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Multidisciplinary care in the intensive care unit for a patient with Prader-Willi syndrome: a dental approach

Assistência multiprofissional em unidade de terapia intensiva ao paciente portador de síndrome de Prader-Willi: um enfoque odontológico

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This study was conducted at the Intensive Care Unit of the University Hospital "Maria Aparecida Pedrossian" - Universidade Federal de Mato Grosso do Sul - UFMS - Campo Grande (MS), Brazil.

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ABSTRACT

Prader-Willi syndrome is a genetic neurobehavioral disease affecting children's development and resulting in obesity, reduced height, hypotonia, endocrine disorders and cognitive deficits, which may impair oral integrity. This study aims to report on a case involving a white male 15-year-old patient with Prader-Willi syndrome whose oral examination revealed bacterial plaque, gingivitis, poor occlusion, viscous salivation and multiple lip, jugal mucosa, inserted gum and tongue ulcerations. An excision biopsy revealed oral ulcerations typical of herpes, which were considered to be likely to correlate with herpes encephalitis. This result demonstrates that a large portion of the deleterious effects of Prader-Willi syndrome can be attenuated by appropriate diagnosis and early therapeutic intervention, highlighting the role of an integrated multidisciplinary team in the development of therapeutic protocols for Prader-Willi syndrome patients.

Keywords: Prader-Willi syndrome; Herpes simplex; Dental care for person with disabilities; Morbid obesity; Intellectual disability; Intensive care; Case reports

INTRODUCTION

Prader-Labhart-Willi or Prader-Willi syndrome (PWS) was first reported in 1956, and additional syndrome details have been elucidated since then. This syndrome is primarily characterized by severe neonatal hypotonia, dysmorphic changes, retarded physical and cognitive development, early hyperphagia, obesity, sleep disorders, psychiatric and behavioral disorders, growth deficit and hypogonadism. Its population incidence ranges from 1:15,000 to 1:50,000 live births, independent of gender, race or social status.[1-5] This syndrome is considered to be a complex neurobehavioral genetic disorder caused by 15q11.2-q13 chromosome changes.[2,3]

Among oral features, prominent examples include hypoplasia of the enamel of the teeth, rampant cavities, taurodontism, delayed dental eruption, excessively worn teeth, facial hypotonia resulting in severe poor occlusion, biofilm accumulation, gingivitis, *Candida albicans* infections, geographic tongue and skin lesions.[4,5]

PWS newborns typically have difficulty in sucking due to facial

muscle hypotonia. At the age of approximately 2 to 4 years, the child begins to develop behavioral disorders and insatiable hunger (hyperphagia), which are more likely ascribable to hypothalamic disorders.[2,3]

PWS has dental aspects, as reduced salivary secretion may result from endocrine and behavioral disorders and may lead to increased susceptibility to oral infections.[4]

Bailleul-Forestier et al. emphasize the importance of early care by a multidisciplinary team to prevent PWS complications.[5] The team's dentist should be familiar with factors predisposing to opportunistic oral bacterial, fungal or viral infections, such as herpes simplex lesions.

Therefore, considering the relevance of this issue, by reporting this case, this article aims to highlight the relevance of a multidisciplinary team to the care of PWS patients.

CASE REPORT

A male, white, 15-year-old patient was admitted with dyspnea and mucopurulent discharge coughing to the emergency room of the Universidade Federal de Mato Grosso do Sul, Brazil's university hospital. The patient's mother reported the patient to have Prader-Willi syndrome and to be poorly cooperative, bedridden and easily exasperated. The patient was continuously treated with the drugs sibutramine, haloperidol and Levozine® and was monitored at an outpatient clinic. The mother reported that the patient was admitted several times due to aspiration pneumonia. The physical examination revealed periorbicular cyanosis, respiratory distress, tachypnea with auscultation of sibilation and morbid obesity (190 kg) (Figure 1).

Upon oral examination, all teeth were present with a large amount of bacterial plaque, gingivitis, poor occlusion, viscous saliva and the residual root of tooth 27 (Figure 2). The patient was diagnosed with aspiration pneumonia and remained for 16 days in a regular ward, where he was treated with ceftriaxone and clindamycin plus topical 0.12% chlorhexidine digluconate as an adjuvant for oral hygiene; this treatment was continued at home after hospital discharge, as recommended by the team's dentist.

