RARE DISEASES AND THEIR ORAL CLINICAL MANIFESTATIONS IN TWO HOSPITAL CENTER IN YAOUNDE

SUMMARY

Introduction

In 2012, rare diseases occurred in one case out of two thousand people in the world. These rare diseases are mainly of genetic origin and have mostly buccal manifestations, hence motivating consultations. These oral lesions can be useful in the diagnosis and monitoring of these rare diseases in people living with them. Globally in Africa and particularly in our context, these events are not yet very well known. The purpose of this work was to determine the epidemiological and clinical aspects of oral manifestations of rare diseases based on the rare disease groups found in our environment, the affected tissue and the type of dentition.

Methods

We conducted a cross sectional, prospective and descriptive study over a four month duration (January 2016- April 2016) within the service of neuropaediatric of two different hospitals: the Obstetrics, Gynecology and Paediatric Hospital of Yaounde and the Chantal Biya Mother and child Care Foundation. We recruited patients diagnosed with these rare diseases who complained or not of oral symptoms. Once a participant included, the principal investigator proceeded to his identification and the clinical examination of the oral cavity in search of lesions associated with the disease group. The neurologist later conducted a general review and classification of the patient according to the clinical groups (according European Organisation of Rare Diseases, Orphanet and SMITH'S classifications). At the end of this general examination, the buccal diagnosis found were recorded on our data sheet. Statistical analysis was performed using SPSS Version 18.0 software and the chi-square test allowed us to make associations. The significance level was set below 5 %.

Results

Totally 37 patients were examined, out of which 36 patients were carriers of at least one oral manifestation (36/37 or 97.2 %). Patients with oral lesions had predominantly a

temporary dentition (59.45 % or 22/37) with a sex ratio of 1: 1.1.5. The most represented disease group was that of the hamartoses with 14 cases/ 37 is 37.83%. Patients aged between 17 months and four years were the most represented (54.05 % or 20/37).The most represented region of origin was the west region (19/37 or 51.4 %). Patients having a temporary dentition showed more abnormalities in dental shape and position (tooth taper 7/22 or 32%), while those having a permanent dentition were more affected by caries(7/10 or 70%) and dental structure abnormalities (tooth wear 4/10 or 40%)..

A significant relationship existed between the type of teeth , the occurrence of dental structure (P = 0.001) and tooth number (P = 0.018) .From the same difficulty chewing (P = 0.023) and suction (P = 0.033) were related to disease groups.

Conclusion

We conclude that rare diseases present in our environment, manifest orally and this varies between the disease groups, but also according to the type of dentition. Functional abnormalities of the oral sphere of these patients are associated with dental damage.

References

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