

ORAL CAVITY RARE LESIONS: 15 YEARS CASE HISTORIES

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SUMMARY

Oral cavity rare lesions: 15 years case histories.

Objectives. Oral cavity rare diseases include a various group of uncommon morbid conditions. For this reason they are often called "orphan diseases", as they are not interesting for research and the description of their natural history is not easy. The aim of our study is to analyze the prevalence and the distribution of oral cavity rare diseases in order to increase their knowledge and allow a fast therapeutic approach.

Methods and material. 3144 patients took part to our study, they were chosen according to specific criteria and included in a experimental program; they all were prepared for oral biopsy surgery at Fatebenefratelli Hospital - Tor Vergata University of Rome. Following the results of the histological diagnosis, patients have been grouped.

Results. From 1996 to 2010, we observed 1635 men and 1509 women, average age was 53 years, higher for women (55y.) and lower for men (52y.).

Conclusions. Nevertheless the low level of accordance and the difficulty in description of natural history of diseases reported in literature, we can conclude that, according to our study the onset of rare diseases shows a percentage of appearing statistically significant.

Key words: incisional biopsy, excisional biopsy, rare lesions.

RIASSUNTO

Lesioni rare del cavo orale: 15 anni di casistica.

Obiettivi. Le lesioni rare della cavità orale sono un gruppo vario di non comuni condizioni morbose. Per questo motivo sono spesso chiamate "malattie orfane", in quanto non sono interessanti per la ricerca e perché la descrizione della loro storia naturale non è facile. Lo scopo del nostro studio è quello di analizzare la prevalenza e la distribuzione delle malattie rare del cavo orale, il tutto al fine di aumentare la loro conoscenza e di permettere un rapido approccio terapeutico.

Metodi e materiali. 3144 pazienti hanno preso parte al nostro studio, sono stati scelti in base a criteri specifici e inseriti in un programma sperimentale, tutti i pazienti sono stati sottoposti a chirurgia orale bioptica presso l'Ospedale Fatebenefratelli - Università Tor Vergata di Roma. I pazienti, a seguito dei risultati della diagnosi istologica, sono stati poi divisi nei diversi gruppi di patologie.

Risultati. Dal 1996 al 2010, abbiamo osservato 1.635 uomini e 1.509 donne, l'età media è stata di 53 anni, più elevata per le donne (55y.) ed inferiore per gli uomini (52y.).
Conclusioni. Il nostro studio, nonostante il basso livello di conformità e la difficoltà nella descrizione della storia naturale delle malattie rare del cavo orale, ci porta a concludere che la loro percentuale risulta essere statisticamente significativa.

Parole chiave: biopsia incisionale, biopsia escissionale, lesioni rare.

Introduction

Oral cavity rare diseases include a various group of uncommon morbed conditions. Biopsy represents the most useful and less invasive method for the diagnosis of rare diseases. It is a common procedure in surgery, consisting in taking a tissutal fragment from a living organism, to get a microscopic examination. This operation is frequently used in the oral cavity, for purpose of histopathologic diagnosis of an unknown lesion. Biopsy is also important for the forensic medicine, because the sample or slide can be rivalued subsequently, if needed. Furthermore biopsy can be used to evaluate a wide benign lesion, when the patient refuses its complete removal for aesthetic and functional consequences of the surgical operation.

Material and methods

The aim of our study is to analyse prevalence and distribution of rare pathologies in the oral cavity by a retrospective and prospective study on histological reports taken by incisional and excisional biopsies from 1996 to 2010. An Univariate analysis was performed to evaluate the prevalence of pathologies. The Chi-squared test was used to compare different groups and the t-test between means. A P-value of <0.05 (two tailed) was considered statistically significant. All the analyses were performed with SPSS for Windows (release 9.0; SPSS Inc., Chicago, IL). We used an experimental program on 3144 patients prepared for oral cavity biopsy surgery in the Fatebenefratelli Hospital oral surgery department - Tor Vergata University of Rome. All the found pathologies, were divided into different groups according the histopathologic diagnosis. The patients participating to our experimental program were chosen according to the following criteria:

1. Patients affected by a pathology needing a biop-tical removal
2. Patients without absolute contraindications to sur-gical operation

3. Patients not treated in the previous 5 years by che-motherapy and/or radiotherapy in the district cer-vical-maxillary-facial.

