

Multidisciplinary care of a patient with duchenne Muscular Dystrophy: case report

Cuidado multidisciplinar de um paciente com Distrofia Muscular Duchenne: relato de caso

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ABSTRACT

Duchenne muscular dystrophy (DMD) is an inherited recessive disease that impairs muscular neuromotor development and affects the daily kinetic activities of both upper and lower limbs. DMD induces systemic implications such as for overweight, gastric alterations, tachycardia, osteoporosis, respiratory insufficiency, and arterial hypertension. This case report aimed to describe the clinical profile of a DMD patient and the 5-year multidisciplinary treatment of systemic implications by dentists and other healthcare professionals. It highlights the importance of the drugs to prevent systemic diseases and adequate multidisciplinary treatment.

Keywords: Muscular dystrophy, duchenne, muscular dystrophies, neuromuscular diseases.

RESUMO

A distrofia muscular de Duchenne (DMD) é uma doença recessiva herdada que prejudica o desenvolvimento neuromotor muscular e afeta as atividades cinéticas diárias dos membros superiores e inferiores. A DMD induz implicações sistêmicas tais como sobrepeso, alterações gástricas, taquicardia, osteoporose, insuficiência respiratória e hipertensão arterial. Este relatório de caso teve como objetivo descrever o perfil clínico de um paciente DMD e o tratamento multidisciplinar de 5 anos de implicações sistêmicas por dentistas e outros profissionais da saúde. Ele destaca a importância dos medicamentos para prevenir doenças sistêmicas e o tratamento multidisciplinar adequado.

Palavras-chave: Distrofia muscular, Duchenne, distrofias musculares, doenças neuromusculares.

1 INTRODUCTION

Duchenne muscular dystrophy (DMD) is an inherited recessive disease that impairs muscular neuromotor development and affects the daily kinetic activities of both upper and lower limbs. This disease mostly affects males (1 per 3,500 births) and its defect is located on the short arm of the X chromosome at the Xp21 region (MORAES *et al* 2011; MARIANO *et al* 2009; NASCIMENTO *et al* 2015)

Diagnostic tests reveal low incidence or absence of the dystrophin protein in the sarcolemma of the muscle fibers, which favors their dystrophy and progressive locomotion issues for DMD patients. The diagnosis is based on familial history investigation, clinical examination, DNA analysis, serum creatine kinase dosage, and muscle biopsy. The replacement of necrotic muscle fibers by adipose tissue over the years results in a notably overweight appearance of patients with DMD. In addition, DMD induces other systemic implications such as gastric alterations, tachycardia, osteoporosis, respiratory insufficiency, and arterial hypertension (MARIANO *et al* 2009; MELO e CARVALHO, 2009; FRANZINI *et al* 2012) Regarding dental treatment, antibiotic

prophylaxis before tooth extraction is crucial to prevent mandibular/maxillary osteonecrosis since DMD patients usually take bisphosphonates and corticosteroids (MORAES *et al* 2011; WISKI e SOUZA, 2015; NAVARRO e LOPES, 2006)

Despite genetic advances and alternatives such as kinesiotherapy, the palliative nature of all treatments results in a poor prognosis for DMD patients, which usually need locomotion assistance and wheelchairs, (ALBUQUERQUE *et al* 2012; FORTES *et al* 2018; NASCIMENTO *et al* 2015; FONSECA *et al* 2007). Respiratory insufficiency is the major cause of death in patients with DMD; thus, during the course of the disease, patients can have cough and difficulty in breathing up to respiratory infections and cardiac involvement.

The following case report aimed to describe the clinical profile of a DMD patient and the 5-year multidisciplinary treatment of systemic implications by dentists and other healthcare professionals.

2 CASE REPORT

A 6-year-old brown-skinned boy and his mother attended the Integrated Service of Oral Diagnosis and Dental Care for Special Patients (SIDOPE) of the Federal University of Pará (UFPA) and complained about decayed teeth and chewing difficulties (Fig. 1). After a detailed anamnesis, the clinical examination revealed some caries and the need for tooth #82 extraction; in addition, a fissure at the labial commissure region was reported by the mother as a result of trauma. The treatment plan involved tooth cleaning, caries removal, and fillings, in addition to tooth #82 extraction. Moreover, several routine lab exams such as blood count, coagulation profile, serum glutamic-oxaloacetic transaminase (SGOT), serum glutamate pyruvate transaminase (SGPT), triglycerides, and echocardiogram were requested to confirm the clinical suspicion of a syndromic alteration.

The high values of SGOT and SGPT evidenced a liver disease that was under investigation; in addition, altered values of Aldolase A/B revealed a severe skeletal muscle alteration that also indicates hepatic disorder. The altered cholesterol and Hydroxyvitamin D levels suggested that the patient has been gaining weight; thus, his mother was instructed to provide a low-carb diet based on fruits and vegetables to prevent obesity, arterial hypertension, diabetes, and early infarction.

Since the diagnosis of DMD, the patient has received dental treatment in addition to a multidisciplinary follow-up (therapist, nutritionist, geneticist, psychologist, and

speech therapist) over the course of 5 years; however, a remarkable muscular involution, calves dystrophy, significant weight gain, and locomotion difficulty have been observed (Fig. 2). Although one of the most common systemic implications, the patient did not present significant cardiovascular alteration, was classified as ASA surgical risk and preventive drugs were prescribed. A panoramic radiograph revealed a mixed dentition with preserved teeth, bones, and muscles (Fig. 3).

Figure 1. Patient with regular teeth and anterosuperior diastemas.



Figure 2. After a 5-year follow-up, the patient needed to use a wheelchair due to significant weight gain, muscular involution, and calves dystrophy.



Figure 3. Panoramic radiographic revealed regular structures of the oral cavity.



3 DISCUSSION

Considering that the understanding of both clinical and functional profiles is of fundamental importance to provide specific healthcare for DMD patients, this case report described potential systemic alterations found in the current literature that can influence dental treatment. It must be emphasized that the participation of dentists in the multidisciplinary treatment of patients with DMD is essential for adequate healthcare. There is scarce evidence in the literature regarding the oral implications presented by these patients; however, dentists must remain attentive and cautious. This report of a 5-year follow-up of a DMD evidenced locomotor difficulties that confirmed the initial clinical diagnosis. Therefore, the dentists requested routine lab exams every six months in addition to regular medical updates by cardiologists, therapeutics, and speech therapists. This constant interaction between dentists and other healthcare professionals is encouraged to improve treatment. It is worth mentioning that this case report described a very atypical profile of DMD patients since he presents only mild systemic alterations for his age; however, continuous use of palliative drugs was prescribed to mitigate further implications. Dentists must be aware of their importance for the early diagnosis of patients with DMD as well as to provide adequate care.

4 CONCLUSION

DMD patients can present several functional alterations during dental care and need specific management; in addition, their family members must be properly instructed about the limitations of this syndrome. Therefore as health promoters, dentists must keep a constant dialogue with other healthcare professionals to establish a valuable multidisciplinary treatment.

INFORMED CONSENT STATEMENT

The authors have obtained the individual's and his mother signed informed consent statement to publish this case report and respective images.

CONFLICT OF INTEREST STATEMENT

The authors declare no proprietary, financial, or other personal interest of any nature or kind in any product, service, and/or company.

ETHICAL APPROVAL STATEMENT

This case report did not require the approval of a formal ethics committee since an informed consent was obtained at the time of the initial dental care.

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