Two days after hospital discharge, the patient was readmitted with reduced consciousness (Glasgow 6), cyanosis and a large amount of food residue in his mouth. He had a cardiorespiratory arrest, trismus and seizures and was successfully resuscitated after 5 minutes. He was referred to the intensive care unit (ICU) under mechanical ventilation with a diagnosis of septic shock secondary to nosocomial pneumonia. Respiratory and hemodynamic support, fluid resuscitation, noradrenalin, ceftazidime, sedation and midazolam plus fentanyl were provided. In addition to the medical and physiotherapy care, the patient was assessed by the ICU's team dentist, who confirmed the previous admission diagnosis and started specialized care. On the patient's 4th day in the hospital, several plain sores with erythematous bed, off-white halo and elevated borders were observed on the lips, jugal mucosa, inserted gum

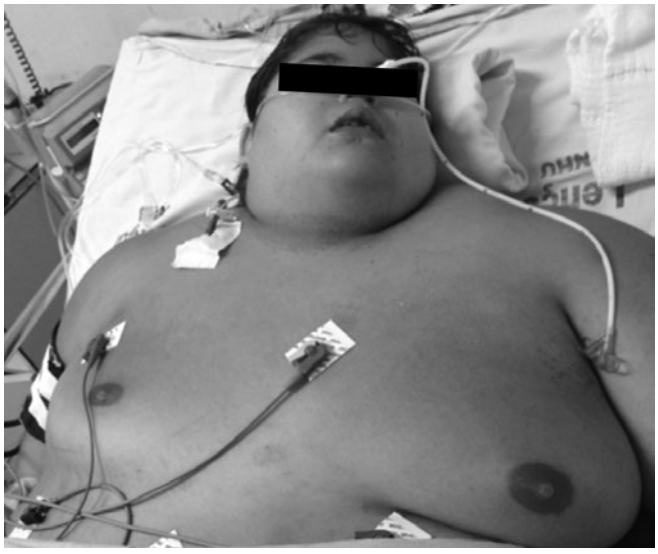

Figure 1 - Clinical aspect of a Prader-Willi syndrome patient.

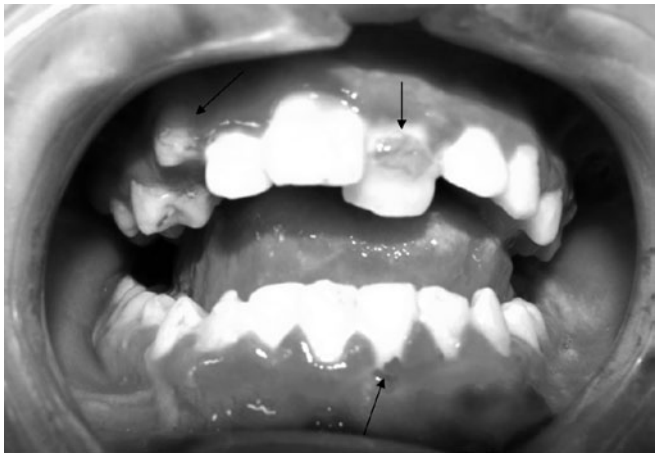

Figure 2 - Clinical aspect of the oral cavity before intubation. Presence of gingivitis, food residues and dental overcrowding (arrows).

and back of tongue, in addition to ulcerated papular lesions on the lateral aspect of tongue (Figure 3). A bedside biopsy was conducted on the 8th hospital day (Figure 4), indicating the aspects described in figure 5; these findings were confirmed on the 13th day of the patient's hospital admittance and suggested a herpes simplex (herpes ulcers) diagnosis, precluding malignant disease. Skin lesions over the sternal manubrium and subclavian regions were also observed, initially diagnosed as traumatic injuries and later considered to be skin herpes simplex lesions (Figure 6).

Empirical topical aciclovir therapy was started, with good outcome. The skin lesions, initially considered to be traumatic, were cleaned with saline and treated with topical chlorhexidine. The next days after the biopsy, the patient progressed, with worsened neurological status, deepened coma, neck stiffness, bilateral Babinski sign and generalized tonic-clonic seizures. Herpes encephalitis was suspected on the 13th day in the hospital. Head computed tomography and cerebrospinal fluid examination were requested but could not be performed due to the patient's excessive weight. Despite the treatment, which included hemodynamic and respiratory support, antibiotics, antifungals, vasoconstrictors, analgesia and sedation, in addition to nutrition support, physiotherapy and dental therapy, the patient died on the 14th day of his hospital stay.