The day of the operation we did:

1. the recording personal data patient's;
2. medical and odontoiatric anamnesis;
3. evaluation of removal location;
4. the recording in department;
5. photograph of the lesion.

After this, the surgical operation was done, (in ad-mission or in day-hospital).

All the biopsies were made using a cold blade scal-pel, because the electric lancet could cause electric-cauterization and perilesional tissutal edges altera-tion.

The surgical technique for the biopsy foresaw:

1. Careful disinfection of perioral cutis with a io-dine-povidone solution;
2. Oral cavity disinfection with mouthwash clore-ridine 0,2% for 1 minute;
3. Local anaesthesia with carbocaina 3% without va-soconstrictor at about 1 cm from site removal;
4. The interested area was stabilized by a suture in-side the lesion keeping the thread stretched by an hemostatic forcep;
5. Incision by Bard-Parker n° 5 lancet with 15T bla-de, doing an elliptic wedge included between two curved incisions converging under the lesion in the deep healthy tissue (where possible);
6. The safety edge in the clinical benign lesion was 2-3 mm; while in the possible malignant lesions the edge was increased to 8-10 mm;
7. After biopitic tissue removing, the surgical wound edges were sutured to help healing by first in-tention. The suture thread used was a Vicryl Plus 4/0 with SH-1 plus ½ circle cylindrical section needle;
8. The surgical fragment was immediately put in a hermetic box with closing screws, that contained formalin in buffered solution 10% with a for-maldehyde quantitative of 4% stabilized at no more than 5° C temperature.

The patient was dismissed after a careful and pro-tracted control of the hemostasis and of their gene-ral condition (or dismissed the following day in case of difficult operation); we recommended the patient doing mouthwashes with a hydrogen peroxide so-

Table 1 - Rare diseases we found in our study.

Actinomycosis (5)	Condyloma Acuminatum (9)	Paget's disease (6)
Adenocarcinoma (15)	Erythroleukoplakia (7)	Neurinoma (8)
Adenocystic c. (7)	Bowen's disease (7)	Nevus Spongiosus Albus (6)
Intrabone c. (11)	Histycytosis X (5)	Odontoameloblastoma (6)
Keratoacanthoma (9)	Non Hodgkin Lymphoma (8)	Ameloblastic Sarcoma (5)
Epidermoid Cyst (4)	Small Cell c. (4)	Malignant Teratoma (3)
Globulo-maxillary Cyst (5)	Odontogenic Myxoma (8)	Squamosus Odontogenic Tumor (5)
Bone Cyst Aneurismatic (6)	Wegener's granulomatosis (8)	

lution 3% 10 Vol, diluted in 50% of water, twice a day for 15 days, from the day after the operation. We prescribed antibiotic therapy for 5-6 days. After 12 days, we controlled the patients and removed the stitches, even if the suture thread was absorbable. After about 15 - 21 days the patients went back for another control and we evaluated the anatomo-pathological report and according to it we decided the most suitable therapeutic program.

Results

At the end of our study the 5% of whole sample, 157 subjects (Table 1), showed diseases with a prevalence lower than 5 for 10.000 inhabitants. In all observed lesions, we noticed the presence of 23 different rare lesions grouped in three classes: infectious and inflammatory lesions (14 cases) (Table 2), dysplastic lesions of benign origin (44 cases) (Table 3), neoplastic lesions of malignant origin (99 cases) (Table 4). In our study there weren't differences about sex, of 157 subject, 80 men and 77 women, while the rare lesions prevalence increased with age, except for patients over 81. From 1996 to 2010 rare diseases prevalence increased from 1.9% to 8,9% with an odd

Table 2 - Inflammatory and infective lesions.

Inflammatory and infective lesions	N° Pz
Actinomycosis	5
Condyloma Acuminatum	9

Table 3 - Dysplastic - neoformative lesions.

