Original article

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Orofacial manifestations of scleroderma. A literature review

Manifestations orofaciales de la sclérodermie. Revue de la littérature

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Summary

Introduction. Scleroderma is a rare disease of the connective tissue (50 to 200 patients/1 million people; 60,000 patients in France). We conducted a literature review about the orofacial manifestations of scleroderma that have been little studied.

Material and methods. The 45 articles found in 6 different databases by using the keywords "scleroderma", "systemic sclerosis", "oral medicine", "face" and published between 1944 and 2016 were selected, for a total of 328 patients.

Results. A total of 1187 orofacial manifestations of scleroderma were identified, occurring mainly in women (84.5%) with a mean age of 40.2 years, 10 years on average after the first manifestation of the disease. The main ones were limitation of mouth opening (69.8%), widening of the periodontal ligament (67.3%), xerostomia (63.4%), telangiectasia (36.2%) and bone lesions (34.5%). Dental root resorptions, pulp and nose calcifications were also reported but with no evident link with scleroderma.

Discussion. Orofacial manifestations of scleroderma are probably more common than reported. They mostly affect women with a mean age of 40. The most common oral manifestations are limitation of mouth opening, widening of the periodontal ligament and xerostomia. Because of the handicap they may be responsible for, these manifestations must be detected early in order to

Résumé

Introduction. La sclérodermie est une maladie rare du tissu conjonctif (50 à 200 patients/million d'individus ; 60 000 patients en France). Nous avons mené une revue de la littérature concernant les manifestations orofaciales de la sclérodermie qui ont été peu étudiées.

Matériels et méthodes. Les 45 articles trouvés dans 6 bases de données différentes en utilisant les mots-clés *scleroderma*, *systemic sclerosis*, *oral medicine* et *face* et publiés entre 1944 et 2016 ont été sélectionnés, totalisant 328 patients.

Résultats. Un total de 1187 manifestations orofaciales rattachées à la sclérodermie ont été répertoriées, touchant majoritairement les femmes (84,5 %) âgées de 40,2 ans en moyenne et survenant en moyenne 10 ans après la première manifestation de la maladie. Les principales manifestations étaient la limitation d'ouverture buccale (69,8 % des cas), l'élargissement du ligament alvéolo-dentaire (67,3 %), la xérostomie (63,4 %), les télangiectasies (36,2 %) et les atteintes osseuses (34,5 %). Des résorptions radiculaires, des calcifications pulpaires et nasales ont été retrouvées mais sans lien évident avec la sclérodermie.

Discussion. Les manifestations orofaciales de la sclérodermie sont probablement plus nombreuses que les cas publiés. Elles concernent les femmes de 40,2 ans en moyenne. Les manifestations les plus fréquentes sont la limitation d'ouverture buccale, l'élargissement du

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prevent from functional impairments and from dental and periodontal lesions.

Keywords: Systemic sclerosis, Scleroderma, Oral medicine, Face

ligament alvéolo-dentaire et la xérostomie. En raison de leurs caractères potentiellement invalidant, ces manifestations doivent être dépistées précocement, pour faciliter la prévention des troubles fonctionnels et des lésions dentaires et parodontales.

Mots clés : Sclérose systémique, Sclérodermie, Oral, Visage

Introduction

Systemic sclerosis, also known as scleroderma (SD), is a chronic connective tissue disease characterized by skin fibrosis and potentially extremely serious vascular and visceral lesions. Matsuis described the disease in 1924 and indicated for the first time a possible visceral involvement [1].
SD affects the quality and mobility of the skin, particularly at the extremities. Raynaud's phenomenon, characterized by a painful digital ischemia, is one of the earliest symptoms to be observed [2].
SD can involve lungs, heart, kidneys, gastrointestinal tract and bones [3].
Numerous oral manifestations (OM) of SD have been reported since 1949, mainly through clinical cases. Tongue rigidity and hardening of the skin of the face, leading to the typical "mask appearance" [4], were first described. Limitations of mouth opening and mandibular bone resorption were also reported.
Our aim was to conduct a literature review about the OM of SD.

Material and methods

A systematic review was performed in the 6 different databases accessible from our university (PubMed, Scopus, Science Direct, Wiley Library, Google Scholar and Dentistry & Oral Sciences sources) using the following keyword combination: "scleroderma" ± "systemic sclerosis" ± "oral medicine" ± "face".
Because of the lack of meta-analyzes and the low number of publications on OM of SD, all the 217 articles matching with the keywords were initially included *(fig. 1)*. Among these 217 articles we selected the articles published between 1944 and 2016 dealing specifically with OM of SD leading to a set of 45 articles (*table I*, [5–25]).
The number age and sex of patients and the time elapsed between diagnosis of SD and OM were recorded and analyzed.

Results

A total number of 392 patients (mean age: 40.2; extreme: 12–62) was found. Sixty-four cases were poorly described and excluded resulting in 328 patients in whom 1187 OM were reported *(table II)*.
OM were observed mainly in women (284/328; 87.1%) and occurred 10.1 years on average (1–33 years) after the diagnosis of SD.
Limited mouth opening (LMO), defined as less than 40 mm, was found in 229 cases (69,8%), mainly in women (88.6%). It occurred early (1 year on average) after the diagnosis of SD. The youngest patient with LMO was 12 years old. LMO lasted up to 33 years after the diagnosis.
Widening of periodontal ligament was found in 67.3% of the cases, mainly in women (89.1%), and occurred 1 to 33 years after the diagnosis of SD.
Xerostomia was found in 63.4% of the cases and seemed to appear later than the previous OM, 4 years on average after the diagnosis of SD. It concerned mainly women (76.9%).
Telangiectasia of the oral mucosa was found in 36.2% of the cases, mainly in women (84%).
Four hundred and ten bone damages were found in 34.5% of the cases, mainly in women (83.9%), and concerned the mandibular angle (49.1%), the mandibular coronoid process (36.9%), the mandibular condyle (22.6%), the mandibular ramus (15.8%), and the zygomatic arches (0.6%). They occurred 7 to 33 years after the diagnosis of SD.
Other symptoms have been occasionally reported but were not included in the OM of SD because of the lack of proof that

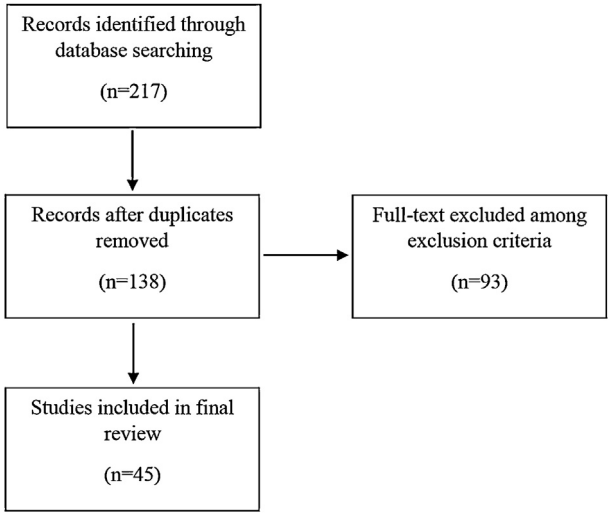

Figure 1. Process of article selection for this study.