospasm and / or laryngospasm. Thus, it was considered prudent not to perform the procedure under these conditions, since the risk to the patient's health would be greater than the benefit.

After further discussion, the remaining alternative was removing only the supernumerary tooth under oral sedation with benzodiazepine, performed in an outpatient setting at, by buccomaxillofacial surgeon. It was given 7.5 mg of Midazolam 60 minutes prior to the procedure. After the stipulated time it was observed that the drug of first choice had a rebound effect on the patient, who was very agitated, making it impossible to perform the surgery.

The following week, we chose to perform oral sedation with 7.5mg of Lorazepam 60 minutes before surgery. Thus, the patient became sleepy and calm, allowing their stabilization in the dental chair with tissue bands and the consequent surgery under local anesthesia with lidocaine 2% + epinephrine 1:100.000.

Patient returned for postoperative review after 1 week and then after 1 month and the healing process was in accordance with normality (Figure 4) however, he presented great quantity of dental biofilm and once again the hygiene instruction was reinforced. The patient is in regular follow-up and preventive treatment in order to maintain good oral health, avoiding the need to perform invasive and highly complex procedures.

Figure 4 - Aspect of the region where the surgery was performed after a month of follow-up.

Source: Authors.

4. Discussion

Autosomal recessive congenital ichthyosis (ARCI) is a heterogeneous group of disorders that present at birth with generalized involvement of skin and lack of manifestations in other organ systems (Ramar et al., 2014). In this group, there is an ichthyosis lamellar, which involves a mutation in the TGM1 gene on chromosome 14, as well as other genes such as Ichthyin, ALOX3 / 12B, FLJ39501 and ABCA12 (Matsuno et al., 2014). According to the report, there is no history in the family and the patient being referred is the first case. However, he was taken to a geneticist, months after birth, and the mother was warned about the 25% chance of having a child with the same disease if she got pregnant again.

This disease has many cutaneous conditions, which can lead to ectropion, eclabium, rudimentary and glued ears, scarce, dry, ringy and brittle hair, hypohidrosis in varying degrees and palmoplantar keratoderma, leading to deformities such as microdactyl (Fitzpatrick et al., 1999). However, there is still a lack of information in the literature about its oral manifestations and necessary dental care.

Ramar et al. (2014) have reported that the enzyme transglutaminase plays a vital role in salivary secretion, which may reduce it, since patients with lamellar ichthyosis have an inadequate function of this gene, resulting in a xerostomia and consequently increase the risk of caries in these patients. This reduction of the salivary flow can still be attributed to the difficulty in the normal movements of the tongue and, consequently, the delay in speech, a condition present in this report.

In addition, other manifestations may include abnormal deciduous and permanent teeth (Pranitha et al., 2014). However, in this case, there is no clinical or radiological evidence of caries or other not ordinary signal, but he presented a supernumerary tooth and biofilm accumulation in the upper incisors, which can also be influenced by the fact that the patient is an oral respirator.

Contrasting with the present study, where the presence of supernumerary teeth was observed, Tewari et al. (2020) found agenesis of some deciduous teeth in a 5-year-old patient with LI. However, in the mentioned study, there is an

association between LI and rickets, so it is suggested that the latter condition is responsible for missing teeth and other factors, such as bone loss.

This case reports how difficult a dental management can be, especially in case of invasive procedures. After the unsuccess with conscious sedation with NO_2O_2, the first-choice management to carry out the propose planning would be general anesthesia but it was not possible since it would be risky due to the patient's conditions, so treatment plan was revised and only the supernumerary procedure was performed under oral sedation It should be noted that this was only possible in the second attempt.

According, Matsuno et al. (2014), respiratory insufficiency is a complication that can occur due to the restriction of thoracic expansion by the lack of elasticity of the skin. In relation to newborns with LB, administering general anesthesia is a challenge, since their breathing is often restricted and the individual is immunocompromised (Akiyama et al., 2006; Prasad et al., 1994). Yet, the most frequent causes of death in patients with ichthyosis are respiratory infections and sepsis, although a decrease in the mortality rate has been documented due to the improvement of neonatal intensive care. Strict hand washing and antisepsis measures are indicated in the handling of these patients, mainly in newborns. To prevent infection, it is also important to restrict, as much as possible, the placement of intravenous catheters and the performance of excessive examinations, which can allow the entrance of microorganisms (Taieb et al., 2002).

A recent case, reported by Reis et al. (2022) showed a 2-year-old girl with LI and several extensive caries and white spot lesions who needed general anesthesia due to behavior management problems. However, this patient did not present any risk reported by the anesthetist, and the entire procedure was successfully performed under general anesthesia. The patient reported in this article, unlike the other reported by Reis, was forbidden to undergo the procedure under general anesthesia due the risk of manipulating the patient's airways. These differences highlight how patients with LI can present varied characteristics and prognoses, why it is important to be aware of all these possibilities to create the best treatment plan.

The prognosis of lamellar ichthyosis varies with the presence or absence of associated complications. Lamellar desquamation causes a breakdown in the dermatological barrier, constituting a gateway to localized or generalized secondary infections (Crisóstomo et al., 2016). Another complication is ectropion, which can promote ulceration through continuous exposure of the cornea, and eclabium, which can make oral feeding very difficult, so that nutrition might need to be performed by venous route, nasogastric or nasoenteral tube (Aigner et al., 2008).

Ectropion, in these patients, has a frequency of 45% to 80% (Uthoff et al., 1994), most often bilateral, with more severe involvement of the lower eyelid (Singh et al., 2005), which corroborates with that seen in the present case report. The treatment of this condition is performed, initially, by eye drops and gels to lubricate the region and prevent the appearance of ulcers and possible complications.

In the present report the patient makes use of acitretin, which is an oral synthetic retinoid, acting in a way that controls deregulated cellular proliferative activity (Orfanos et al., 1997). However, it may present some adverse effects if used for a long term, such as induction of toxic effects on bone tissue, cheilitis, dryness of mucous membranes, hair loss and itching (Gulasi et al., 2016).

It’s essential to discuss the lack of knowledge of health professionals about patients diagnosed with LI. As it is a rare disease (Yang et al., 2001) the possibility of error or doubts during diagnoses and treatment choices increases. In the present case the patient had a professional permission to surgery after a surgical risk exam, but while waiting to enter the operating room, the anesthetic responsible for the case did not allow the procedure due the serious risks of bronchospasm and / or laryngospasm. This shows the disparity of knowledge among professionals in the area and the need to expand information regarding patients with LI, since even a small error can be life-threatening for these patients.

5. Conclusion

The most important of these cases is the existence of a multidisciplinary approach to define the best treatment, considering the limitations of the patient, and under what conditions this should be done so that possible complications can be avoided. In addition, it is essential that the patient is regularly under dental supervision and preventive treatment, so both, patient and responsible, can receive guidance about oral health and consequently prevent the possibility of opportunistic oral infections, dental caries, periodontal diseases, and the need for invasive treatment that may cause some risk to the patient. Finally, it is important to encourage future reports and studies of patients with IL to provide greater knowledge about this condition to all health professionals, in order to offer better treatment to these patients and minimize the chance of possible complications.

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