Dysplastic - neoformative lesions	N° Pz
Keratoacanthoma	9
Epidermoid Cyst	4
Fissure Cyst	5
Bone Cyst Aneurismatic	6
Weneger's Granulomatosis	8
Paget's Disease	6
Nevus Spongiosus Albus	6

Table 4 - Neoplastic lesions.

Neoplastic lesions	N° Pz
Adenocarcinoma	15
Adenocystic Carcinoma	7
Intrabone Carcinoma	11
Erythroleukoplakia	7
Bowen's disease	7
Histycytosis X	5
Non-Hodgkin Lymphoma	8
Small Cell Carcinoma	4
Odontogenic Myxoma	8
Neurinoma	8
Odontoameloblastoma	6
Ameloblastic Sarcoma	5
Squamosus Odontogenic Tumor	5
Malignant Teratoma	3

ratio of 5 adjusted for sex and age (Fig. 1). The highest prevalence observed of the rare lesion was in 2003, when 17 subjects, on 350 evaluated, showed rare lesions. During years trend was not regular because there were some sudden decreases (Fig. 2).

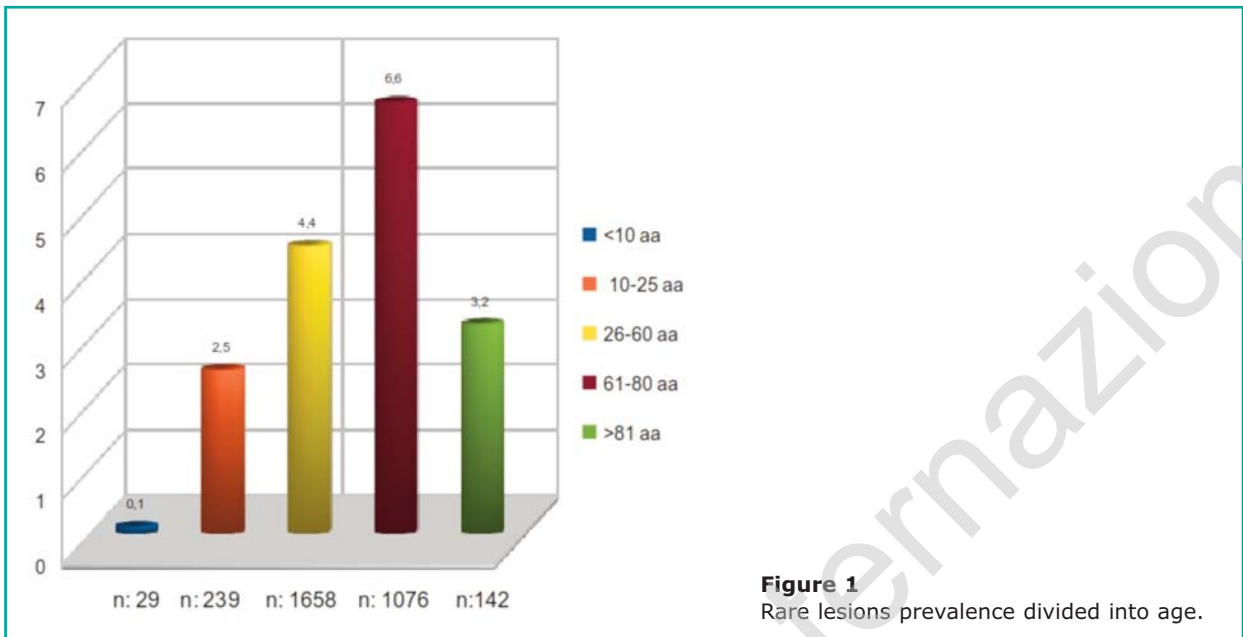


Figure 1
Rare lesions prevalence divided into age.