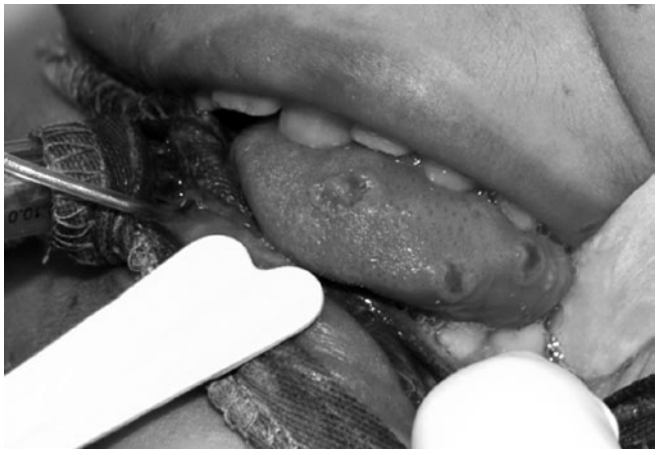

Figure 3 - Clinical appearance of tongue herpes lesions.

Figure 4 – Skin herpes lesions (arrows).

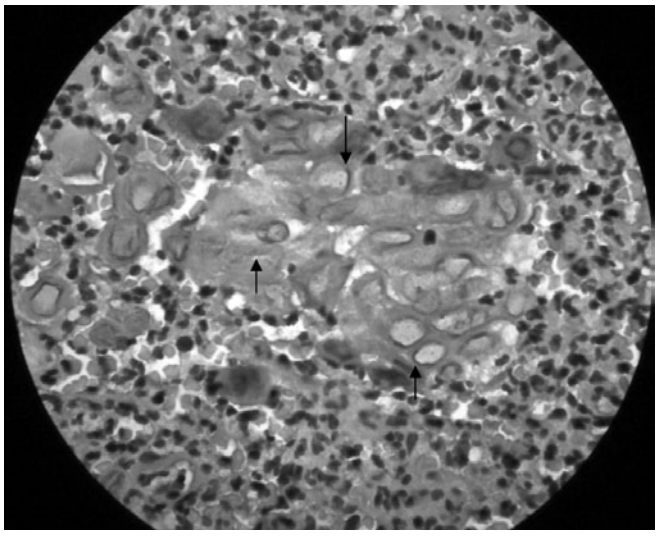

Figure 5 - Slide showing dense connective tissue with mixed inflammatory infiltration of neutrophils and lymphocytes mixed with eosinophilic material. Epithelial cells with ballooning degeneration, acantholysis, and clear and increased nuclei compatible with viral inclusions (HE, 1000x).

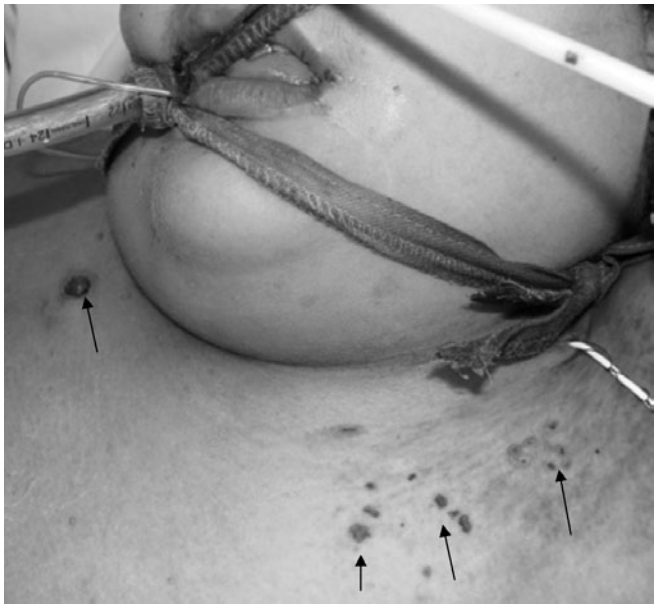

Figure 6 - Macroscopic aspect of a lesion excised from the tongue.

DISCUSSION

The involvement of a dentist as part of the ICU team is suggested as a way to address issues

related to oral health. This concern is particularly relevant for patients with special needs, hospitalized and requiring intensive care due to increased risks of colonization by organisms, especially in the oral cavity, in close contact with tubes, fixations, oral devices and others. It should be additionally considered the deficient oral self-cleaning, normally provided by speaking, chewing and appropriate salivation. As these defensive mechanisms are absent, biofilm accumulates, and the oropharynx is colonized by nosocomial organisms, which, in turn, may cause infections such as nosocomial pneumonia.[6-8]

According to Araújo et al., the nursing team may neglect oral hygiene, as it is not considered to be a priority for critically ill patients. Even the assessment of the oropharynx is considered to be difficult to conduct in mechanically ventilated patients.[7] However, there is evidence that oral hygiene with antiseptics such as chlorhexidine may prevent oral infections and their progression to the respiratory tract; this intervention is primarily aimed at preventing bacterial proliferation, therefore reducing the incidence of mechanical ventilation-associated pneumonia.[7] No side effects have been documented; this procedure is considered to be safe and well tolerated.[8]

In this reported case, poor oral hygiene was connected with the patient's overall condition. Although few reports are available on oral conditions in PWS,[4,5] the deficient cognition and self-care associated with the behavioral disorder are evidenced by the poor oral hygiene (Figure 2) and missing multidisciplinary care. This deficiency may have contributed to the opportunistic infection.

The patient's lesions led to an oral herpes diagnosis. This diagnosis was late due to the difficulty in retrieving information from the patient's family. Herpes is one of the most common human infections and manifests more severely in neonates and immunocompromised patients.[9] In such patients, herpes may range from limited vesicular lesions on the orofacial and genital regions to dissemination to the mucosa and central nervous system (CNS) involvement, that may cause sequels,[10] as in this reported case.

As the herpes virus is neurotropic, in this case, oral herpes was considered to be associated with nervous dissemination. Indeed, herpes encephalitis was the suspected cause of death (although not confirmed). This diagnosis was suspected and supported by the association between oral and cutaneous lesions and the outburst of signs suggesting involvement of the CNS: reduced consciousness level, neck stiffness, Babinski sign and generalized tonic-clonic seizures; however, the diagnosis could not be confirmed. This case supports the idea that oral assessment by a dentist, who is a part of an integrated with the ICU multidisciplinary team, is essential and complements the care of critically ill patients, especially in patients with oral conditions or who have their overall condition aggravated by oral conditions, such as patients with syndromes or coma.

In this case, the diagnosis of oral herpes resulted from the participation of a dentist in the ICU multidisciplinary team.

CONCLUSION

Several diseases may be ameliorated by appropriate diagnosis and early therapeutic and educational interventions, in addition to multidisciplinary care. This care includes the participation of a dentist as part of the ICU multidisciplinary team. Prader-Willi syndrome has specific odontology aspects, as its endocrine and behavioral disorders are associated with serious oral harms; in turn, these oral conditions may cause or aggravate the patient's clinical condition. Therefore, the development of treatment protocols and additional studies on the management of PWS patients can facilitate better care and outcomes.

RESUMO

A síndrome de Prader-Willi (SPW) é uma doença neurocomportamental genética que afeta o desenvolvimento da criança, resultando em obesidade, estatura reduzida, hipotonia, distúrbios endócrinos e déficit cognitivo que podem comprometer a integridade da cavidade oral. O presente estudo tem como finalidade apresentar um caso de paciente branco, masculino, 15 anos de idade portador da referida síndrome cujo exame clínico intra-oral evidenciou presença de placa bacteriana, gengivite, má-oclusão, salivação viscosa e múltiplas lesões ulceradas em lábio, mucosa jugal, gengiva inserida, dorso e ventre lingual e lesões papulares ulceradas em borda lateral da língua. Após realização de biópsia excisional, foi constatada a presença de lesão herpética em cavidade oral e lesões cutâneas típicas do herpes que foram associadas a possível causa de encefalite herpética. Assim, observou-se que grande parcela dos efeitos deletérios da SPW podem ser amenizados com

o diagnóstico correto e intervenções terapêuticas e educacionais precoces, sendo importante a atuação de uma equipe multiprofissional integrada e o desenvolvimento de protocolos assistenciais para melhor manejo dos pacientes portadores da síndrome de Prader-Willi.

Descritores: Síndrome de Prader-Willi; Herpes simples; Assistência odontológica para pessoas com deficiências; Obesidade mórbida; Deficiência intelectual; Terapia intensiva; Relatos de casos

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