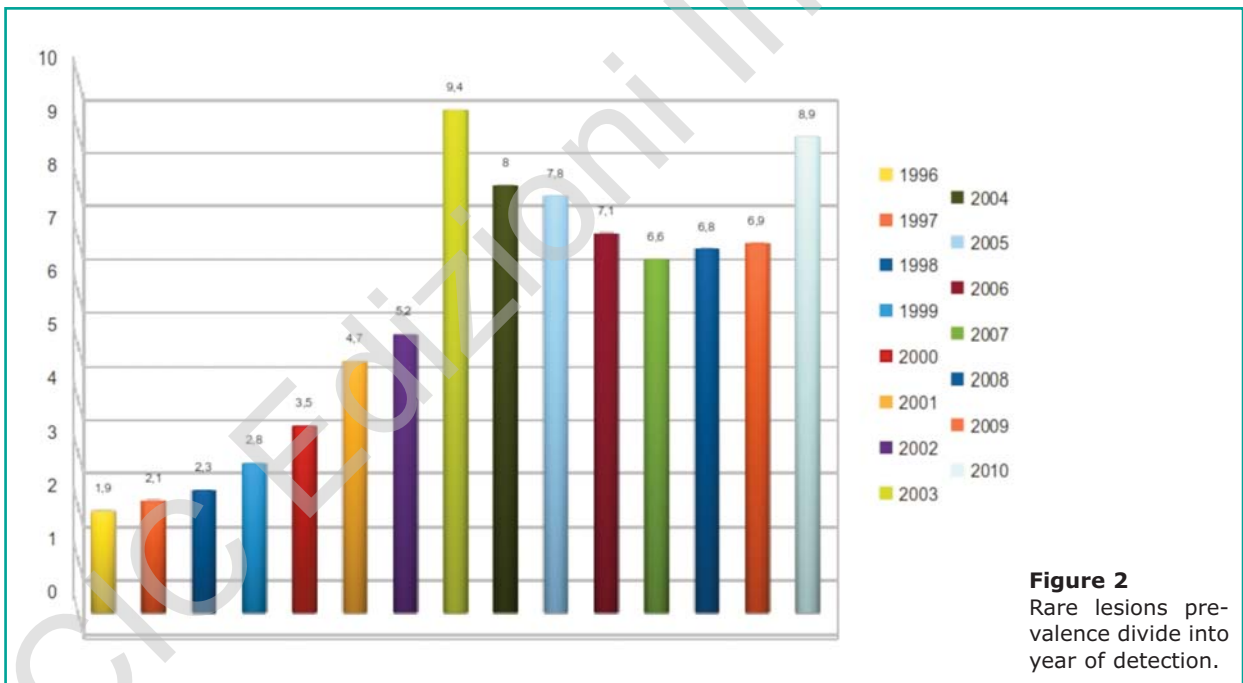


Figure 2
Rare lesions prevalence divide into year of detection.

Discussion

From data come out of the international literature revision, rare diseases are not very frequent unhealthy conditions. They are also called orphan diseases, be-

cause not very interesting for experimental research. The rarity of diseases causes greater difficulty in describing the natural history of pathology with the possible variants and planning some valid clinical research. In other words, the low prevalence causes less knowledge than the possible one (2). Therefore we spe-

ak about not very examined diseases and frequently without adequate therapy. So patients suffer a double damage: at first suffering an almost always severe pathology and then because they aren't adequately recognized, diagnosed and treated. Because of these considerations they have begun giving some resources to help such pathologies. This process started in USA and then involved all western countries and WHO. Because of these system it's necessary to know exactly how much all rare diseases affect the population and interest health services. The rare diseases group includes several different diseases, because these pathologies aren't a well known nosologic group, but a heterogeneous group characterized only by low prevalence (3). There isn't an exact definition of rare diseases. The congress USA gives the only official definition of rare diseases that is a disease affects 200.000 persons at the most, therefore one person every 1200 (4). The action program of the European community for rare diseases 1999-2003 defines rare the diseases with a prevalence lower than 5 for 10.000 inhabitants (5,6). The aim of our work is to analyze the epidemiological characteristics of different oral cavity rare diseases, gathered by a bioptic exam with an histological interpretation.

Conclusion

We think that the increase of the average life and heterogeneity of population, owing to the emigration, will cause increase rare diseases in the future (7). Biopsy allows a right recognition and an effective differential diagnosis with a moderate biological and healthy price (8). In conclusion we can affirm that biopsy is a methodology clinical-diagnostics essential

for diagnosis of oral cavity rare diseases.

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